SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES AND TECHNOLOGY

THIRUVANANTHAPURAM

CLINICAL FEATURES, IMAGING FINDINGS AND MANAGEMENT OF SPINAL VASCULAR MALFORMATIONS

PROJECT REPORT FOR

DM NEURORADIOLOGY

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CERTIFICATE

*This is to certify that the work contained in this thesis have been carried out by **Dr. Jitender Saini** in the Department of Imaging Sciences and Interventional Radiology, Sree Chitra Tirunal Institute of Medical Sciences and Technology, Trivandrum, during his rotatory postings as per schedule, under my guidance and is to my satisfaction.*

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DECLARATION

I, **Dr. JITENDER SAINI** hereby declare that I have actually carried out the project ***"Clinical Features, Imaging Findings And Management Of Spinal Vascular Malformations"*** independently under supervision and guidance in this institute.

Date – Sep 2009.

THIRUVANANTHAPURAM

Dr. JITENDER SAINI

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ABBREVIATIONS

AVM	-	arteriovenous malformation
AVF	-	arteriovenous fistula
CT	-	computed tomography
CISS	-	constructive interference in steady state
DSA	-	digital subtraction angiography
FOV	-	field of view
HHT	-	hereditary hemorrhagic telangiectasia
IBCA	-	isobutyl 2-cyanoacrylate
MRI	-	magnetic resonance imaging
MRA	-	magnetic resonance angiography
NBCA	-	n-butyl 2-cyanoacrylate
PMF	-	perimedullary fistula
PVA	-	poly vinyl alcohol
SAMS	-	spinal arteriovenous metameric syndrome
SAH	-	subarachnoid hemorrhage
SAVS	-	spinal arteriovenous shunt
SCAVM	-	spinal cord arteriovenous malformation
SCAVF	-	spinal cord arteriovenous fistula
SDAVF	-	spinal dural arteriovenous fistula
T2W	-	T2-weighted
T1W	-	T1-weighted
TSE	-	turbo spin echo

INDEX

Introduction

INTRODUCTION

Spinal vascular diseases are a rare form of vascular neurologic pathologic conditions. Most common entities include spinal dural arteriovenous fistulas (SDAVF), perimedullary fistulas (PMF), spinal cord arteriovenous malformation (SCAVM), epidural and paraspinal arteriovenous malformation (AVM) and cavernomas. They may be congenital like SCAVM or acquired like SDAVF. These lesions can cause acute, subacute, or chronic spinal cord dysfunction, which may be either complete or partial. The variability in clinical presentation reflects differences in pathophysiology of spinal cord vascular malformations.[1,2]

Due to their low incidence, there is lack of large-scale clinical studies assessing various aspects of diagnosis, natural history, and management. Although spinal vascular malformations are pathologically and biologically similar to their counterparts within the brain and cranial meninges, often their clinical impact, both in terms of natural history and response to therapeutic intervention, has been comparatively worse. While acute manifestations of spinal vascular malformations typically lead to a diagnosis early in the course of the disease, the subacute venous congestion might lead to non specific neurological symptoms which in turn delay proper diagnosis. Clinical manifestations are often non specific, leading to delay in definite diagnosis. This leads to prolonged ischaemia to the cord and irreversible damage.[1, 2]

Although digital substraction angiography remains the gold standard, MRI constitutes first line investigation in a suspected case of spinal vascular malformation of the spine as it shows both abnormal flow voids as well as cord signal changes. Associated lesions or evolutive complications, such as hematomyelias, oedema, thrombosis, or cord atrophy are also well depicted on multi-planar sections in MR imaging Magnetic resonance angiography may also be useful in determining the level of feeding artery and characterizing the lesion especially in dural fistula, however its role in intradural shunts remains debatable.[3, 4].

Treatment modalities available include endovascular treatment and surgery. There is no role of radiotherapy in treatment of spinal vascular lesions.[4] In our centre endovascular treatment is the first line treatment for all the cases of spinal vascular malformation.

Our centre is a major tertiary referral centre for management of all types of neurovascular disorders. Our department of IS & IR has been actively involved in the endovascular management of various neurovascular lesions. We hereby review our experience in the endovascular management of spinal vascular malformations over the last 25 years.

Review of Literaure

REVIEW OF LITERATURE

Spinal vascular malformations are rare and still underdiagnosed entities that, if not treated properly, can lead to considerable morbidity with both acute and progressive spinal cord symptoms.

HISTORICAL PERSPECTIVES

Hebold and Gaupp are credited with the first descriptions of isolated spinal cord vascular malformations in 1885 and 1888, respectively.[5,6] Gaupp reported on the case of a 45-year-old woman with slowly progressive lower extremity weakness and paresthesias attributed to "a large varix of veins arising from the pia which pressed upon and flattened the spinal cord." They referred to these lesions as hemorrhoids of the pia mater and recognized them as potential cause of sub arachnoid hemorrhage. First clinical description of the spinal vascular malformation was given by Berenbruch[7] in 1890. Over the years many case reports appeared in literature describing the clinical features of the spinal vascular malformations, which mimicked those of spinal tumors. Surgeons performed exploratory laminectomies in patients with progressive paraparesis who were subsequently found to have vascular malformations. The first attempt at surgical treatment of these lesions was made by Krause[8] in 1906 in a 34-year-old man who had presented with a 7-year history of progressive lower extremity symptoms. The first successful operation for a spinal cord vascular malformation was reported by Elsberg[9] in 1916. He operated on 6 patients of which two patients showed significant improvement in symptoms. Foix and Alajouanine[10] described the entity of subacute or acute progressive myelopathy associated with abnormal spinal cord vasculature and necrosis in the absence of hemorrhage. Several years later, Lhermitte et al[11] noted the association between this syndrome and an AVM in the spinal cord. Wyburn-Mason[12] realized, for the first time, the similarity of two patients reported by Foix and Alajouanine[10] and proposed that these cases probably represented the endstage progression of the same vascular lesion. Introduction of Lipiodol myelography aided significantly in the diagnosis of these lesions. In the mid-1920s, several patients who underwent myelography, which showed a block to contrast presumably due to a spinal cord tumor, were found, instead, to have vascular lesion at surgery. Perthes J[13] was the first surgeon who preoperatively recognized an spinal cord vascular malformation (SCVM) by myelography and successfully excised the lesion.

Though cerebral angiography was pioneered by Egas Moniz in 1927, 30 years elapsed before spinal angiography became a critical diagnostic and research tool to identify SCVMs. The first cases of a cervicomedullary AVM diagnosed by vertebral angiography were reported in 1958 by Hook and Lidvall[14]. A few years later Rend Djindjian and associates[15] introduced spinal aortography in France at the Lariboisiere Hospital. In 1965, Doppman and DiChiro[16] took the lead in the United States by demonstrating the importance of subtraction angiography in the diagnosis of SCVMs.

THE ARTERIAL CIRCULATION OF THE SPINAL CORD[17,18]

Unlike the brain, which receives its blood supply from arteries having a common origin in the aortic arch, the spinal cord is supplied by arteries of diverse origin The arterial architecture is organized around horizontal axes; the radicular arteries, which anastomose with the longitudinal axes, the anterior, and posterior spinal axes. The density of arterial supply to the spinal cord varies according to each level. The cervical and lumbosacral segments are richly supplied whereas the thoracic segment has few arteries.

Radicular arteries: There are 33 pairs of radicular arteries. Each radicular artery penetrates the spine following the spinal nerve and then its roots. It has an ascending movement whose obliqueness depends on the spinal level in question. Each radicular artery penetrates the spine following the spinal nerve and then its roots. It has an ascending course whose obliqueness depends on the spinal level in question. Certain arteries regress and others hypertrophy resulting in the adult disposition. The radicular artery penetrates the dura mater, which it supplies with blood, and disappears quite deeply in the vertebral canal supplying blood to the roots to the spinal nerve, the pia mater, and in certain cases the spinal cord. The radicular arteries have been classed into three groups: 1) radicular arteries, 2) radiculopial arteries and 3) radiculomedullary arteries.

1. **Radicular arteries** represent the minimal contribution of the embryonic segmental system to the neural crest derivatives. The arteries disappear in the roots before reaching the pia mater and the spinal cord. They are not involved in supplying blood to the spinal cord.

2. **Radiculopial arteries** correspond to a group of arteries that reach the spinal cord pial network. They supply the spinal nerve root and run ventral to its anterior root or ventral to its dorsal root to reach the spinal cord surface. They participate in the pial network, which gives off radial perforators centripetally to the spinal cord. Radiculopial arteries do not participate in the supply of the anterior spinal axis but may be anastomosed with the anterior spinal axis. They participate in the posterior arterial axis. The radiculopial arteries vary in number between 10 and 20. Their caliber is much smaller than the anterior ones. It is often difficult to clearly identify their course because they divide into numerous branches in the pial network.

3. **Radiculomedullary arteries** represent the arteries that contribute in both the segmental and longitudinal supply to the spinal cord. The

radiculomedullary arteries vary in number between five and ten. They participate in constituting the pial network before feeding the ventral spinal axis. The caliber of the anterior radiculomedullary arteries varies. It is smaller at the thoracic spinal level and larger at the cervical and lumbar segments. At the lumbar region, one of the anterior radiculomedullary arteries is particularly developed; the artery of the lumbar enlargement (arteria radicula magna of Adamkiewicz). The origin of this artery varies greatly but it originates most often between levels T9 and T12, most frequently on the left.

Three principal longitudinal axes exist, represented in front by the anterior spinal axis and behind by a pial network across which derive two privileged circulation pathways: the posterior spinal axes. At cervical level, the anterior spinal axis is organized from the anterior spinal artery formed by the union of two branches that originate in the intracranial vertebral artery. Then at thoracic spinal level, the anterior spinal axis is often discontinuous. At lumbar level, the anterior arterial axis is larger and fed by the branches of the superior and inferior division of the anterior arteria radicula magna (of Adamkiewicz). The dorsal part of the spinal cord is covered by a large arterial plexus, supplied by the radiculopial arteries. At cervical spinal level, these predominant currents anastomose with the vertebral system. Indeed, two posterior spinal arteries originate bilaterally from each side of the vertebral artery or from the proximal part of the posterior inferior cerebellar artery to join the posterior axes. The posterior arterial axis ends caudally at the conus medullaris, where it anastomosis with the anterior arterial axis. Intrinsic blood supply to the spinal cord is organized around two large systems: the sulco-commissural arteries and the radial perforating arteries.

VENOUS CIRCULATION OF THE SPINAL CORD[17,18]

Venous circulation differs from the arterial circulation in two important ways: the veins are not satellites of the arteries inside the parenchyma and vein density is greater on the dorsal side than on the ventral side of the cord. The intraspinal veins drain the venous blood towards the extrinsic venous axes. They can be divided into three groups: 1) central or sulco-commissural veins, 2) posterior veins, and 3) peripheral veins. Contrary to the arterial system, venous drainage occurs predominantly towards the posterior part of the spinal cord. The extrinsic system comprises the 1) pial venous network and 2) longitudinal collectors. The anastomotic pial venous system is located on the surface of the cord, in the layers of the pia mater. As is the case for the arterial system, there are two types of principal longitudinal pathways: the ventral spinal venous axis and the dorsal spinal venous axis. The radicular veins drain the venous blood from the perimedullary system towards the vertebral plexus. They vary in number and are generally larger than the radicular arteries. There are an estimated 25 anterior radicular veins and 30 posterior radicular veins. It is exceptional to find a radicular artery and a radicular vein at the same level. The radicular ventral veins most notably drain the anterior median venous axis. The dorsal radicular veins anastomose between themselves and with the median dorsal axis, which principally drains the pial plexus.

CLASSIFICATIONS:

Most significant advance in the diagnosis of spinal cord vascular malformations was brought about by the advent of selective angiography in the 1960s. It laid the foundation for new scientific classification systems for spinal vascular malformations. Furthermore, it laid the groundwork for the modern surgical and endovascular approach to these lesions. Initial classifications based on microscopic appearance, structure, topographic relationship and pathological nature were confusing and complexity of the nomenclature made it difficult to understand and compare various lesions. Wyburn-Mason[12] recognized two main types of vascular malformations, the venous type and arteriovenous type. Venous type showed abnormal pial veins on dorsal aspect of the cord and were usually found in lower thoracic spine. Arteriovenous type located in cervical and lumbar enlargement, showed fistulous capillary bed. Arteriovenous type presented early in life while venous type was seen in old age patients.[12]

Modern classification of the malformations was first proposed by DiChiro, Doppman, and Ommaya in 1971.[19] They divided vascular malformations into single coiled vessel lesions (currently recognized as spinal dural AVFs or type I), the typical high-flow glomus type of intramedullary AVM (type 1I), and the more extensive AVM that tends to occur in younger people and frequently extends into the vertebral body and paraspinal tissue ("juvenile" or type III). The most common of pattern was extramedullary and dorsally located lesion described as being fed by a single feeding artery in the majority of cases, with very sluggish flow being characteristic. Many other specialists contributed to the further development and subclassification of different groups of spinal cord vascular abnormalities. In 1977, Djindjian et al[20] first described the intradural perimedullary AVF. Later, Merland et al[21, 22] reported their experience with such lesions and then proposed their sub classification into three different types on the basis of caliber, length, and number of feeding and draining vessels. In 1986, Heros et al[23] reported a patient with an intradural perimedullary AVF and proposed that this type of spinal vascular malformation be classified as a type IV spinal vascular malformation.

In 1976, Aminoff[24] stated that these lesions could not be divided satisfactorily in to venous or arterio-venous type based on either angiography or pathology. However he noted that lesions with significant shunting presented early and showed high incidence of hemorrhage. Similar Djindjian and Hurth et al[20] noted differences in the cervical lesions compared to those in the thoracic and lumbar regions. They observed that lesions having a direct supply from the artery of Adamkiewicz presented early compared to posteriorly located lesions.

In 1977 Kendall and Logue[25] published the first description of spinal AVFs. They reported on 9 cases of extradural spinal AVMs draining into the coronal spinal venous plexus The authors noted that the malformation derived its arterial supply from 1 or 2 segmental arteries and that all had 1 or 2 arterialized veins that penetrated the dura 2–3 mm from the nerve root sheath to drain into the coronal plexus of veins. Merland and associates[21], Oldfield et al[26], and Symon et al[27] suggested that lesions previously classified as extramedullary AVMs were in fact dural AVFs.

Gueguen et al[22], had divided ventral fistulas into three subtypes: 1, 2, and 3. Anson and Spetzler [28] reclassified the subtypes as IV-A, IV-B, and IV-C. From an anatomical point of view, it is important to distinguish paraspinal (including vertebrovertebral fistulas) and epidural locations from dural and intradural locations of the AVM or AVF, as the neurological symptoms will nearly always be related to the venous drainage pattern of the lesion no matter where the nidus is situated. Spetzler et al[29] again classified these lesions into three primary or broad categories: neoplasms, aneurysms, and arteriovenous lesions. The third category, spinal cord arteriovenous lesions, is divided into arteriovenous fistulas and arteriovenous malformations (AVMs). Arteriovenous fistulas are subdivided into those that are extradural and those that are intradural, with intradural lesions categorized as either dorsal or ventral. Arteriovenous malformations are subdivided into extradural–intradural and intradural malformations. Intradural lesions are further divided into intramedullary, intramedullary–extramedullary, and conus medullaris, a new category of AVM.

Spinal vascular malformations lesions are dynamic in nature and changes will occur in the malformation itself that will modify its clinical behavior, for example appearance of intranidal aneurysm. Rodesch et al[30] classified vascular malformations of spinal cord in three main groups. The first group includes genetic hereditary lesions that are caused by a genetic disorder affecting the vascular germinal cells. Spinal cord malformations associated with hereditary hemorrhagic telangiectasia fall into this category. The second group includes genetic nonhereditary lesions that share metameric links, such as Cobb syndrome (or spinal AV metameric syndrome), that affect the whole myelomere. Patients typically present with multiple shunts of the spinal cord, the nerve root, bone, paraspinal, subcutaneous, and skin tissues. Klippel-Trenaunay and Parkes- Weber syndromes also belong to this group. The third group includes single lesions that may reflect the incomplete expression of one of the previous mentioned situations and includes spinal cord, nerve root, and lesions of the filum terminale. Intradural direct AV shunts were classified in to micro AVF and macro AVF. Another classification scheme divides spinal vascular malformations, similar to vascular malformations of the brain, into true inborn lesions and the acquired lesions, the latter being dural AV fistulae, whereas AVMs and cavernomas constitute inborn lesions of the spine[31]. In conclusion there have been number classification schemes proposed for spinal vascular malformations and they reflect our growing knowledge in terms of their morphology, pathophysiology, embryology and genetics.

Comparison of various classification schemes of spinal vascular malformations:

DiChiro et al, 1971	**Spetzler et al, 2002**	**Rodesch at al, 2002**	**Krings et al, 2005**
Type I-dural AV fistula	dorsal intradural AVF		Dural AV fistula
Type II-AVM Glomus type	Intramedullary AVM	3. focal intradural SCAVS (nidus)	AVM-Glomerulus
Type III-AVM-Juvenile type	Extradural intradural AVM	2. genetic nonhereditary SCAVS-metameric	AVM-juvenile
Type IV-perimedullary fistula Type A, B &C	ventral intradural AVF	1. genetic hereditary lesions-macro AVF classified Type IVC PMF 3. focal intradural SCAVS (micro AVF)	AVM-perimedullary fistula I, II, III

EPIDEMIOLOGY:

Spinal dural arteriovenous fistulas: Among all the spinal cord malformations, spinal dural arteriovenous malformations (SDAVF) are the commonest one, with a proportion of 60-80% [21, 26, 27]. This disease entity is probably underdiagnosed[32,33]. In its review of 614 patients who suffered from a spinal cord lesion not due to trauma Jellima[32] found at least 3 patients in whom either a SDAVF or a cerebral fistula with spinal drainage was present. Out of these three patients one was diagnosed only on autopsy.

Patients affected by SDAVF are mostly middle-aged men. In a review of all reported series with more than 5 patients; there were 968 men against 210 women (ratio almost 5: 1). The mean age at the time of diagnosis was 55–60 years; patients under 30 years of age are rarely reported and they constitute approximately 1% of all the cases[34]. The youngest patients reported were 22 years at the time of diagnosis[35, 36].

Most SDAVFs are located in the thoracolumbar region. Cervical and sacral are uncommon locations and constitute approximately 6% of all the lesions. Multiple SDAVFs in a single patient are uncommon and are seen in less than 1% patients[34]. Two other series reported one patient (2%) and two patients (4%) with a double SDAVF among their patients[37,38].

Spinal cord arteriovenous malformations: In contrast to SDAVF spinal cord AVMs usually present in the 3rd decade of life, but they can be diagnosed in children < 16 years old in 20% of the cases[35,39]. When the pediatric population is included, a male predominance is shown, but no sex predominance is found in the adult population. Rosenblum et al[35] studied 81 patients of spinal vascular malformations comprising 27 (33%) had dural AV fistulas and 54 (67%) had intradural AVM's. The 54 patients with intradural AVM's were subclassified as having intramedullary AVM's (43 patients) or direct AV fistulas (11 patients). There was male predominance associated with intradural lesions comprising of 70% patients. Intradural AVM's occurred more diffusely along the longitudinal spinal axis than dural AV fistulas and occurred more commonly in the cervical or thoracolumbar region. Patients with symptoms from AVM's of the spinal cord were younger, aged 4 to 58 years (27 ± 12.1 years). Ten (19%) patients of intradural AVM were associated with other vascular abnormalities. In a clinical study comprising 155 patients of spinal cord AV shunts by Rodesch et al[30], there were 81% single lesions and 19% multiple; among these, 59% were true intradural shunts with metameric features. Ten cases of Cobb syndrome, three cases of Klippel-Trenaunay syndrome, and two cases of Parkes-Weber syndrome, all with associated cord lesions, were observed. Spinal dural AVF are not associated with other vascular abnormalities. Similar associations of spinal AVM with genetic vascular diseases was noted in the study of Rosenblum et al[35].

Perimedullary fistulas: Type IV spinal AVMs or intradural AVFs arise most frequently in the thoraco-lumbar spine[44,45,46]. Type IV vascular lesions are diagnosed mainly in the third or fourth decade of life[46]. Patients suffering from such perimedullary fistulas, however, might already become symptomatic during childhood[46,47]. Perimedullary fistulas show male predominance[48,49]. In the series of Cho et al[45] mean age was 28 yrs and 17 of 19 lesions were located in conus medullaris region. Nine lesions were type IVa, 6 IVb and remaining were type IVc. Three of four patients of Type IVc were children. In their series Rodesch et al[30] reported nineteen percent of SCAVSs were fistulae; 23% of those were macrofistulae, of which 83% were related to Rendu-Osler-Weber disease.

Epidural (paraspinal and vertebrovertebral fistulas): Paraspinal shunts are rare lesions and show a female predominance[50]. Although vertebrovertebral arteriovenous fistula are a type of epidural lesion, however because of there distinct clinical presentation and treatment they are considered a separate entity by many authors. In the series of Goyal et al there were 10 patients (7 male and 3 females) of paravertebral AVM with a mean age of 33 yrs (1-73 yrs)[51]. In another study comprising of 45 patients of vertebral arteriovenous fistulas there were 21 males and 24 female patients (Mean age 44 yrs)[52]. These lesions may post traumatic or spontaneous and can be associated with fibromuscular dysplasia or neurofibromatosis[51,52].

Overall spinal vascular malformations are rare lesions and constitute 3-4 % of all the spinal mass lesions[50]. They constitute 3.3% to 11% of all the spinal masses[53,54].

PATHOLOGY:

There are two principle categories of the spinal cord vascular malformations are seen. In the first variety there are aggregates of thin walled vessels which may be compacted, like a honeycomb, or dispersed throughout the parenchyma. The vascular caliber ranges from capillary to cavernous. They produce symptoms by either hemorrhage or by compression of the cord[55,56]. Second type malformations consists of tangled large serpentine channels that throw vascular loops about the cord and its nerve roots. Most vessels are situated on the dorsal surface of the cord and are accompanied by smaller hyalinized vessels within the underlying parenchyma. They contain thickened draining veins and feeding arteries, resembling cerebral arteriovenous malformations[57,58]. In the cases of venous congestion secondary to vascular malformations there may be necrotic and myelomalacic changes within the cord with evidence of concentric hyalinization of the intramedullary vessels[34, 57,58].

PATHOPHYSIOLOGY OF SYMPTOMS OF SPINAL AVS. NATURAL HISTORY. RELATIONSHIP WITH ANGIOARCHITECTURE

Spinal vascular malformations are a heterogeneous group of vascular lesions sharing the characteristics of abnormal blood vessel morphology. Clinical manifestations may be acute, subacute or chronic. The variability in clinical presentation reflects differences in pathophysiology of spinal cord vascular malformations requiring specific treatment options. Veins represent the key factor in the onset of neurological symptoms in any type of SCAVS. Arterial steal and cord ischemia of arterial origin has also been advocated to explain progressive impairment occurring at long term follow up in patients with SCAVSs but is considered unlikely mechanism by most of the studies[46,47,48].

Epidural, paraspinal and vertebrovertebral fistulas: They are frequently of a fistulous type and drain in ectatic veins located outside the spine. They present usually with progressive neurological symptoms. Venous ectasia in these condition behave like mass, invaginating in to the spinal canal and causing slow compression of the spinal cord. As the paraspinal veins communicate with the radicular and perimedullary venous drainage they may lead to the venous congestion in the cord[3,49,50]. Paraspinal AVSs can also lead to hemodynamic disorders like cardiac overload and congestive heart failure[59].

Dural arteriovenous shunts: The onset in middle age suggests that SDAVF is an acquired condition, in contrast to intradural ventral fistulas or AVMs, which are assumed to be congenital abnormalities[35]. They always reveal by progressive myelopathy, which tends to be more severe if both anterior and posterior drainage is present, The clinical history usually begins with paraparesis. Symptoms are progressive in 80%, acute onset paraplegia or paraparesis in 10% and progressive with remissions in the remaining[3]. Patients with SDAVF very rarely present with spinal haemorrhage in contrast to patients with AVMs[34,35]. Aminoff and others proposed in 1974 that venous hypertension and not vascular steal, cord compression or haemorrhage was the main pathophysiological factor[24]. The shunt is most often formed within the dorsal surface of the dural root sleeve in the intervertebral foramen, where the radicular vein pierces the dura, together with one or more dural branches of the radicular artery. However, the shunt is sometimes situated along the dura between two adjacent nerve roots[3,4,25,46]. The shunt results in venous hypertension in the spinal cord, because the intramedullary veins and the radicular vein share a common venous outflow. The reduced arteriovenous pressure gradient results in a decrease in tissue perfusion and venous infarction[60]. This possibly explains the worsening of symptoms with exercise, which leads to increase in the arterial pressure. Scarcity of the venous outflow channels in the lower thoracic cord leads to transmission of venous congestion in caudo-cranial direction throughout the spinal cord and that the first symptoms of myelopathy tend to reflect dysfunction of the the conus medullaris[61].

Intradural spinal cord arteriovenous shunts (SCAVS): Bleeding is one of the distinctive features of the intradural AV shunts. Other manifestations include acute non hemorrhagic neurological deficits and chronic progressive neurological impairment. Berenstein and Lasjaunias[46] in 1992 were among the first to analyse symptoms, evolution and architecture of intradural SCAVSs. They confirmed the spontaneous progressive impairment of spinal cord SCAVSs after diagnosis, emphasised the role of the veins in the onset of the symptoms and suggested the relationship between associated aneurysms and haemorrhage. In their analysis of 155 cases, Rodesch et al[62] concluded that venous congestion and presence of false aneurysms are the only architectural features correlating with the symptomology. They found no significant differences in the angio-architecture between hemorrhagic and non hemorrhagic SCAVS, except for that occurrence of pseudo-aneurysms. Venous congestion was responsible for progressive symptoms. Acute deficits unrelated to bleeds were due to intralesional thrombosis or hemodynamic changes. There was difference in the natural history of the lesions in childrens and adults as bleeds and hematomyelia were more commonly seen in the children. They postulated the hypothesis that this is possibly due to the fragile and unstable equilibrium between the lesional and normal vasculature and latter fails to compensate for the shunts. In 72% patients with bleed, there was spontaneous recovery without early aggressive treatment. Rebleed rates were low. Presence of associated genetic diseases with these conditions could be another factor responsible for underlying biological weakness of the vasculature resulting in hemorrhage. Acute non-hemorrhagic deficits could be seen in: 7.5% of adults and 22% of children. These abrupt deficits represent thrombosis of parts of the malformation or acute hemodynamic changes occurring within the lesion, disrupting the equilibrium between the SCAVS and the cord. Neurological symptoms in SCAVSs were most often progressive.

About 69% of patients in the cohort of Hurth et al[63] presented with a stepwise course, ending in significant impairment of spinal function. In his study patients with thoracic and lumbar AVMs were more rapidly disabled compared to the cervical lesions which were well tolerated. Reexamination at 5, 10, and 20 years revealed severe neurologic deterioration in 13%, 20%, and 57% of the patients, respectively. He found that two factors undoubtedly affected the progression of the disease: the position (extramedullary or intramedullary) of the shunt and the localization of the lesion. Major neurological impairment occurred within 4-6 years in cases with extramedullary malformations. Lesions in the midthoracic location had the worst prognosis, with 66% of these patients having major disabilities at 20 years. This is may be due to the poor vascular supply and collateral circulation in this region of the cord.

Aminoff et al[64] reported 8-year follow-up on 60 patients with untreated AVMs of the spinal cord. Within 3 years from onset of symptoms, 36% of patients less than 41 years and 48% of patients 41 to 60 years were confined to bed or wheelchair. Nine patients in this study died due to complications of chronic paraplegia.

The progressive symptomatology in cases of intradural vascular malformations is also due to the venous congestion producing progressive myelopathy, with gradual congestion of the intrinsic and extrinsic draining channels[65]. Because of transmedullary anastomoses and the richness of these venous networks, 'referred symptoms' remote from the shunt and different from the territory of the arterial feeders can be explained.

CLINICAL FEATURES:

Spinal Dural arteriovenous fistula: The clinical features of SDAVF can be distinguished in those at the onset of the disease, in retrospect, and those present at the time of diagnosis. Initial symptoms are often nonspecific. They include gait difficulties, symmetrical or asymmetrical sensory symptoms such as paraesthesias, diffuse or patchy sensory loss, and even radicular pain. Disturbances of micturition and defaecation may occur at the start but most often they develop in later phases of the disease[35,37,66,67,68]. Course of the disease is usually progressive, however acute onset symptoms are recorded in 5-18 % patients[67]. Acute onset of symptoms or clinical worsening may occur following exercise, prolonged standing, bending over or eating[24,27,35]. Intraspinal haemorrhage is distinctly rare, since it occurred in none of the 1178 patients described in the cumulative series with more than 5 patients[34]. Patients may present with acute headache as a result of intracranial subarachnoid headache (SAH), mostly in patients with a dural fistula at the craniocervical junction, but may rarely be observed in lower thoracic or thoracolumbar lesions[34]. At the time of diagnosis two-third of patients show a combination of gait difficulties, sensory disturbances and involvement of sacral segments (micturition, defaecation or sexual dysfunction). Erectile dysfunction exists in 11–80% of men patients[34,67].

Spinal cord AVM: Hemorrhage is the most common presentation, occurring in about 50% of all patients with spinal cord AVMs, and it is often associated with sudden onset of new neurological deficits or with worsening of preexisting deficits[3,4,35]. The typical syndrome of spinal hemorrhage is characterized by acute severe back pain spreading along the spinal axis and legs. Motor and sensory symptoms and bladder and bowel dysfunction can occur. In a small percentage of patients (25%), preexisting spinal cord or nerve root symptoms are present. Intracranial SAH can occur with severe headaches and di sturbance of consciousness and an intracranial origin can be wrongly suspected. The hemorrhage rate seems to be different in the pediatric population in which hemorrhage and hematomyelia are more frequent[62]. Non-hemorrhagic symptoms include root or back pain, weakness, sensory changes, sexual, bowel, and bladder dysfunction, and rarely a bruit[24,35,69].

Perimedullary fistula: In their study of 32 cases of intradural arteriovenous fistulas, Rodesch et al [70] found 10 cases in pediatric population and remaining in adults. Seven of the pediatric lesions presented with hemorrhage which included 3cases of micro-AVF and 4 of macro-AVF. All patients with macro AVF were associated with HHT. In adult group there were 21 cases of micro-AVF and only one case of macro-AVF. Hemorrhage was present in 6 of the 22 cases in adults. Overall 9 of the 25 cases of micro AVF presented with hemorrhage, 3 with non hemorrhagic acute symptoms and remaining had features of chronic myelopathy, while in cases of macro AVF 4 patients presented with acute hemorrhage while in the remaining chronic progressive myelopathy was noted. In the study of Cho et al[45], most common presentation was progressive weakness followed by backache. Acute presentation was noted only in the 3 patients. In another study, majority of patients presented with progressive illness (32 of 35); hemorrhage was noted in 7 cases (20%)[42].

DIAGNOSTIC IMAGING OF THE SPINAL VASCULAR MALFORMATIONS:

Imaging of SVMs has always presented a formidable challenge because their clinical and imaging presentations are similar to neoplasms, demyelination diseases, and infection[4,71,72]. The gold standard of imaging of SVMs has traditionally been catheter-based 2D angiography because of its ability to produce images with superior temporal and spatial resolution[4,46,69]. However, spinal 2D DS angiography is not without risks. Selective catheterization of arteries supplying an SVM can often be time consuming, require multiple catheterizations, involve long radiation exposure times, and use large volumes of potentially nephrotoxic contrast agents. Furthermore, selective catheterization of segmental arteries can lead to spinal cord infarction due to their embolization or occlusion[73]. Therefore, advancements in noninvasive imaging modalities (MR and CT angiography) have improved the ability to diagnose these conditions non invasively[1,4,73,74,75,76,77].

Although the imaging hallmarks of SVMs are typically less apparent when CT is used, they are similar to those seen on MR imaging, namely prominent or enlarged subarachnoid vessels. Magnetic resonance imaging, however, has the distinct advantage of examining the spinal cord tissue and supporting structures in detail which, on T2-weighted images, often demonstrates increased signal intensity, representing spinal cord edema and/or hematoma. They do not provide any hemodynamic information regarding the vasculature of the spinal column[1,3,73]. Noninvasive angiography techniques (CT and MR angiography) have recently allowed the acquisition of dynamic information, which has greatly assisted in not only the diagnoses of SVMs, but also the localization of the fistulous connection(s)[1,74,75,76,77]. Lai et al. reported 8 cases of spinal DAVFs in which the fistulous connection, feeding arteries, and draining veins were localized with great consistency when compared with spinal DS angiography[75]. However, the spatial and temporal resolution was still inferior to spinal digital subtraction angiography and made the angioarchitecture of complicated SVMs very difficult to characterize. Computed tomography angiography also has the added disadvantage of exposing patients to radiation and potentially nephrotoxic contrast agents[73].

To overcome the limitation of temporal resolution of noninvasive imaging, rapid multiphase dynamic MR angiography with parallel imaging has been developed with a temporal resolution of 3–6 seconds and a spatial resolution of ~ 1 mm The advantages of rapid time-resolved MR angiography of the spine are the improvement of temporal resolution through parallel imaging while maintaining a high spatial resolution, allowing hemodynamic visualization of the SVM, acquiring a large FOV, allowing a wide survey of possible fistulous connections, and eliminating patient exposure to radiation and iodinated-contrast agents. Over all MRI and MRA help in the diagnosis of SVMs and can be used to complement, not supplant, spinal DS angiography[73,78].

In paraspinal arteriovenous shunts, T1 or T2 signal void serpiginous or tubular structures, corresponding to large draining veins, are located outside the spinal canal. The cord may be affected because of mechanical compression or congested due to venous hypertension. Differential diagnosis is difficult and presence of epidural, vertebral and paraspinal flow voids helps in localization and characterization of these lesions[51].

Intramedullary high signal intensity is present due to the venous congestion. Intravenous injection of gadolinium-DTPA enhances the dilated veins. It is the combination of these dilated veins with the high signal intensity of the cord that leads to the diagnosis of dural shunt. An isolated high signal intensity without perimedullary venous structures is either related to a non-vascular cause or may suggest the diagnosis of a micro shunt draining exclusively in the subpial anterior spinal vein and not in posterior veins that are subarachnoid[3,4,34,79]. Cord enhancement is moderate in the early post injection period, and more significant in delayed phase after 45 min; it is believed to be due to the venous stasis resulting in oedema, ischemia or venous infarction of the cord[3]. Cord hyperintensity is reversible if underlying condition is treated at early, however later it may be permanent and cord atrophy takes place. Patients with cord enhancement carries poorer prognosis when compared with the patients with no enhancement[65].

MR enables the making of the diagnosis of an intradural SCAVS by showing abnormal cluster of vessels and the secondary changes in the cord, however it often fails to reveal the the precise nature of the shunt and its exact location and their association with myelomeric lesions. Large lesions are easier to depict. Associated lesions or evolutive complications, such as hematomyelias, oedema, thrombosis, or cord atrophy are well depicted on multi-planar sections[3].

The role of MR angiography in intradural spinal cord arteriovenous shunts remains debatable. The level of origin of the arterial feeders and the distribution of the draining veins can be determined but no detailed information regarding regional or 'lesional' angio-architecture can be obtained by this promising technique[3,4].

THERAPEUTIC MANAGEMENT OF SPINAL ARTERIOVENOUS SHUNTS

Introduction and subsequent refinement of spinal arteriography by Djindjian, Doppman and DiChiro, and others permitted progression of knowledge concerning the pathophysiology and subsequent treatment of the spinal vascular malformations[15,16,19,80,81]. Doppman and colleagues generated successive advancements through the addition of percutaneous embolization to the therapeutic armamentarium, treating a 16-year-old boy by embolizing an arterial feeder with steel pellets[82]. Subsequently, angiography, in addition to its position as the gold standard of diagnosis, has experienced a continued technical and technological evolution principal components of therapeutic enhancement have been the advent of microcatheters and the continued introduction of novel substances for embolization The newer microcatheters allow the neurointerventionalist to navigate small and tortuous vasculature with increased safety and to promote canalization of previously inaccessible vascular territories[2].

In field of embolic agents also we have seen significant advancement with the use of liquid adhesives such as IBCA and NBCA resulting in substantial improvement in recanalization formerly demonstrated with particles such as organic fragments, silk, and PVA[2,83]. Additional benefits include adjustable polymerization time and superior vessel permeation; however, despite these advantages, the liquid embolic agents are not without problem, including the risk of catheter adhesion, variability in physical behavior, and most significantly an increased risk of spinal cord infarction[2].

Newer liquid embolic agent Onyx has additional advantages over NBCA as an embolic agent; it can be delivered slowly in a more controlled fashion over several minutes, carries a lower risk of premature venous occlusion or occlusion of a large intradural portion of the draining vein, and carries a low risk of a retained catheter. However because of angiotoxic and neurotoxic effect of the solvent used, safety of this new agent remains to be clearly established in spinal cord[84,85,86].

TREATMENT OF SPINAL DURAL ARTERIOVENOUS FISTULAS:

The choice of treatment is between endovascular embolization and surgical ligation of the fistula. Initially spinal dural arteriovenous fistulas were treated using particle embolization, which was found to be safe and gave good immediate results. However soon it was realized that occlusion following particle embolization are not permanent. In the series of Morgan and Marsh[87], 14 patients underwent PVA particles embolization. Despite safe and complete angiographic obliteration, in over 12 of 14 patients 13 recurrences were noted in the the follow-up. Results of embolization were more durable with the use of liquid embolic agents. In the study of Niimi et al[66], adequate angiographic result was achieved in 87% patients where penetration of the liquid embolic agent (both IBCA and NBCA) into the fistula or draining vein was seen. Authors concluded that because this procedure could be safely performed, embolization with liquid cyanoacrylate should be the first-line treatment provided that angiography does not demonstrate a common pedicle for both the feeder and spinal artery.

Song et al [88] retrospectively reviewed twenty seven patients who underwent treatment for DAVF. Seventy-four percent (20 patients) had lesions amenable to NBCA-based endovascular intervention, and 90% (18 patients) had complete angiographic occlusion. Over an average follow-up period of 3.1 years (range 1 month–8.9 years) 15% of the patients suffered recurrence of the fistula. van Dijk et al[38] reported angiographic cure in only 25% of their patients of spinal DAVF. In meta-analysis of Steinmetz et al[89], the published literature from 1966 to 2003 was reviewed and in 10 studies included, definitive treatment was possible in only 46% patients. However the use of NBCA was not an inclusion criterion and particles were used in some of the included studies which we already know, demonstrate recanalization on follow up.

There is very limited experience with newer liquid embolic agent Onyx. Nogueira et al[90] recently reported their experience with Onyx in three patients of spinal dural AVF and they concluded that endovascular treatment of SDAVFs with the Onyx is feasible, safe, and highly effective, as it allows for a controlled penetration of the embolic agent into the draining vein. However there are concerns about its long term results which need to be addressed with follow up studies.

In patients without a common origin for both the feeding artery and spinal artery, embolization with liquid acrylic agents represents a safe initial treatment option, providing durable cure in up to 70–85% of patients when the agent penetrates the proximal draining vein[3,4,88,91].

Embolization of SDAVF is not possible in every patient. First, if the arterial feeder of the fistula is a segmental medullary artery, embolization entails a high risk of spinal cord ischaemia. Secondly, technical difficulties such as arterial wall dissection of the feeding vessels during the embolization procedure may prohibit introduction of the microcatheter close enough to the fistula. If embolization is possible, then the success of treatment depends on endovascular occlusion of the draining vein, which means that recanalization will rarely occur when the draining vein is filled with glue[92]. The reason is probably that the fistula is often made up of several small feeding arteries and a single draining vein, so that occlusion of only (one of) the arterial feeder(s) to the fistula will generally lead to development of new arterial feeders[92,93]. In cases of failure of embolization or cases where it is contraindicated, surgery plays important role. Surgical occlusion of the intradural vein that receives the blood from the shunt zone is a relatively simple and safe intervention with the exception of sacral fistulae[94,95]. In the meta-analysis of Steinmetz et al success of surgery was 98% compared to 48% for endovascular procedure[89].

TREATMENT OF SPINAL AVM:

The therapeutic approach of asymptomatic AVM is difficult since data concerning the spontaneous prognosis are not available; however, in symptomatic AVMs therapy ameliorates the prognosis of the patient. Therapeutic decision making requires consideration of the patient's medical comorbidities, lesion location, and angioarchitecture, as both microsurgical excision and endovascular embolization carry significant risk of neurological injury[96].

Treatment options for spinal cord AVMs include excision, endovascular obliteration, partial targeted therapy, or conservative management. The primary goal in the management of spinal cord AVMs is to preserve neurological function. Cure of these lesions is seldom obtained without morbidity. Partial targeted treatment to obliterate weak portions like arterial or nidal aneurysms, size reduction, decreased flow, and decongestion of the venous drainage may modify the natural history and improve clinical outcome. Ideally all symptomatic lesions should be completely obliterated. However, it is of utmost importance to identify the cases in which total anatomical cure is not possible without worsening the neurological status of the patient. Under such circumstances, an anatomical goal or objective should be defined before starting the treatment. Obliteration of a high-risk portion of the lesion, such as an associated aneurysm, the occlusion of a direct AVF within the nidus, or of a portion draining into a territory with outflow restriction, can result in a favorable long-term outcome[4,69].

Early aggressive treatment is rarely warranted in cases of spinal AVM. Exception being presence of high risk feature like presence of pseudoaneurysm in which case early intervention is indicated because of high risk of rebleed. Most patients show gradual improvement over a period of time[62,69].

Embolization can be done with particles or liquid embolic agents. Role of particles lies in lesions that are inoperable due to location (that is, ventral thoracolumbar lesions) or unsuitable angioarchitecture, and/or when the nidus cannot be reached with current microcatheter technology. Particulate embolization, in addition to facilitating surgical intervention, reduces the risk of hemorrhage, provides symptomatic improvement, and can delay symptomatic progression[97]. Clinical benefits of the particulate embolization were shown in the study of Biondi et al[97], where 35 patients with thoracic intramedullary AVMs, both glomus and juvenile type were followed-up up to 5 years, 158 procedures were performed to treat recanalization; however, 63% of the patients exhibited clinical improvement compared with baseline findings. There were fewer complications and these procedures were performed with a low incidence of adverse effects. They observed 23 complications (15%), of which 12 (8%) resulted in clinical worsening, Use of liquid embolic agents give permanent and durable exclusion of the abnormal vasculature. In the series of Rodesch et al[98] comprising of 69 patients, 99% embolization were performed using acrylic glue. Anatomical obliteration of the shunt was obtained in 16% patients while in the remaining patients >50% reduction was achieved. No recanalization was noted on follow-up angiograms. Good clinical outcomes were obtained in 83% of the patients: 15% of them were asymptomatic, 43% were improved, and 25% were stable. In 4% of patients, embolization failed to stabilize the disease. Transient deficits were seen after embolization in 14% of the patients, and permanent severe complications occurred in 4% of the patients. Mild clinical worsening was seen in 9% of the patients. In another series outcome was excellent in 49% of patients and good in 28%; 77% of the cases had a favorable result. None of the patients with a previous fixed deficit returned to their normal condition. Permanent worsening directly related to the procedure occurred in 11% of the cases, and transient complications occurred in another 11%. Immediate complete obliteration of the malformation was achieved in almost half of the patients by embolization alone. In the remaining patients, partial targeted embolization or combined approaches associating embolization and surgery were used[69].

There is limited data on the efficacy of Onyx in the treatment of intramedullary AVM patients. In one study consisting of 17 patients, good angiographic outcomes included total AVM obliteration in six patients (37.5%), subtotal obliteration in five patients (31.25%), and partial obliteration in five patients (31.25%). Improvement in neurological and/or functional status was noted in 14 patients, resulting in an 82% rate of overall good clinical outcome[99].

In the surgical series, Yaşargil and colleagues[39] reported good results in 41 patients with spinal cord AVMs who underwent surgical treatment. Clinical improvement occurred in 48% of the cases and clinical worsening in 19.5%; there was 1 death directly related to surgery. Spetzler et al[29] reported on their surgical experience that 68% of the patients (27 spinal cord AVMs) improved and 29% remained the same, with 92% of complete resection. In 8 patients clinical worsening was noted.

PERIMEDULLARY AV FISTULA:

Perimedullary fistula may lead to hemorrhage or slowly progressive myelopathy due to spinal cord congestion. Rarely enlarged venous sac as seen in cases of type 3 PMF or macro AVF can cause mechanical compression of the spinal cord.

The location and angioarchitecture of perimedullary fistulas are critical in management strategies for these lesions. For Type I fistulas surgery is considered treatment of choice because embolization of type I perimedullary spinal arteriovenous fistulas (AVFs) can be difficult, because of tortuosity and the small diameter of the feeder and distal location of the fistula site. Higher grade complex fistulas and/or those in difficult regions for surgical exposure are probably best approached by endovascular methods[100]. Surgery is considered the treatment of choice only when embolization seems to be too hazardous or impossible because of small and tortuous feeders or remote location of the shunt as in case of filum terminale lesions[101].

Aim of treatment in this condition is should always be complete obliteration of the fistula, however even partial treatment of the lesions results in clinical stabilization and good clinical outcome[101]. Therapeutic decisions will be based on angiography, which diagnoses the fistula, and treatment must be focused on the shunt itself and not on its hemodynamic or biological consequences. Some lesions considered at first sight to be intramedullary-type arteriovenous malformations are in fact simple mAVFs with reflux and congestion of the intrinsic venous network, giving a "pseudonidus" appearance. Surgical treatment involves interruption of the arterial supply by applying a clip followed by disconnection of the draining vein as near to the fistula as possible. In their experience of treatment of 32 cases of perimedullary fistulas Rodesch at al[101] reported successful embolization of 86% macro AVF and 48% of micro AVF. 67% were completely cured in both the categories and remaining were partially embolized. However all patients improved clinically despite partial treatment in few of them. Recently, Oran et al[102] reported successful embolization of 4 of 5 patients with type I perimedullary AVFs.

In the study of Cho et al[45] patients underwent endovascular treatment, complete angiographic obliteration of fistula was performed in 5 and partial obliteration in 4. Symptomatic improvement or arrest of progression was achieved in 5 of 9 patients with complete or partial occlusion. Embolization failed in two patients. With surgery most patients (9 of 10) were improved or stable. In the series by Barrow et al[40] patients underwent surgery and one was treated by embolization. Following surgery one patient became normal, 5 improved while in one case neurological status remained unchanged. One case treated by endovascular means regained normal clinical status.

TREATMENT OF PARASPINAL AVM AND VERTEBRAL ARTERIOVENOUS FISTULA:

In the case series reported by Goyal et al[51] combination of methods were used to treat paraspinal vascular lesions. Patients with medullary reflux underwent surgery while in other cases combination of trans arterial and transvenous embolization were done using both liquid embolic agents as well as coils. Authors concluded that in this condition treatment needs to be individualized and decided according to the pathophysiology in each case.

Vertebral arteriovenous fistula is a rare entity and only few large series are available. Beaujeux et al[52] reported 45 cases of vertebral arteriovenous fistula. Thirty four were treated by endovascular means. Using detachable balloons authors reported cure rate of 91% and they sacrificed the vertebral artery in 3 cases. All the patients were symptomatically better following the procedure.

Aims and Objectives

AIMS AND OBJECTIVES

1. To review all types of vascular malformations of spine that were investigated and treated in our hospital over a period of ~25 years.

2. To analyze different clinical presentations of spinal vascular malformations in relation to their types.

3. To study the angioarchitecture of various types of spinal vascular malformations.

4. To study the details of the procedure and management protocol for different types of spinal vascular malformations.

5. To analyze different approaches for endovascular management of various types of spinal vascular malformations

6. To analyze the outcome of the endovascular management of spinal vascular malformations.

7. To analyze the outcome of the endovascular management of spinal vascular malformations, in relation to different types of spinal vascular malformations treated.

8. To analyze the complication rates in relation to various types of spinal vascular malformations treated.

Material and Methods

MATERIAL AND METHODS

This was a retrospective & prospective study carried out at the department of IS & IR, SCTIMST, Trivandrum. For the purpose of this study, patients who had undergone investigation and treatment for spinal vascular malformations at the department of IS & IR, SCTIMST, from 1985 to 30th September 2008 were reviewed. There were 74 patients in total including all types of malformations (Spinal cavernomas were excluded). The clinical data of the patients was obtained by reviewing their case sheets obtained from the Medical Records Department (MRD) & the imaging data was obtained from the DSA lab records. From these, the data regarding clinical presentation, angiographic characteristics, and details of the embolisation procedure, post procedural details and also the follow up data was collected. The patients were advised follow up in the interventional radiology outpatient department by appointment.

A total of 74 cases of spinal vascular malformation were investigated and managed at our institute since 1985. All patients underwent a complete neurological evaluation prior to the procedure. The patients had a baseline imaging done – either a CT or MRI of the spine. From 1996 till date we have been using Advantx digital substraction angiography unit by GE (GE Milwaukee, USA). A wide variety of catheters and embolic material was used for the procedures as was best suited for the individual patient depending on the angioarchitecture.

Initially 1985-93 the diagnostic angiography was done under local anaesthesia after premedication (Inj. Pethidine 50mg i.m & Inj. Phenergan 25mg i.m for adults, and as per body weight for children). Since 1994 all diagnostic procedures are done under general anaesthesia. For procedures carried out under general anesthesia, the patient was monitored in the neurological intensive care unit. All therapeutic procedures were carried out under general anaesthesia only.

PATIENT EVALUATION

Complete records of the patient were reviewed and proforma was completed. Format of the proforma is attached. Clinical symptoms were assessed before and after treatment using the modified Aminoff– Logue grading scale (ALS) for myelopathy. The patients were followed up with clinical evaluation for improvement in their symptoms. Follow up angiograms and MRI was performed where possible.

CT: CT does not form part of our routine protocol for evaluating cases of the spinal vascular malformation.

MRI: MRI is the first line investigation in our hospital for evaluating cases of spinal vascular lesions. Our protocol includes T1W tra/sag [TR/TE/NEX

(495/12/2), matrix 256x512, slice thickness 3mm], T2W tra/sag [TR/TE/NEX (5360/125/3) matrix 173x256, slice thickness 3mm], 3D TSE sag [TR/TE/NEX (1500/142/2), matrix 317x320, slice thickness 0.9mm], post contrast T1W images following injection of Gadobenate dimeglumine I.V 0.1mmol/Kg body weight and GRE T2W [TR/TE/NEX (1090/22/2), matrix 173x256, slice thickness 4mm]. Gradient images are done to detect presence of blood.

Myelography: It was routine investigation prior to 1992, however we no longer use it for evaluating these case.

Angiography procedure: All the spinal angiography are done via right common femoral artery route. For diagnostic procedures, 6F sheath is used. Following introduction of sheath, 2500 units of heparin is given (in children 50 units/Kg body weight is used). Dose is repeated 100 units every hour. Initially aortogram is done at arch and descending aortic level using 4-5F pigtail catheter (Boston scientific, Tijuana, Mexico). Selective catheterization of the intercostal and lumbar arteries is done using 4F/5F left coronary (Cordis, Miami, USA), right coronary (Cordis, Miami, USA), SHK (Cordis, Miami, USA), Sims catheter (Cordis, Miami, USA) and Cobra (Terumo, Tokyo, Japan). Vertebral artery, external carotid artery angiography are done using 4F/5F vertebral Glidecath (Terumo, Tokyo, Japan).

The angiographic protocol included catheterization of the vertebral, costocervical, thyrocervical, external carotid arteries and segmental arteries, including the lower lumbar arteries, median and lateral sacral arteries, and internal iliac arteries for lower lumbar or sacral lesions. Selective angiography of the suspected feeding arteries was then performed. Following angiography patients sheath is removed in lab and hemostasis achieved using compression technique. Antibiotics are not given following diagnostic spinal angiogram.

Embolization procedure:

- All embolization procedures were performed with the patient under general anesthesia.
- The procedure was performed in Advantx digital substraction angiography unit by GE (GE Milwaukee, USA). Systolic blood pressure during the procedure was controlled at less than 100 ± 10 mmHg.
- Catheterization was performed by a transfemoral approach using standard coaxial techniques, however on one case direct puncture technique was used for embolization.
- An intravenous bolus of 5000IU of heparin was given to each patient and maintainence dose of 1000IU hourly.

- The angiographic protocol included catheterization of the vertebral and segmental arteries, including the lower lumbar arteries, median and lateral sacral arteries, and internal iliac arteries for lower lumbar or sacral lesions. Selective angiography of the suspected feeding arteries was then performed.

- All microcatheterization was done under roadmap guidance. Once microcatheterization as the close as possible to the nidus/fistula was achieved, a nonsubstracted single-shot image of the microcatheter with the guidewire was obtained when catheterizing the feeding pedicle to have a trace of the microcatheter trajectory and to be able to control precisely the reflux within the feeder. The guidewire was then removed, and a superselective arteriogram was performed via the microcatheter.

NBCA (glue) injection technique:

- When NBCA comes in contact with an ionic solution of alkali pH, it solidifies quickly into a hard paste by an exothermic reaction.

- To prevent this, bolus of glue was preceded by a low pH, non ionic flush solution with 5% dextrose.

- In case of wedge position within the nidus without very early venous drainage, a dilution of 14% to 80% was used. (NBCA diluted with lipiodol) depending on the rapidity of shunt.

- Highly concentrated glues (50% to 80%) was used in large caliber direct arteriovenous fistula without interposed arterioles or when the origin of vein which should not be occluded is very close to the catheter tip or when distance from the tip of catheter to normal arteries is short and reflux along the tip of the catheter must be perfectly controlled.

- Injection was done very slowly till the glue comes out of the catheter to avoid the formation of multiple little drops of glue exiting too rapidly and spreading quickly in the veins.

- First drop of glue was pushed very gently from the tip of the catheter to the distal artery and nidus.

- Injection was stopped for a few seconds and then resumed when the glue was seen penetrating into the origin of draining vein.

- At the end of injection the glue was aspirated back within the syringe, guiding catheter pulled back and the microcatheter was withdrawn rapidly into the guiding catheter to avoid migration of some drops of glue in the normal circulation.

For embolization, a mixture of n-butylcyanoacrylate (NBCA) and iodized oil, hydrogel, polyvinyl alcohol particles and Gelfoam are used. Glue was injected via a microcatheter coaxially inserted through the guiding catheter. For particulate embolization selective catheterization of the feeding arteries is done and embolization done till complete devascularization is achieved. When selective catheterization was difficult distal occlusion of the intercostal artery was carried out using fibre coil thus facilitating superselective microcatheterization and embolization. In case where selective catheterization cannot be done, distal intercostal artery is occluded with fibre coils and particles injected using 4F catheter.

A decision to treat was made in all patients with angiographically confirmed diagnosis of vascular lesion. In a small number of selected cases, endovascular treatment was repeated due to partial occlusion after the first session and configuration of the lesion that favored endovascular treatment. Onyx was used only in two patients. PVA particles used were 250 μ or above in size. Size of the particle was decided based on the lesion as well as microcatheter used. Particles were admixed with contrast and no saline was added as it could lead to change in the size of particles. Injection was carried out slowly continuously looking for stasis as well as intermittent angiogram for looking at obliteration of the nidus/fistula. In few patients detachable coils or fibre coils were also used.

Post procedural management:-

The patient was shifted to the neurological ICU for 24 hours after the procedure. We routinely give antiobiotics for 3-5 days. Injection Methyl prednisolone 1gm I.V is given for 5 days. If a fresh neurological deficit is seen following procedure, high dose methylprednisolone (5.4mg/Kg body weight) is administered intravenously over 24 hrs. Steroids are tapered slowly and then stopped. In cases where significant venous stasis is noted Injection heparin 5000 units 6 hourly or Inj Flaxiparin 0.3-0.4 mL 12 hourly is injected for 2 days.

Following data was collected for each patient for the purpose of analyses.

Name :

Age :

Sex :

Address :

Hospital No :

Co-morbid illness :

Clinical features :

First symptom :

Presenting complaints:

Duration :

Onset : Sudden Vs progressive

Weakness: Symmetrical/symmetrical/paraparesis/Quadriparesis/monoparesis, onset, progression

Sensory impairment: present/absent, paresthesias

Neck pain & backache: Yes/No, Radicular/non-radicular, site [cervical /thoracic /lumbar]

Bladder : hesitancy/retention/incontinence, history of catheterization

Bowel : constipation/incontinence

History suggestive of CNS involvement: headache/loss of consciousness /seizure

Clinical course :

Examination :

GCS :

Tone/Spasticity : Upper limb/Lower limb, Grade

Power : Upper limb/Lower limb, Grade 1-5

DTR : Upper limb/lower limb/abdominal reflexes

Clonus : yes/no

Plantars :

Sensory : Touch/pain/vibration/position/temperature,Level

Spinal shock :

Preprocedure modified Aminoff and Logue scale score of disability

Classification of gait disturbance

Grade 0 : normal gait and activity

Grade 1 : leg weakness or abnormal gait, no restricted activity

Grade 2 : grade I with restricted activity

Grade 3 : requires cane or similar support for walking

Grade 4 : requires walker or crutches for walking

Grade 5 : unable to stand, confined to bed or wheelchair

Classification of micturition

Grade 0 : normal

Grade 1 : hesitance, urgency, or frequency

Grade 2 : occasional urinary incontinence or retention

Grade 3 : total urinary incontinence or retention

Classification of defecation

Grade 0 : normal

Grade 1 : slight constipation, react to laxation

Grade 2 : occasional incontinence or severe constipation

Grade 3 : total incontinence

MRI :

Level/signal alteration/cord swelling/ atrophy/extent/ flow voids/enhancement/ hemorrhage

Angiogram:

Type/Number of feeders /feeders –radiculomedullary/radiculopial,ASA/PSA Angioarchitecture of nidus- fistula/nidus/feeding artery aneurysm/angiopathy features/ intranidal aneurysm / pseudoaneurysm

Venous drainage: venous pouch/venous ectasia/aneurysm /drainage/thrombus / venous congestion

Treatment:

Date :

Embolization/Surgery :

No of treatment sessions :

Embolizing material :

No of feeders embolized :

Microcatheter / microwire :

Complication : Per-operative/ post operative

Post op management :

Steroids : Yes/No

Heparin/Flaxiparin : Yes/No

Ventilator support :

Management of complication :

Per-op :

Post-op :

Modified Aminoff and Logue scale of disability:

- Post procedure
- At discharge

Follow up

Date

Duration

Modified Aminoff and Logue scale of disability

Results

RESULTS

We analyzed 74 cases of spinal vascular malformations evaluated at our hospital in last 25 yrs. These included 26 cases of spinal dural arterio venous fistula or Type 1 lesions, 17 cases of perimedullary fistulas or Type IV lesions, 28 cases of spinal AVM and 3 cases of paraspinal lesions. The following are the results of each type presented separately.

SPINAL DURAL ARTERIOVENOUS FISTULA:

A total 26 patients (24 males;2 female) of SDAVF were included in our study. Mean age of the patients at the time of diagnosis was 52 yrs (Age range 29-81 yrs). Only one patient was less than 30 yrs at the time of diagnosis. Mean duration of illness was 18 months (range 7 days-6 yrs). Maximum number of patients were in there 6^{th} decade at the time of diagnosis (9 patients).

Early symptoms were consistent with myelopathy, typically worsening from initial presentation to the time of diagnosis. At the time of initial presentation, patients reported lower extremity weakness (10 patients, 38%), sensory symptoms (4 patients, 15%), back pain (10 patients, 38%), and urinary symptoms (3 patients, 11%). Radicular pain was present in 6 patients. Typical neurogenic claudication pain was noted in one patient. By the time diagnosis was made weakness was present in 25 patients (96%), urinary symptoms 25 patients (96%), sensory symptoms 23 patients (92%), paresthesias 8 patients (31%) and erectile dysfunction 4 patients (15%). Due to claudication symptoms in one patient diagnosis of lumbar canal stenosis was made and hence laminectomy was done. Following this patient symptoms were aggravated and further evaluation showed vascular lesion. In another case initial urinary complaints were attributed to the prostate disease and he underwent transurethral resection of prostate. Two patients underwent surgical exploration as possibility of tumor was considered. One patient was having spinal tuberculosis and was receiving ATT.

In all but one patient disease onset was slow and progressive. However in one case there was short history of acute onset weakness with sphincter involvement. In 11 patients symptoms were asymmetric while remaining 15 had symmetrical clinical features involving both sides of the body equally. Clinical features were usually progressive however fluctuating course of illness was noted in one patient where history of spontaneous improvement and worsening of symptoms was seen.

Diagnostic work-up included MRI in 24 patients. It most commonly showed increased signal intensity in the center of the spinal cord with peripheral sparing on T2-weighted images and abnormal blood vessels on surface of the spinal cord. Based on imaging findings presumptive diagnosis of vascular malformation was made in 22 patients. In three patients, only cord hyperintensity was seen on MRI. In two patients initially diagnosis of spinal cord tumor was considered. On surgical exposure definitively diagnosis of SDAVF was made in one case and in another case cord biopsy was taken. In both the cases subsequent angiography led to the final diagnosis of SDAVF. Myelography was used to make the initial diagnosis in two patients. This investigation revealed irregular dilation of the subarachnoid veins. In both the patients, spinal angiographic results confirmed the diagnosis. Intramedullary enhancement on immediate post contrast study was seen in 5 patients. Long term follow up was available in one of these patients in whom clinical recovery was not significant despite successful closure of fistula.

Angiography successfully showed the fistula in 25 cases including two cases where MRI findings were equivocal and diagnosis of tumor was considered. In one patient diagnostic angiogram showed common segmental artery supplying both the anterior spinal artery (artery of Adamkiewicz) and the fistula, endovascular curative embolization was not attempted. Follow up angiogram showed spontaneous closure of the fistula and mild clinical improvement was noted. However, on subsequent follow up patient remained bed bound. Definitive Patient refused for repeat MRI and angiography. In another case MRI showed features of congestive myelopathy with abnormal flow voids. However angiography done was found to be normal. Repeat MRI done showed evidence of thrombosis in the vessels and decreased conspicuity of flow voids. Definitive diagnosis could not be made however in view of history low backache and radicular symptoms possibility of spontaneously closed SDAVF was kept.

Feeding intercostals artery were usually seen coming from the mid and lower thoracic spine D6 to D12 vertebrae. In two cases lumbar arteries were the source of feeding dorsospinal artery and in one case lateral sacral artery gave rise to the feeding artery. No cases of cervical dural fistula were seen in our study. None of the cases showed clinical or radiological evidence of bleed. Feeders were noted from right side in 14 cases and left side in 11 cases. In one case angiogram was normal. Fistula was seen being supplied by single feeder in 24 patients, while in one case two feeders were noted.

Venous drainage was ascending as well descending type. Ascending drainage in to the posterior fossa was seen in 2 patients; however no clinical or radiological evidence of subarachnoid hemorrhage was noted in these two patients.

In all the cases embolization was done using Glue. In none of the cases of SDAVF, particles or onyx were used. Glue concentration used, ranged from 15% to 33%. In 22 patients single embolization session was carried out. In remaining case two sittings were needed as during first attempt there was feeder perforation which was managed conservatively and procedure was abandoned. During second sitting embolization was successfully done. In one of the patient following embolization initially good clinical improvement was noted, however on follow up there was again deterioration. Repeat angiogram showed filling of the fistula through above intercostal artery, so there was recurrence possibly due to the failure of occlusion of the draining vein. Second sitting of embolization was done. In two cases only partial embolization could be done and patients were referred for surgical treatment. Over all in 24 patients (92%), 25 therapeutic procedures were carried out, in the remaining two spontaneous cure occurred. In two patients (8%) treatments was partial and were referred for surgery. Recanalization occurred in two patients (8%) of which one underwent repeat embolization and improved while other did not return for MRI and angiogram.

Minor complications were noted in 3 patients (12%). They included feeder perforation and transient worsening of motor power after the procedure. In the first case heparin was immediately reversed and procedure abandoned. In the second case possible reason was slowing of flow in the venous channels resulting in the increased cord congestion. Patient was started on heparin for two days and there was spontaneous improvement in the weakness.

Clinical follow up of greater than 3 months was available in 14 treated patients. Mean follow up duration was 19 months (range 1 month to 8 yrs).

Follow up angiogram after successful embolization was done in 2 patients and in both cases it was normal. In three patients MRI was done, which was normal in one patient and showed regression of abnormalities in other two. Comparison of admission Modified Aminoff Logue score with the post embolization follow up score revealed complete cure in 2 patients (~8%), improvement in 15 patients (57%), unchanged status in 7 patients (26%) and in the remaining two patients procedure was not done. Improvement in gait and motor function was seen in 12 patients while urinary symptoms improved in 10 patients. Mean ALS at admission was 5.11 while mean score at follow up was 3.66.

Table 1: Chi square test comparing pre-procedure gait disability scores with post procedure gait disability

	Value	**Df**	**Asymp. Sig. (2-sided)**	**Exact Sig. (2-sided)**	**Exact Sig. (1-sided)**
Pearson Chi-Square	15.385[a]	1	.000		
Continuity Correction[b]	12.346	1	.000		
Likelihood Ratio	19.605	1	.000		
Fisher's Exact Test				.000	.000
Linear-by-Linear Association	14.769	1	.000		
N of Valid Cases	25				

Risk estimate

		95% Confidence Interval	
	Value	**Lower**	**Upper**
For cohort P0G1 = 0	4.333	1.606	11.691
N of Valid Cases	25		

Improvement noted was statistically highly significant with p value <.001 and odds ration >2 (4.33).

Table 2: Chi square test comparing pre-procedure micturition disability scores with post procedure disability

	Value	Df	Asymp. Sig. (2-sided)	Exact Sig. (2-sided)	Exact Sig. (1-sided)
Pearson Chi-Square	11.111[a]	1	.001		
Continuity Correction[b]	7.910	1	.005		
Likelihood Ratio	12.655	1	.000		
Fisher's Exact Test				.002	.002
Linear-by-Linear Association	10.667	1	.001		
N of Valid Cases	26				

Risk estimate

		95% Confidence Interval	
	Value	**Lower**	**Upper**
For cohort POS1 = 0	2.250	1.084	4.671
N of Valid Cases	25		

Improvement in urinary disability noted was statistically highly significant with p value .001 and odds ration >2 (2.25).

Duration of symptoms at the time of diagnosis and gait disability score were compared using Chi square test to look for any association between them.

Table 3: Chi square test comparing pre-procedure gait disability scores with duration of symptoms

	Value	**Df**	**Asymp. Sig. (2-sided)**	**Exact Sig. (2-sided)**	**Exact Sig. (1-sided)**
Pearson Chi-Square	3.033[a]	1	.082		
Continuity Correction[b]	1.839	1	.175		
Likelihood Ratio	3.095	1	.079		
Fisher's Exact Test				.128	.087
Linear-by-Linear Association	2.921	1	.087		

Risk estimate

		95% Confidence Interval	
	Value	Lower	Upper
Odds Ratio for Duration1 (0 / 1)	4.050	.812	20.200
For cohort PPG1 = 0	1.938	.879	4.274
For cohort PPG1 = 1	.479	.194	1.182
N of Valid Cases	25		

Results show existence of association between gait disability at the time of presentation and duration of illness as indicated by Odds ratio however it has not reached statistical level of significance possibly due to small sample size.

Table 3: Chi square test comparing pre-procedure micturition scores with duration of symptoms

	Value	Df	Asymp. Sig. (2-sided)	Exact Sig. (2-sided)	Exact Sig. (1-sided)
Pearson Chi-Square	.022[a]	1	.883		
Continuity Correction[b]	.000	1	1.000		
Likelihood Ratio	.022	1	.883		
Fisher's Exact Test				1.000	.598
Linear-by-Linear Association	.021	1	.885		
N of Valid Cases	26				

		95% Confidence Interval	
	Value	Lower	Upper
Odds Ratio for Duration1 (0 / 1)	.889	.186	4.244
For cohort PPS1 = 0	.957	.536	1.711
For cohort PPS1 = 1	1.077	.403	2.879
N of Valid Cases	26		

Results of Chi square test indicate no association between the severity of urinary symptoms and duration of illness.

PERIMEDULLARY FISTULAS:

In our study there were 17 cases of perimedullary fistula or Type IV lesions (15 males and 2 females). They included 9 patients (52%) of pediatric age group (7 males and 2 females). Mean age was 26 yrs (range, 6-62 yrs). Eleven patients (64%) had an acute onset of illness; while in the remaining 6 progressive symptoms were noted. In 8 (46%) patients there was clinical evidence of underlying hemorrhage as the cause of acute symptoms. In five of eight patients evidence of hemorrhage was seen noted on diagnostic imaging studies. In 2 patients, clinical features resembled intracranial hemorrhage and one of them underwent cerebral angiogram. In the other case frank SAH was noted on CT scan. Eight pediatric patients had an acute onset of clinical symptoms. Mean duration of illness was 18 months (1 week to 9 yrs). Clinical course of the illness was episodic or fluctuating type in 5 patients, which was interspersed with episodes of improvement and clinical deterioration. Most common clinical symptom was weakness (88%) followed by sensory impairment 14 patients (82%), backache 11 patients (64%) which was associated with radicular symptoms in 2 patients. Sphincter disturbances were noted in 10 patients (58%).

On MRI imaging most commonly observed imaging findings were flow voids and increased signal intensity on T2W images. In 4 patients evidence of hemorrhage was noted. In one patient who had presented with acute onset symptoms, MRI showed evidence of hyperintensity in the draining veins suggestive of hemodynamic changes in the shunt or possibly thrombosis. While in another case large draining venous sac showed evidence of peripheral thrombus. Two patients underwent myelography which revealed irregularity and serpenginious filling defects in subarachnoid contrast column.

Final diagnosis was reached on the basis of diagnostic angiogram. There were 10 cases of Merlands Type IV a, 5 cases of Type IVb and 2 cases of Type IVc. In 15 patients lesions were single while in two patients multiple lesion were noted. (2 in each case). By revised classification of Rodesch et al; 14 of our cases were micro AVF and three can be classified as macro AVF. In none of our 3 patients harboring macro AVF association with HHT was found.

A total of 19 lesions were seen in 17 patients of which 11 were seen in relation to the conus and 2 in filum terminale, one each in relation to cauda equina root and cervical spine. While remaining lesions were noted in the dorsal spine. The patient harboring two type 1 lesions one in relation to conus and other in the cauda equine roots may be a case of SAMS. Supply was noted from both ASA and radiculopial feeders. In two patients feeding artery aneurysms were seen of which one was Type IVc and other was Type IV b lesion. Angiopathy features were not seen in any case. Evidence of venous thrombosis could be appreciated in two patients while two patients showed presence of venous aneurysms. One of the patient with small distal aneurysm had SAH in the past. Pial venous reflux and intrinsic venous congestion could be seen in 5 patients. Three of these five patients had acute presentation.

Table 4: Chi square test comparing venous ectasia/venous pouch and presence of hemorrhage

	Value	Df	Asymp. Sig. (2-sided)	Exact Sig. (2-sided)	Exact Sig. (1-sided)
Pearson Chi-Square	2.837[a]	1	.092		
Continuity Correction[b]	1.418	1	.234		
Likelihood Ratio	2.915	1	.088		
Fisher's Exact Test				.153	.117
Linear-by-Linear Association	2.670	1	.102		
N of Valid Cases	17				

Risk estimate

		95% Confidence Interval	
	Value	**Lower**	**Upper**
Odds Ratio for Venous ectasia (0 / 1)	5.833	.696	48.873
For cohort hem = 0	2.450	.709	8.463
For cohort hem = 1	.420	.146	1.208
N of Valid Cases	17		

Test results indicate association between presence of venous ectasia/venous pouch on angiogram and clinical event of hemorrhage. However association is not statistically significant possibly because of the small sample size.

Sixteen patients were treated by endovascular means while one patient underwent surgery. Sixteen patients underwent a total of 19 treatment sessions. Total nineteen feeders were embolized. Nine anterior spinal artery and ten posterior spinal axis embolizations were carried out. Embolic agents used include PVA particles, hydrogel, coils, glue and Onyx. In ten patients only glue was used, in two patients particulate embolization was done. Onyx and coils were used as primary embolic agent in one case each. In the remaining different agents in combination were used.

Complications included transient and permanent weakness following procedure on 5 occasions (26%), feeding artery perforation and severe anterior spinal artery spasm in the one case each. Clinical deterioration was transient or minimal in 3 patients and permanent in two patients. Feeding artery perforation (intercostal artery) was managed by injection of 50% glue proximal to the site of rupture. In another case worsening took place on 10 post procedure and possibly represents a case of progressive thrombosis of the draining veins. He was started on high dose intravenous methylprednisolone and Heparin. Patients overall clinical outcome was poor. Second case of clinical worsening occurred following Onyx embolization in his second sitting. In this patient Type IV a fistula was noted in relation with filum and following 1st embolization, disease was well controlled for ~4 yrs. However there was clinical worsening and repeat angiogram showed filling of the fistula. During second time onyx was used in the ASA; severe spasm leading to stasis of flow in the artery was noted. Patient was aggressively managed with lumbar drainage of CSF and high dose of systemic steroids. Mild clinical improvement was seen following conservative management.

Good outcome i.e. clinical status better than pre embolization was noted in 11(68%) patients where complete radiological closure of the fistula was achieved. In one patient good outcome was noted following first embolization however clinical worsening and poor outcome after 2nd embolization. Poor outcome was seen in another patient who had worsened after 10 days of embolization and possible reason was thrombosis of the draining veins leading to clinical worsening. So overall 2 patients had bad outcome i.e. post procedure clinical status worse than pre-procedure clinical status. Fair result was obtained in remaining 3 patients where disease was well controlled and no clinical worsening took place. One case who underwent surgery had improved clinical status, so overall improvement noted in 72% patients.

Clinical follow up of greater than 3 months was available in 13 treated patients. Mean follow up duration was 24 months (range 1 month to 8 yrs). Follow up angiogram and MRI were done in 3 patients each and they were normal in all the cases thus confirming successful exclusion of fistula in these 6 patients. Clinical outcome was determined by comparison of admission Modified Aminoff Logue score with the post embolization follow up score revealed normal score in 8 patients (47%), improvement in 4 patients (23%), unchanged status in 3 patients (18%) and in the remaining 2(11%) worsening was noted. In one patient where surgery was done and outcome was good with minimal disability on long term follow up.

ARTERIOVENOUS MALFORMATIONS:

There were 28 patients with spinal arteriovenous malformations (16 males, 12 females). Mean age of the patients was 28 years (Age range 8-52 yrs). There were 3 pediatric age group patients. Onset of illness was acute in

10 (35%) patients and progressive disease was noted in the remaining 18 patients. Clinically disease was asymmetrical in 18 patients (64%). In 9 of 10 patients with acute presentation, there was clinical evidence of hemorrhage and in two patients CSF examination showed blood. MRI showed evidence of bleed in 5 of these patients while in one case there was expansion of the conus with subarachnoid filling defects suggestive of hematomyelia with vascular malformation. Clinically weakness was present in 27 patients (96%), sensory impairment was noted in 23 (82%) patients, sphincter disturbances in 22 patients (78%), paresthesias were present in 8 patients (28%) and erectile dysfunction in 5 patients (17%). Four patients were bed bound at the time of presentation. Mean duration of illness was 19 months (7days-17 yrs). Fluctuating course of illness was noted in 8 patients.

Associated conditions include chest wall hemangioma in a case of upper dorsal AVM, skin malformation in upper thoracic and lower cervical dermatomal distribution in one patient. One patient had walking difficulty since birth while another developed it at the age of 9 months. Another patient showed multiple café u lait spots, however no other feature s/o neurofibromatosis were seen. One patient had undergone spinal surgery in the past for lower limb weakness, however details of the surgery and investigative work up were not available.

Diagnostic imaging included MRI examination in 20 patients and myelography in 9 patients. One patient had undergone both the investigations. Intramedullary flow voids could be seen in 17 patients thus confirming the intramedullary nature of the lesion. Hemorrhage could be made out on imaging in 6 patients with evidence of hematomyelia in one case. Other common abnormalities include subarachnoid flow voids, cord signal changes and cord deformation. In myelograms serpengenious filling defects were seen while in one case complete block was noted in lower dorsal spine. In one case there was conus expansion due to hematomyelia. CT scan was done in three patients. It showed hematomyelia with extramedullary AVM in one case, SAH in the other two cases.

In all the cases diagnostic angiogram was done to confirm the diagnosis as well as plan the interventional procedure. In 13 (46%) patients lesion were dorsolumbar or lumbar in location, dorsal in 10 (35%) patients while in the remaining cervical or cervicodorsal lesion were noted. In two patients in addition to the spinal intramedullary lesion adjacent structures of the same myelomere were involved like dural, vertebrae and paraspinal tissue constituting metameric form of the spinal vascular malformation. In one patient there was conus lesion present and in addition a separate nidus with different venous drainage was noted. Arterial feeder was noted from the lateral sacral artery thus possibly representing a cauda equine root lesion which again makes it a metameric form of Spinal AVM. In two patients corresponding dermatomes were showing skin lesions in addition to the spinal AVM, which may be considered a incompletely expressed form of the SAMS.

Feeders were single in 9 patients and multiple in the remaining cases ranging from 2 to up to 10 feeders. Single feeders were radiculomedullary in 6 and radiculopial in 3 patients. In patients with multiple feeders, ASA was seen supplying the nidus in 14 cases. In 4 patients feeding artery aneurysms were seen. In one patients hugely dilated and ectatic radiculopial feeding arteries were seen feeding a large intranidal fistula. In 7 patients intranidal aneurysms were seen. There were two pseudoaneurysms diagnosed on the basis of fluid-2 level and retention of contrast in them till late venous phase. In both these patients there was history of hemorrhage. In five patients harboring intranidal aneurysm hemorrhage was seen. None of the patients harboring feeding artery aneurysm had history of bleed. Hugely dilated venous ectatic sac were seen in 3 patients. In 2 of these patients there was acute clinical presentation and hemorrhage was noted. Intracranial venous drainage was seen in 2 patients and one of them presented with SAH.

Table 5: Chi square test to look for any association between feeding artery aneurysm and hemorrhage

	Value	Df	Asymp. Sig. (2-sided)	Exact Sig. (2-sided)	Exact Sig. (1-sided)
Pearson Chi-Square	2.211[a]	1	.137		
Continuity Correction[b]	.826	1	.364		
Likelihood Ratio	3.410	1	.065		
Fisher's Exact Test				.273	.189
Linear-by-Linear Association	2.132	1	.144		
N of Valid Cases	28				

Risk estimate

		95% Confidence Interval	
	Value	**Lower**	**Upper**
For cohort Hemorrhage = No hemorrhage	.625	.458	.852
N of Valid Cases	28		

Our results indicate that there is no association between feeding artery aneurysm and the hemorrhage in patients of spinal AVM.

Table 6: Chi square test to look for any association between intranidal aneurysm and hemorrhage

	Value	Df	Asymp. Sig. (2-sided)	Exact Sig. (2-sided)	Exact Sig. (1-sided)
Pearson Chi-Square	2.674[a]	1	.102		
Continuity Correction[b]	1.365	1	.243		
Likelihood Ratio	2.551	1	.110		
Fisher's Exact Test				.165	.123
Linear-by-Linear Association	2.579	1	.108		
N of Valid Cases	28				

Risk estimate

		95% Confidence Interval	
	Value	Lower	Upper
Odds Ratio for Intranidal aneurysm (No intranidal aneurysm / Intranidal aneurysm)	4.267	.703	25.878
For cohort Hemorrhage = No hemorrhage	1.778	.731	4.321
For cohort Hemorrhage = Hemorrhage	.417	.154	1.131
N of Valid Cases	28		

Test result indicates association between the presence of intranidal aneurysm and hemorrhage (Odds ratio 4.2), however it has failed to reach the level of statistical significance possibly due to small sample size (p value .165).

Table 7: Chi square test to look for any association between venous ectasia/pouch and hemorrhage

	Value	Df	Asymp. Sig. (2- sided)	Exact Sig. (2-sided)	Exact Sig. (1-sided)
Pearson Chi-Square	1.116[a]	1	.291		
Continuity Correction[b]	.318	1	.573		
Likelihood Ratio	1.065	1	.302		
Fisher's Exact Test				.352	.280
Linear-by-Linear Association	1.077	1	.299		
N of Valid Cases	28				

Risk estimate

		95% Confidence Interval	
	Value	**Lower**	**Upper**
Odds Ratio for Venous ectasia (No venous ectasia / venous ectasia)	2.667	.417	17.046
For cohort Hemorrhage = No hemorrhage	1.455	.628	3.370
For cohort Hemorrhage = Hemorrhage	.545	.191	1.561
N of Valid Cases	28		

Test result indicates weak association between the presence of venous ectasias/venous pouch and presence of hemorrhage. Association is not statistically significant, however odds ratio is 2.667 indicating possibility of weak association.

Fifteen patients underwent endovascular therapy while rest of them either underwent surgery, refused treatment or are awaiting embolization. In one patient embolization was attempted but could not be accomplished. Total of 21 procedures were done and 43 feeders were embolized. Number of treatment sessions ranging from 1 to 3. Glue and particles were used for embolization. In 7 sessions of embolization only glue was used in concentration range from 15%-80% depending on the hemodynamics of the shunt. In 2 patients Glue and particles both were used. In remaining only particulate embolization was done. In 21 treatment sessions major clinical worsening was noted in 6 and transient and mild worsening in 4 patients. In two patients clinical worsening was due to feeder perforation which was inconsequent in one patient while in the second case there was SAH and artery had to be sacrificed by injecting glue. This patient following first embolization had transient increase in the symptoms, however on long term follow up patient remained relatively asymptomatic for 14 yrs, however he again became symptomatic and during attempted catheterization of feeding artery perforation occurred which was controlled immediately. In two cases post embolization cord involvement led to severe disability. Over all in 7 patients good outcome was seen (disability score better than preprocedure score), fair in two patient (unchanged or mild increase in disability) and poor in 6 cases (worse than preprocedure score). Arterial embolic complication led to the bad outcome in one patient and on follow up there was no symptomatology noted pertaining to the spinal lesion. In another case clinical worsening could be attributed to large extramedullary intradural lesion which developed after embolization and was found to be a granuloma, so in this case progressive symptoms cannot be attributed to vascular lesion alone. There were no hemorrhagic episodes on follow up in any of the patients and all patients who deteriorated had progressive worsening.

Five patients underwent surgery and post surgery good outcome was achieved in 3 patients while 2 had poor outcome. Mean follow up duration for treated cases is~ 4.2 yrs (1 month to 17 yrs).

PARASPINAL AVM AND VERTEBROVERTEBRAL FISTULA:

There were 2 cases of vertebral arteriovenous fistulae and one case of paraspinal AVM. There were two males and one female patient (Mean age 30 yrs). All the patients presented with progressive neurological deficit in form of progressive motor or sensory deficit. Sphincter involvement was present in all the cases.

One case of VVF was a known case of NF1 who presented with a 6-month history of humming sound over the left side of her neck, radicular pain in her left upper limb, paraesthesia, and progressive weakness of both upper and lower limbs. An MR imaging study showed an enlarged cervical epidural venous sac extending from C-3 to C-6 that compressed the spinal cord and caused cord hyperintensity on T2-weighted images. Angiography showed a high-flow 12- mm fistula with a large distal venous sac (32 × 36 mm) connecting the left VA and an emissary radicular vein at the level of the C5–6 intervertebral foramen. The fistula drained into dilated cervical epidural venous sacs from C-3 to C-6 and subsequently to extravertebral veins and the internal jugular vein. In addition evidence of vascular dysplasia due to underlying neurofibromatosis could be well made out. Initially surgical trapping was attempted, however could not be done. So subsequently endovascular coil embolization was done. Complete closure of fistula could not be achieved so direct puncture and coil deposition was done. On follow up patient showed remarkable recovery and her initial Aminoff Logue score Grade 5/3/1 improved to Gr1/0/0 at 6 month follow up. Second patient of vertebro-vertebral fistula presented with insidous onset slowly progressive weakness. On auscultation loud bruit was present in left side of neck. CT angiography and DS angiography showed large paravertebral venous sac with two feeders, one enlarged vertebral artery and other one possibly representing duplicated origin of the left VA coming directly from the aortic arch. Venous drainage was in to the dilated epidural venous plexus and then via radicular veins in to the paravertebral veins. Patient is awaiting embolization.

Third case of was of a 20 year old male who presented with progressive weakness and numbness of both lower limbs. In the past patient had undergone surgery twice for a recurrent swelling of infrascapular region. CT scan showed large paravertebral AVM with destruction of vertebrae. DS angiogram showed multiple intercostal and thyrocervical artery feeders. Patient was embolized twice with nearly symptom free 3-year period in between. Following second embolization again significant improvement was noted. Patient again presented 6 years after his onset of symptoms with complains of CHF and underwent surgical resection, however he died in post operative period.

Figures

FIGURES

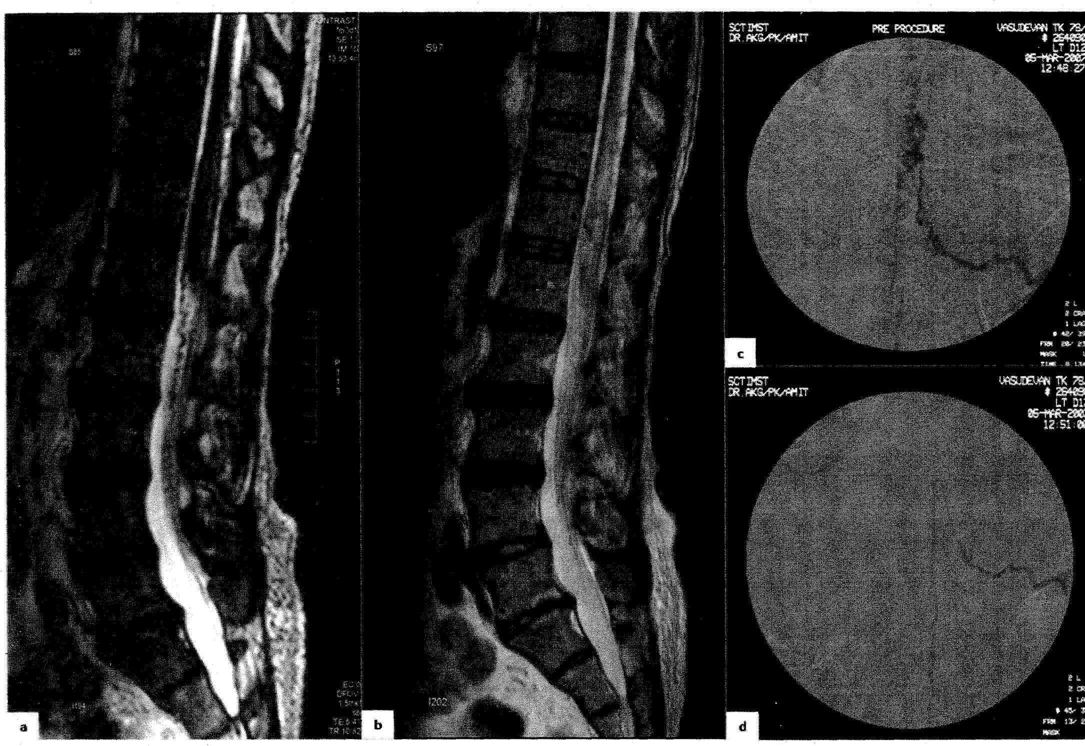

Fig 1: Patient was 78 year old male who presented with progressive weakness and urinary incontinence. a) Sagittal CISS 3D image of MRI shows multiple flow voids on the dorsal surface of the spinal cord and along the cauda equine nerve roots. b) Sagittal T2W image of LS spine shows increased signal intensity as well as expansion of the lower thoracic cord and conus. c) Spinal angiogram: Left D12 intercostal artery injection shows evidence of SDAVF with both feeding artery and anterior spinal artery arising at same level. d) Immediate repeat angiography showed spontaneous closure of fistula.

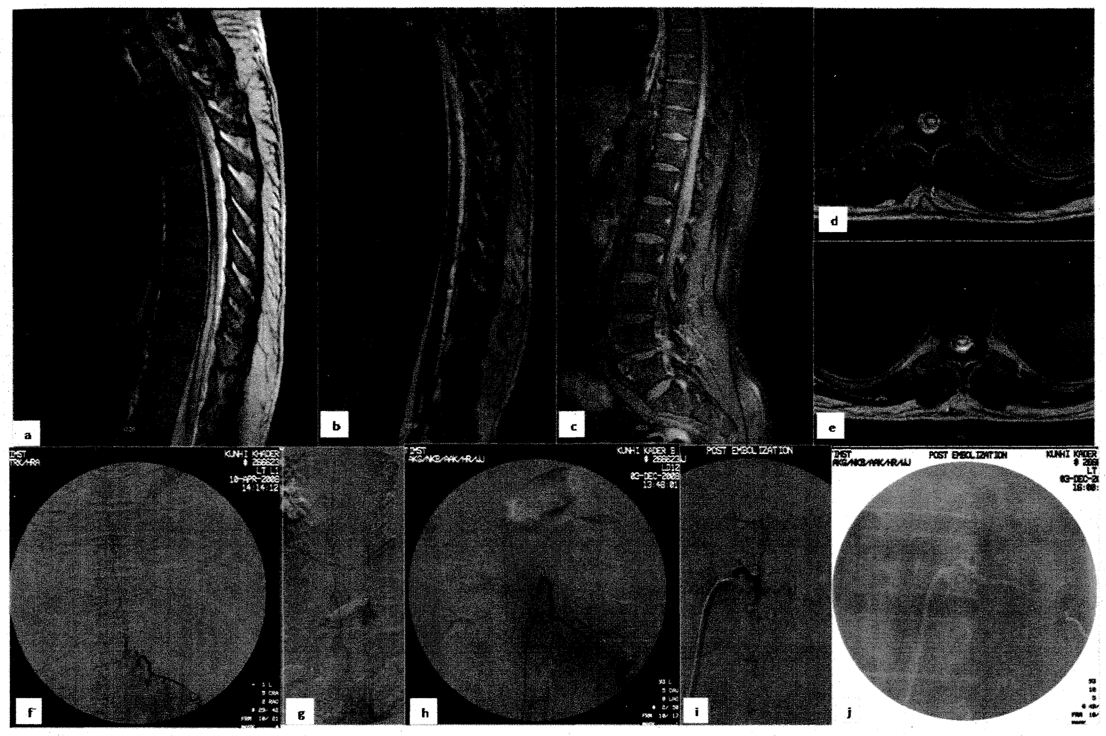

Fig 2: Fifty eight year old male presented with history of chronic backache, progressive weakness and urinary hesitancy. a) Sagittal T2W image of spine shows increased signal intensity of the spinal cord extending up to the mid thoracic cord level. b) Sagittal CISS 3D image very well demonstrate the flow voids on the dorsal surface of the cord. c) Sagittal post contrast T1W image shows mild enhancement of the cord parenchyma. d & e) Axial T2W images of the spine showing increased signal intensity localized to the centre of the cord with peripheral hypointense rim indicative of congestive myelopathy. f & g) Left L1 lumbar artery injection shows SDAVF and ascending drainage in to the perimedullary venous plexus. h) Anterior spinal artery filling noted from a vessel at different level (left D12 intercostal artery). i) Post embolization angiogram shows no filling of fistula. j) Post embolization glue cast can be appreciated in the fistula and draining vein, while distal intercostal artery shows fibre coil, which was deposited for facilitating microcatheterization of the feeding radicular artery.

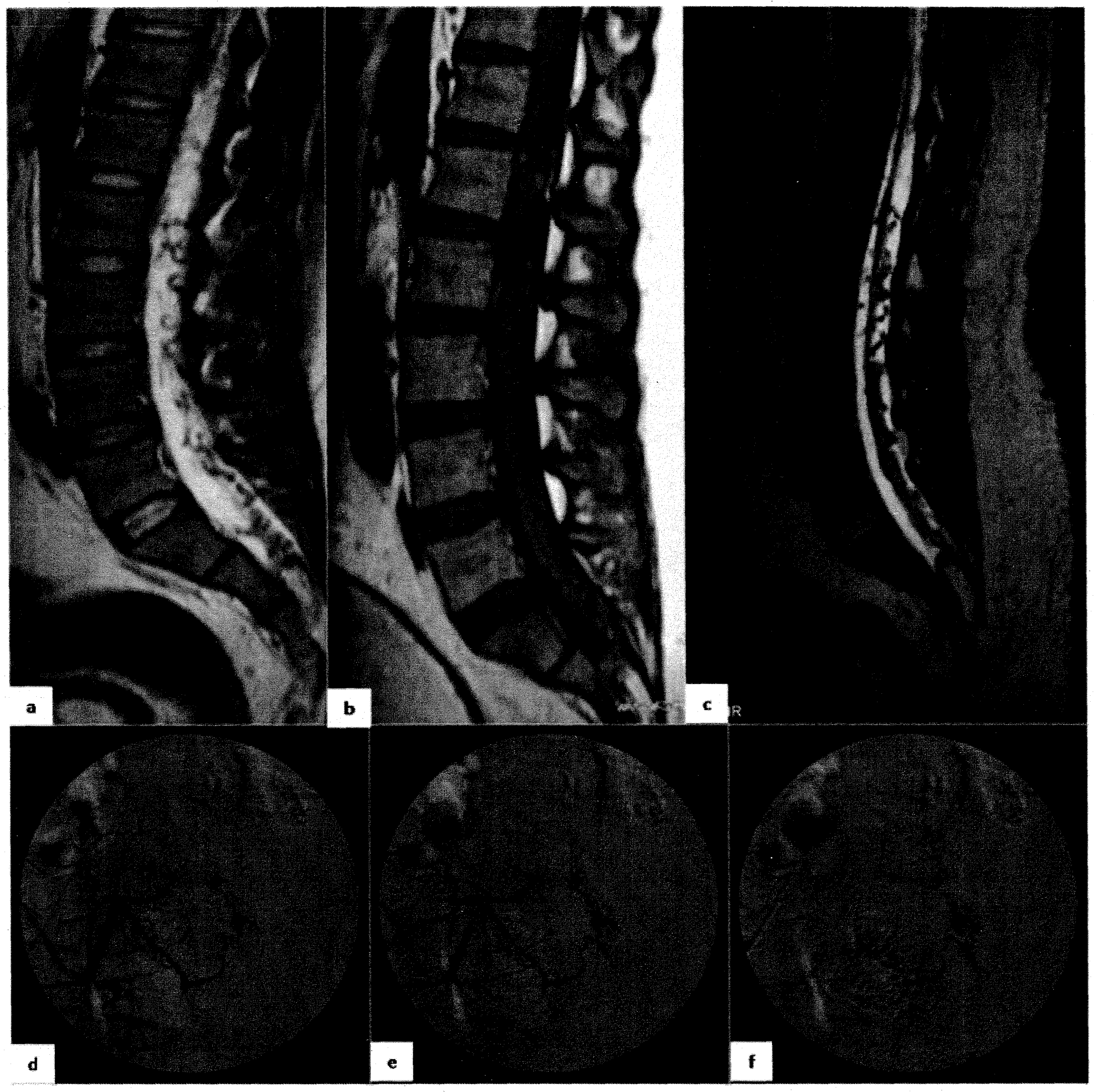

Fig 3: Patient was 35 year old male who presented with acute onset rapidly progressive bilateral lower limb weakness with sphincter involvement. a- c) Sagittal T2W, T1W and 3D T2W TSE sequences respectively showing low lying tethered cord with multiple flow voids in the subarachnoid space. d, e) Internal iliac artery posterior branch angiogram shows SDAVF with two feeders seen arising from lateral sacral artery. f) Angiogram following closure of the upper feeding artery with glue shows residual filling via second feeder. This feeder could not be safely catheterized so patient was referred for surgery. Improvement in gait was noted after partial embolization; however sphincter disturbances persisted despite embolization.

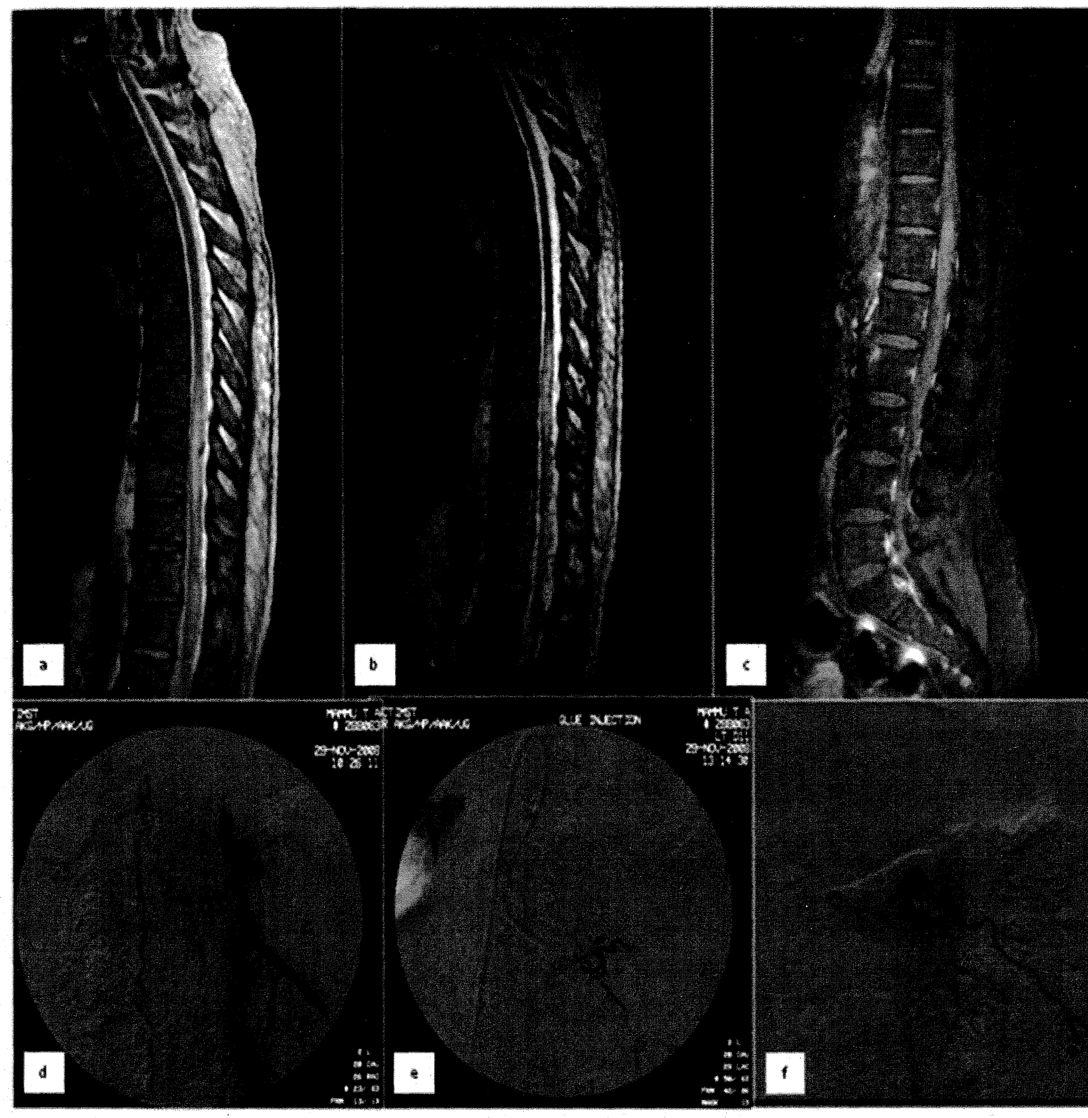

Fig 4: This is a case of 56 year old male patient who presented with insidious onset left lower limb weakness, bladder and sensory disturbances. a) Sagittal T2W image shows cord expansion and increased signal intensity. b) CISS 3D image shows prominent flow voids in the sub arachnoid space. c) Post contrast T1W image showing mild enhancement of the conus. d) Spinal angiogram; left D11 intercostal artery injection shows SDAVF with descending venous drainage. e) Image shows glue injection through selectively positioned microcatheter in the dorsospinal artery feeding the fistula. f) Post embolization angiogram shows no abnormal filling of veins.

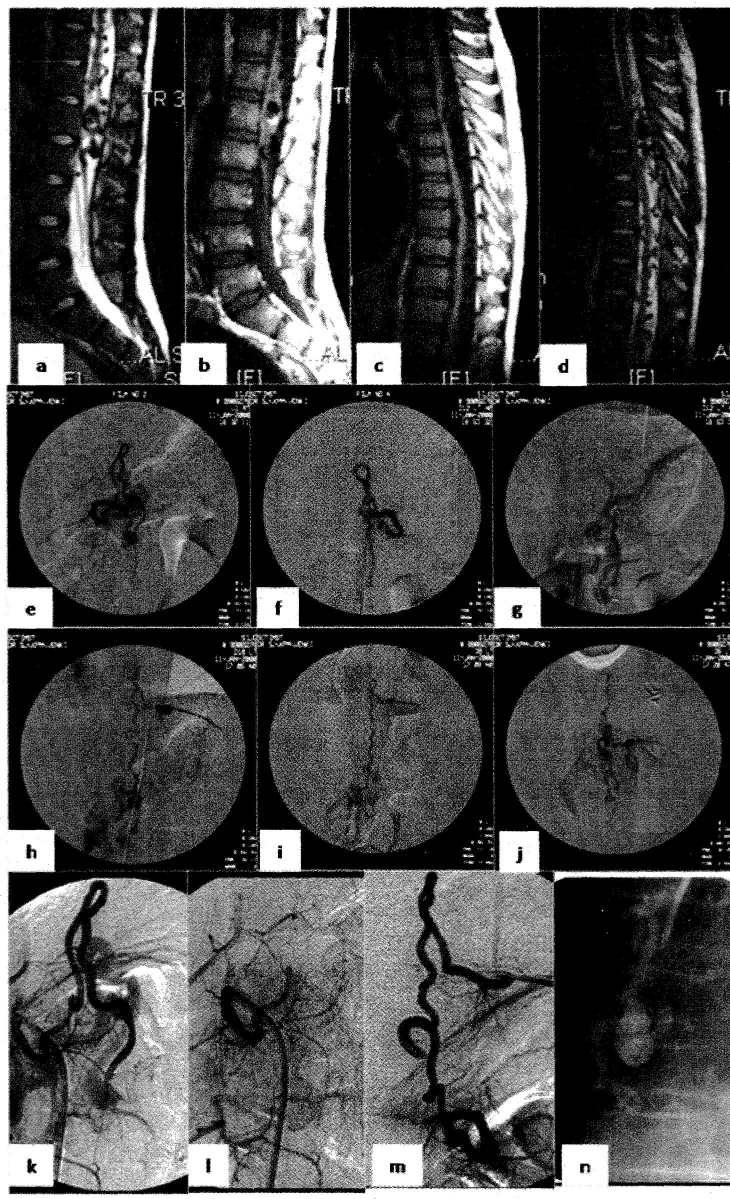

Fig 5: Images of fifteen year old male patient presented with acute onset backache with weakness of bilateral lower limbs. a-d) Sagittal T2W and T1W images of lumbo-sacral and dorsal spine show multiple prominent flow voids in the sub arachnoid space with two large venous sacs seen at L1 and D5 levels. Cord compression noted due to enlarged venous pouch at L1 level. Venous pouch at L1 level also shows evidence of partial thrombosis. There is no evidence of obvious hemorrhage to explain the patient's symptoms. e-i) Spinal angiography shows multiple feeders both radiculopial (e-h) and radiculomedullary (i, j), draining via single hole fistula in to a large venous pouch. Note the dilated venous sac and draining veins. j) Another lesion of similar morphology seen at D5 thoracic level. k) Angiogram with microcatheter positioned just proximal to the fistula. i) Following embolization with glue check angiogram shows no filling of fistula. m, n) Similarly other fistula was embolized and glue cast well seen filling the venous pouch and proximal draining veins in the unsubtracted image (n)

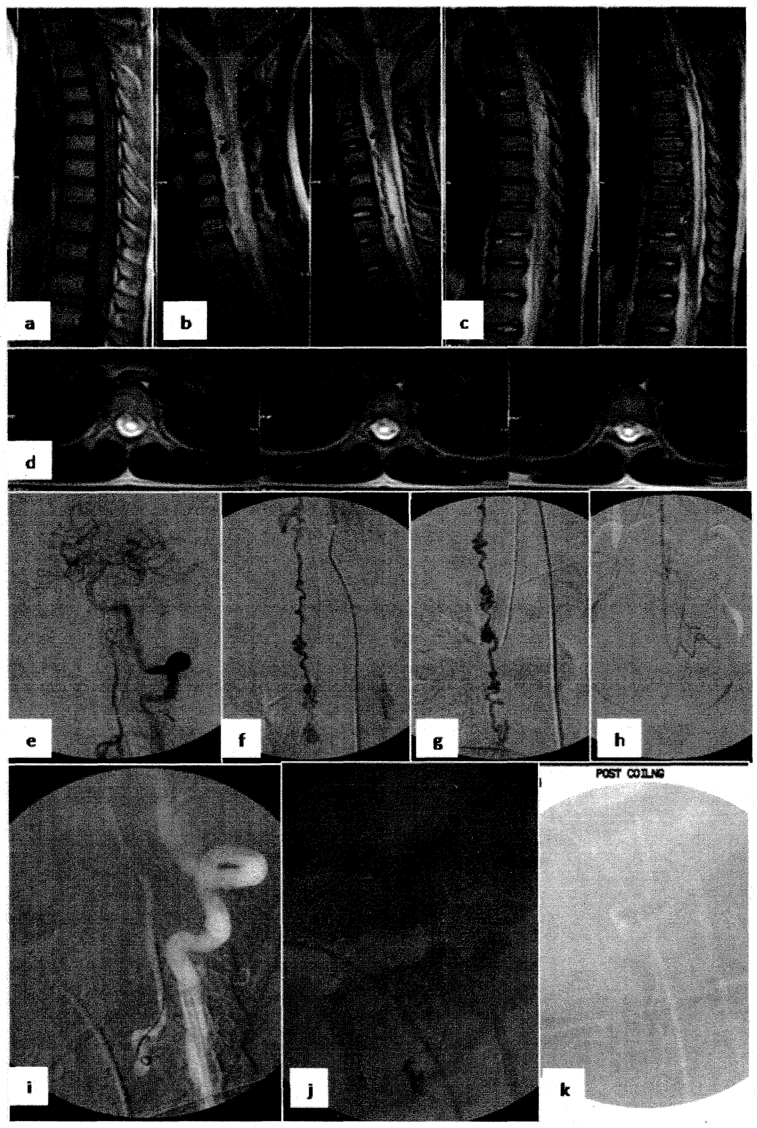

Fig 6: This is a 13 year old male patient who presented with acute onset rapidly progressive weakness of both lower limbs and sphincter disturbances. a) Sagittal T1W images show low intensity in the centre of cord. In addition a prominent flow void seen anterior to the spinal cord. b & c) Sagittal T2W images show extensive signal change in the cervicothoracic cord with a prominent flow void seen in anterior thecal space. d) Axial T2W image shows prominent increased signal in the centre of the cord suggestive of congestive myelopathy. e-h) Vertebral angiogram showed anterior perimedullary fistula at cervical level, supplied by anterior spinal artery, and draining through medullary vein all the way down in to the pelvic veins. Small venous aneurysm can be well seen on the draining vein (g). i-k) Anterior spinal artery was selectively cannulated using microcatheter and fistula was closed using 7 bare platinum coils. Following closure of fistula filling of the distal anterior spinal axis can be seen (k).

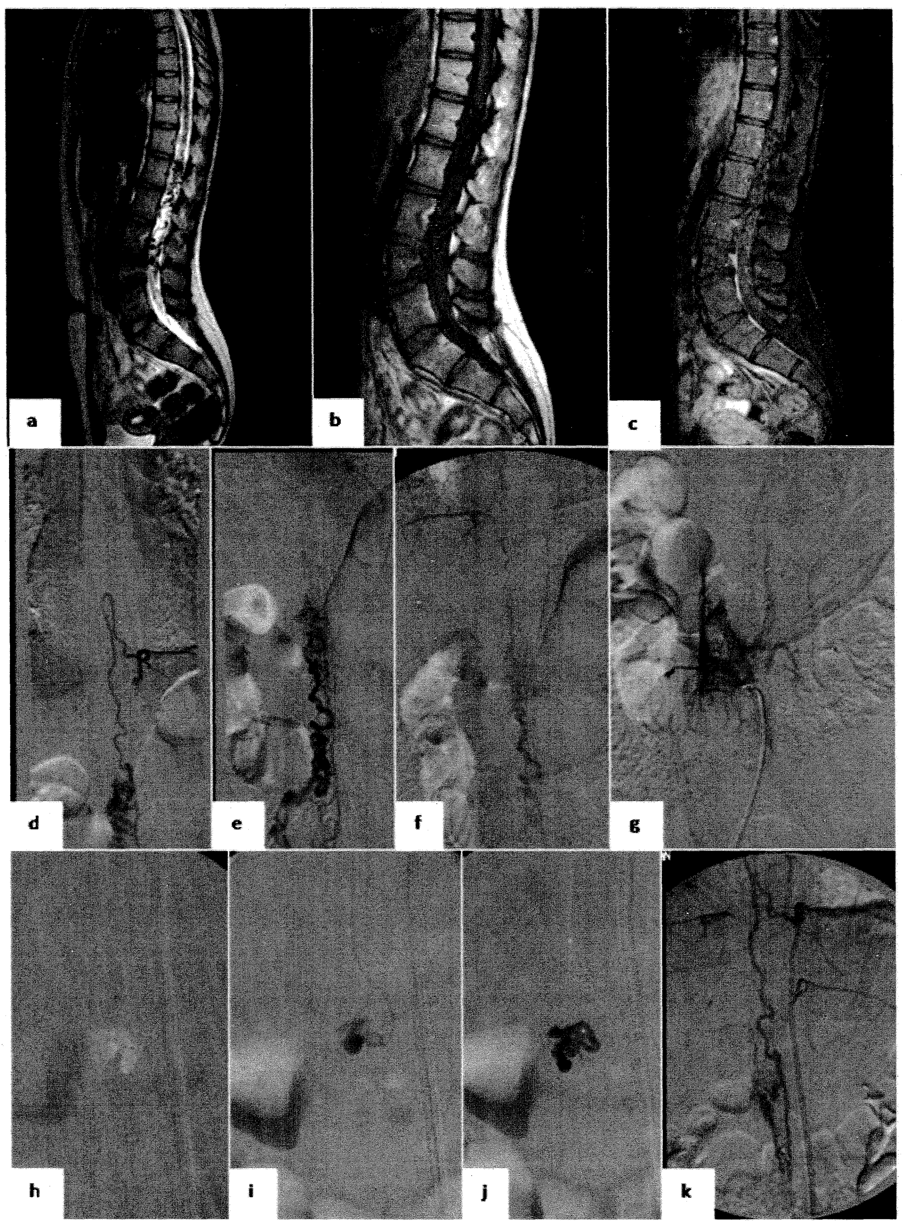

Fig 7: Twelve year old female presented with chief complaint of acute low backache followed by bilateral lower limb weakness. a-c) Sagittal T2W, T1W and postcontrast T1W images shows prominent flow voids in lumbosacral spine. In addition there is abnormal morphology of L4 vertebral body. d-f) Spinal angiogram showed multiple feeders both radiculomedullary (d) and radiculopial (f) feeding the AVM nidus. In addition there is evidence of right L3 level paravertebral AVM which is draining in to the epidural plexus (g). Anterior spinal axis was selectively catheterized and super-selective microcatheter injection showed a intranidal aneurysm (h,i). Glue was selectively injected in to the feeding artery (j). It partially obliterated the nidus and distal anterior spinal axis was preserved (k). Clinically patient improved and is asymptomatic 2 years follow up.

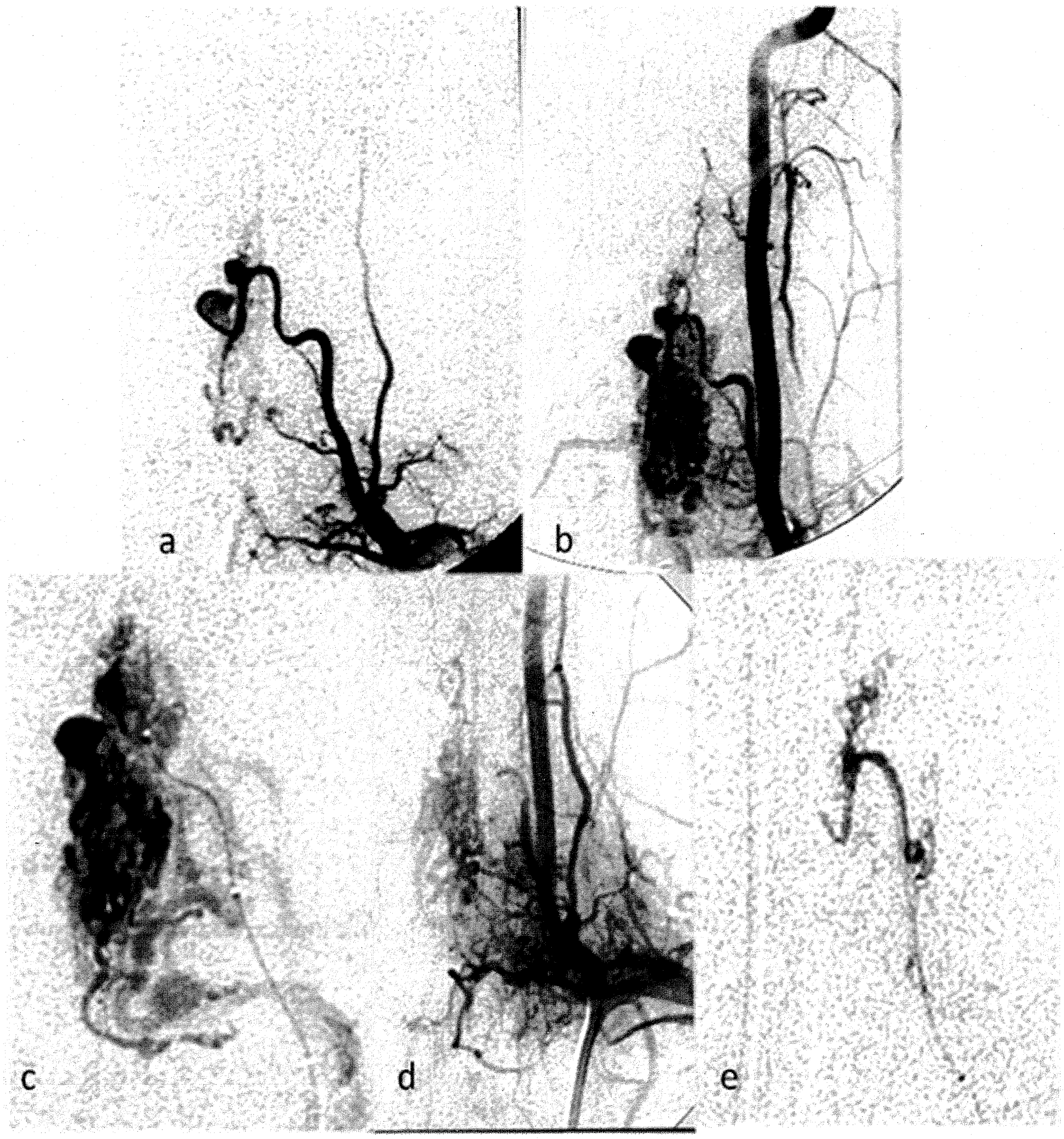

Fig 9: Spinal angiogram images of a 32 year old male who presented with chronic neck pain, paraparesis and urinary hesitancy. There is a large AVM nidus noted in the lower cervical spine, fed by radicular arteries from left vertebral artery (a-c). In addition two feeding artery aneurysms can be noted (a). Selective catheterization of the feeding artery done using a microcatheter and embolization was carried out using 250-355μ PVA particles. Check angiogram following embolization showed significant reduction of the AVM nidus (d,e). However following procedure patient developed worsening of symptoms. After 10 years, patient's motor power remained poor, however sphincter symptoms resolved and there was no hemorrhage or acute worsening in the follow up period.

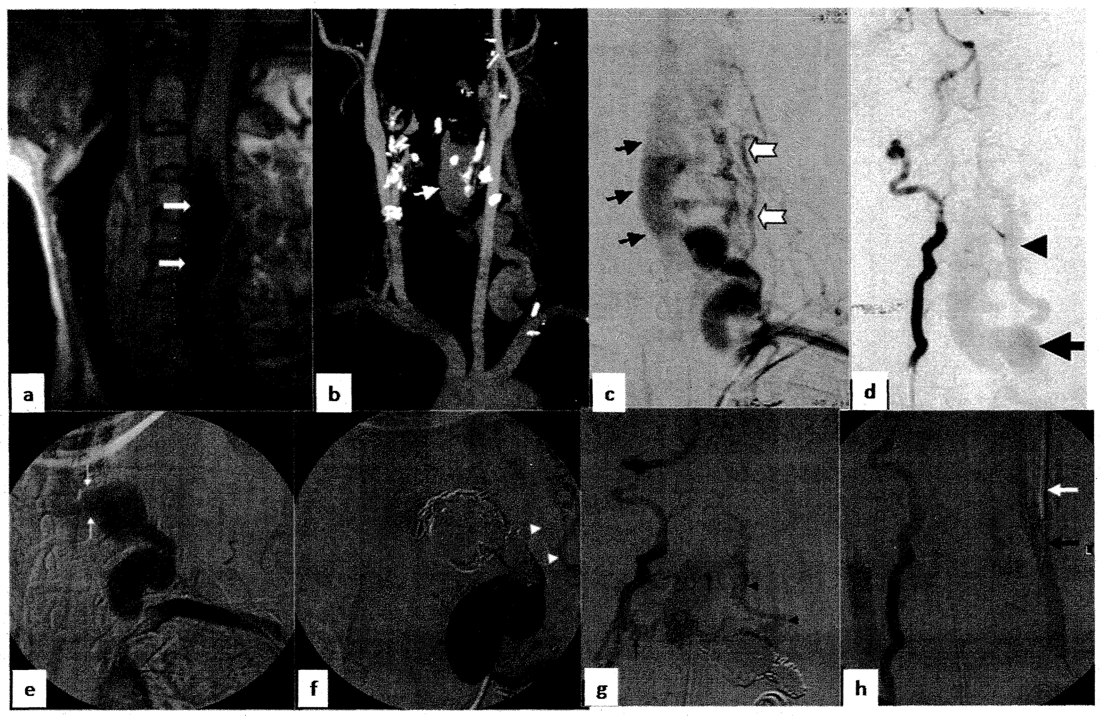

Fig 9: Twenty nine year old female patient with left lower cervical vertebro-vertebral fistula presented with quadriparesis and urinary retention. Sagittal T1- (a) weighted MR images showing a hypointense epidural venous sac from C-3 to C-6 (arrows) compressing the spinal cord. b Post ligation CT angiogram of the neck demonstrating the tightly narrowed left VA origin and prefistulous dilated tortuous artery. Note the cervical epidural venous sac (arrow). c: Post ligation left subclavian artery angiogram revealing the left VVF connecting the dilated cervical epidural venous sac (black arrows). Note the multiple collaterals from cervical arteries filling the fistula (white arrows). d: Right VA injection demonstrating retrograde filling of left VA (arrowhead) and the fistula (arrow) through intersegmental collaterals. e: Image obtained before coiling of the left subclavian artery, showing the high-flow fistula with a large post fistulous venous sac. Shunting of blood to the epidural venous sac can be seen through the left C5–6 neural foramina (arrows). f: Post coiling left VA injection demonstrating absent antegrade filling of the fistula; however, retrograde filling of the fistula can be seen through the cervical collaterals (arrowheads) because of leakage of contrast material into the subclavian artery. g: Right VA image obtained after coiling, revealing retrograde filling of the left V2 segment (arrowheads) and the fistula through intersegmental collaterals (arrow).h: Right VA image following percutaneous coiling demonstrating no filling of the fistula. Note the percutaneously placed cannula (white arrow) with microcatheter (black arrow) in the coil mass.

Discussion

DISCUSSION

Vascular malformations of the spinal cord occur infrequently, yet are a significant cause of morbidity. They form a complex set of disorders that can present with a wide spectrum of signs and symptoms, which may contribute to delays in their diagnosis and treatment. Spinal vascular malformations can be further classified according to multiple features, including anatomical location, angioarchitecture, or flow dynamics.

There are two major types of spinal vascular malformations based on the location of the abnormal vasculature, intradural and dural. Dural type i.e. spinal DAVFs are the most commonly occurring spinal vascular malformations, accounting for 60–80% of all such lesions[32,35]. The nidus of dural AV fistulas is embedded in the dura covering the dorsal nerve root and in the adjacent spinal dura[21, 22,23,34,35]. Arteriography demonstrates an AV nidus of fine vessels in the lateral aspect of the spinal canal and within an intervertebral foramen. Contrast medium flows through the fistula and intradurally, into a tightly coiled, continuous vessel on the cord surface in to the medullary vein in a retrograde manner to the coronal venous plexus, which becomes dilated, tortuous, and elongated[30].

In our series we found 26 cases of spinal dural AV fistula constituting 33% of all the cases of spinal vascular lesions excluding cavernomas. Incidence of spinal dural fistula is much higher as compared to the other vascular lesions. However one possible reason for this descrepency in our study is lack of catastrophic acute events in the natural history of SDAVF and clinical features resembling other common causes of neurological deficits and backache like disc disease or neuromuscular disease[34]. Another important reason may be initial imaging evaluation by CT scan, low field MR and sub-optimal MR imaging. So large number these patients remain undiagnosed or diagnosis is very late leading to under-reporting of this entity. This is evident in our series where 4 patients had undergone previous surgery with suspected diagnosis of other pathology. Similar discrepancy was noted by Rosenblum et al[35] who attributed this to the referral bias and practice of dorsal vein stripping on laminectomy.

There was strong male predominance reported by Rosenblum et al[35] in there series of 27 patients of SDAVF. Men comprised 85% of patients of SDAVF. The age of patients with symptoms from dural AV fistulas ranged from 22 to72 years (Mean age +/- 49+/-11.4 years). In the series of Song et al[88] there were 20 patients (16 [80%] men and four [20%] women aged 47–83 years [mean, 66 years]). Eskander et al[103] reported 26 patients with spinal dural AVFs which included 22 men and 4 women with a mean age of 65 years (range 39–79 years).

In our series we found total 26 patients (24 males; 2 female) of SDAVF with mean age 52 yrs (Age range 29-81 yrs) which is similar to the previously reported literature. Median time from onset of clinical disease to the diagnosis ranges between 12-44 months[34]. In our study mean duration of symptoms was 18 months. In the study of Jellema at al[67] initial symptoms included walking difficulty in 34%, sensory impairment in 24%, backache in 14% and micturition problems in 4%. Song et al[88] found weakness in 75%, sensory deficit in 70%, backache 25% and urinary complaints in 5% patients. In our cases, extremity weakness (10 patients, 38%), sensory symptoms (4 patients, 15%), back pain (10 patients, 38%), and urinary problems (3 patients, 11%) constituted early complaints. By the time diagnosis is made symptoms are more severe as well as present in higher proportion of cases. In the study of Jellema et al[67] paraparesis and urinary complaints were present in ~80%, backache in 28% and erectile dysfunction in 25% patients at the time of diagnosis. Song et al[88] reported paraparesis in 90%, urinary complaints in 85%, backache30% and sexual dysfunction in 15% patients at the time of diagnosis. We too noted similar progression of symptoms in our patients and found weakness in 25 patients (96%), urinary symptoms 25 patients (96%), sensory symptoms 23 patients (92%), paresthesias 8 patients (31%) and erectile dysfunction 4 patients (15%) at the time of diagnosis. Symptoms were progressive either continuously (24 patients) or fluctuating in 1 patient while there was acute onset illness in one patient. No patient had hemorrhagic event. Hemorrhage is uncommon in SDAVF and seen in cervical lesion where venous congestion can progress in to the posterior fossa[34,35,88,104].

'Flow void phenomena' representing tortuous and dilated veins at the dorsal surface of the spinal cord are found in 35–91% of patients undergoing MRI[105,106]. We were able to identify flow voids in 88% patients. Initial MRI did not show flow voids in 5 patients, however repeat MRI with 3D T2W TSE showed this in 2 more patients. Hyperintensity on T2W images is the hallmark of this condition and is seen in 67%-100% cases and we noted this abnormal signal change in all our patients who underwent MRI examination.

In two patients myelography was done which showed linear filling defects in subarachnoid contrast column. MR angiography may be useful in localizing the level of fistula, however exact level cannot always be made out and may give false positive result also. We could identify the level of fistula in two patients with MR angiography which was done in 5 patients.

Also in accordance with previous studies, most SDAVFs arose from lower thoracic and lumbar vertebral segmental arteries. Fistulae at the cervical level are uncommon. Recently Narvid et al[104] reported 3 cases of cervical dural fistulas in there series of 63 patients. We did not find any case of cervical dural fistula in our patients. However we found one case of sacral fistula.

In 25 cases diagnosis was confirmed by spinal angiography. Therapeutic embolization was done using NBCA in all the treated cases. In no case primary surgery was done. Results of embolization in our cohort are comparable to the some of the largest series reported till date.

Table 8: Comparison of the endovascular results for SDAVF

Authors & Year	No. of Patients	Embolization Technique	Successful Embolization (%)	Surgery	Repeat embolization
Guillevin et al[107], 2005	26	NBCA	81	19	0
Rodesch et al., 2005	18	NBCA	72	17	0
Song et al., 2001	20	NBCA	70	25	0
Van Dijk et al., 2002	44	NBCA	25	70	0
Narvid et al., 2008	39	NBCA	69	31	0
Niimi et al., 1997	49	NBCA /IBCA & Embospheres	83	0	N/A
Present study	26 (2 spontaneous cure)	NBCA	87%	8%	1

The difficulties in achieving definitive control of these lesions were shown in the study by Niimi, et al[66]. In their experience with 49 patients, adequate initial embolization occurred in only 39 (80%) of 49 cases. After adequate embolization was achieved, an additional eight patients suffered a recurrence due to collateralization or to the development of a new AVF. Thus overall, only 63% of patients were definitively treated by the initial embolization. In study by Westphal and Koch[37], 47 patients with spinal dural AVF were treated using an interdisciplinary approach combining embolization and surgery. The authors concluded that embolization should be attempted as the first mode of treatment, although only 30% of patients were cured by embolization alone and the remaining 70% required surgery. In our study 24 patients underwent primary therapeutic procedure, in 83% complete exclusion of fistula was achieved. In 2 patients (9%) complete exclusion of fistula could not be achieved so they were referred for surgery while in another 2 (9%) recanalization occurred for which repeat successful embolization was done in one case while other was lost in follow up. No major permanent complication occurred in our study. Overall in 21 patients fistula was successfully cured.

Mean follow up duration was 19 months in our study. Comparison of admission Modified Aminoff Logue score with the post embolization follow up score revealed complete anatomical and clinical cure in 2 patients (~9%, ALS Gr 0/0/0), improvement was noted in 15 patients (57%), unchanged status in the remaining. No procedure related worsening seen.

Westphal and Koch[37] found that there 42% patients improved following treatment. Cenzato et al[108] reported 46% improvement while Song et al[88]. Found that 57% improved following treatment. All studies have reported improvement in the gait and weakness, however micturition disturbances were considered relatively resistant to treatment. However Narvid et al[104] reported improvement in 65% of endovascular patients and 50% of surgery patients by at least one ALS point, and for micturition, 94 and 80%, respectively. There data add evidence to support the equivalence of endovascular and surgical treatment in achieving improvements in ALS grade. There data do not support the idea that deficits in bowel and bladder function resist therapeutic improvement as reported earlier. If we use the criteria used in Narvid`s study; Gait score improved by at least 1 grade in 13 (54%) patients and micturition score improved in 11 patients (45%). Our experience supports the there inference regarding improvement in sphincter disturbances. However one drawback of our study is lack of long term follow up in all the cases as only 6 patients had follow up more than 3 yrs.

PERIMEDULLARY FISTULA:

Intradural AVM's are spinal vascular lesions in which the nidus of the AVM is in the cord or pia and which, together with the spinal cord, receive their blood supply from medullary arteries. lntradural AVM's are subclassified into intramedullary AVM's, in which the nidus of the AVM is within the parenchyma of the spinal cord or pia, and direct AV fistulas, which are intramedullary or paramedullary in location, and in which the transition from artery to vein occurs without an intervening glomus of abnormal vessels. So perimedullary AVFs, or Type IV lesions involve an aberrant connection between the spinal arteries and veins without an intervening vascular network. Typically they are located near the conus medullaris in the ventral subarachnoid space and extend over multiple spinal levels. Given the significant flow present in these structures, aneurysms and ectatic venous malformation are frequent accompaniments.[1,3,4,22,23,29,31,42,69,70]

Perimedullary fistula constituted 17% of the cases of spinal vascular malformation in the series of Mourier et al[42]. They reported 35 cases with mean age of 25yrs (2-42 yrs). There was no sex predominance. There were 4 patients of type IVa, 9 patients of Type IVb and 22 patients of Type IVc. Cho et al[45] in there series reported 19 patients with mean age of 28 yrs (6-52 yrs). There were 12 males and 7 females. The mean duration of symptoms was 22 months. There were 9 Type IVa, 6 Type IVb and 4 Type IVc. In the series of Rodesch et al[70] there was clear male predominance with 10 pediatric patients (<15 yrs) and remaining adults. They classified lesions as micro AVF and macro AVF with 7 cases of macro AVF and remaining being micro AVF.

In our study there were total of 17 cases (22%) of perimedullary fistula or Type IV lesions (15 males and 2 females). They included 9 patients (52%) of pediatric age group (7 males and 2 females). Mean age is 26 yrs (range, 6-62 yrs). By criteria followed in Rodesch et al`s[70] study there were 7 pediatric patients (41%). There were 10 cases of Type IV a, 5 cases of Type IVb and 2 cases of Type IVc (14 micro-AVF and 3 macro-AVF). 2 cases of macro AVF were seen in pediatric patients, however none showed features of HHT. Cho et al[45] also did not find associated HHT in any of his patients. However HHT is difficult to diagnose in pediatric patients because of absence of telengiectsia and scarcely available history of epistaxis.

In Mouriers series, all Type a and Type b lesions were seen at lumbar level and Type c in thoracic (16) and cervical levels (6)[42]. In Cho et al[45] cohort, all lesions were at conus medullaris level, except two which were located at cervical level and were of Type b and Type c respectively. Barrow et al[40] also found majority of his lesions in conus medullaris level (7 of 8). In the series of Rodesch et al[70] majority of lesions were thoracolumbar in location. In our study group majority of lesions were in conus region. Eleven lesions were seen in relation to the conus, 4 in the dorsal spine, two in relation to filum terminale and one each in cauda equina root and cervical spine. In two patients multiple (two each) lesions were noted.

Clinically, 70% of pediatric and 27% of the adult patients presented with hemorrhagic symptoms in the series of Rodesch et al[70]. In two patients acute non hemorrhagic onset was noted while remaining presented with progressive symptoms. Venous architectural factors (either venous ectasias that ruptured or venous pial reflux) were most often associated with hemorrhage. Venous architectural factors (either venous ectasias that ruptured or venous pial reflux) were most often associated with hemorrhage. Venous architectural factors either venous ectasias or venous pial reflux was most often associated with hemorrhage. In study of Cho et al[45] most common presentation was progressive weakness followed by backache. Acute presentation was noted only in the 3 patients. Patients with hemorrhagic manifestation were not mentioned. In Mourier`s series also majority of patients presented with progressive illness (32 of 35) while hemorrhage was suspected in 7 cases[42]. However in the latter two studies angioarchitecture was not evaluated.

In our cohort eleven patients (64%) had an acute onset of illness; while in the remaining 6 progressive symptoms were noted. In 8 (46%) patients there was clinical or radiological evidence of underlying hemorrhage as the cause of acute symptoms. In 3 cases acute non-hemorrhagic presentation was seen. All the pediatric patients (<15 yrs) had acute onset illness, however in one patient after initial episode patient had progressive course over next 9 yrs. In 4 patients definite history suggestive of hemorrhage was present (57%). In adults incidence of hemorrhage was 40%.

Five patients who presented with acute hemorrhagic event had associated venous pouch/venous ectasias or venous aneurysms. Intrinsic cord venous congestion was seen in 4 of these 8 patients. Venous thrombosis as evaluated on MRI was seen in two patients and both patients had acute presentation. Feeding artery aneurysm were seen in two patients both of whom presented with acute hemorrhagic presentation. However in both these cases venous side abnormalities were also noted. Statistical analysis for association between venous abnormalities and hemorrhage showed definite association between the two, however it failed to reach the level of significance possibly because of less number of patients.

Table 9:Comparison of treatment/and outcome in cases of PMF:

Authors & Year	No. of Patients treated	Embolization Technique	Embolization	Surgery	Combined	Angiographic result	Clinical outcome
Mourier et al, 1993	34	IBCA, Balloon, PVA	25 (22 primary endovascular)	6	8	Cure-27(79%) Partial-6(17%) One patient died details not given	Wor-5% Unc-26% I-69%
Cho et al, 2005	19	Glue, PVA	11	8	2	Cure 5(45%) Partial-4(36%) Failure of pr-2	Wor-11% Unc-17% I-72%%
Rodesch et al, 2005	32	Glue	18	17	-	Cure-10 (66%) Partial-5(34%) Failure of pr-3	I-93% Unc-7%
SCTIMST experience	17	Glue, PVA, Onyx, coils	16	1	-	Cure-13(81%) Partial-3(19%) Surgery in one patient-cured	I-12(72%) Unc-3(17%) Wor-2 (11%) Surgery patient-I

I-Improved, Unc-Unchanged, Wor-Worsened

On clinical follow up improvement was noted in 72% patients; 66% of whom had no disability. Mean follow up duration was 24 months (range 1 month to 8 yrs) and is comparable to that reported in previous studies.

Complications occurred in 5 patients and resulted in fixed permanent deficits in two patients. Rodesch et al[70] reported no major deficit although they kept the possibility of dissection in one of their case and procedure was abandoned. Patient had spontaneous closure of fistula on follow up. Other complications in there series were transient in nature. Mourier et al[42] reported clinical worsening in two of their cases while one patient died following embolization. Cho et al[45] reported only 1 transient worsening following procedure in 11 patients who underwent embolization.

SPINAL ARTERIOVENOUS MALFORMATIONS:

Spinal cord AVM constitutes ~80% of all the spinal intradural arteriovenous shunts; rests are perimedullary fistulas. They usually present early in the life most commonly during the 3rd decade. Rosenblum et al found[35] male predominance with 70% cases of intradural AVM seen in male patients. However Rodesch et al[62] found male dominance only in the pediatric age group; in adults no gender bias was seen. In his study mean age of patients of intradural AVM was 27 yrs. In the patient group of Biondi et al there was male predominance (70%). Mean age of the patients was 20 yrs. However in the cohort of Corkill et al intramedullary AVM cases were more ftequently seen females (12 females and 5 males). In our series of 28 patients there were 16 males and 12 females with mean age of 28 yrs.

Hemorrhage is the most common presentation, occurring in 50% of all patients with spinal cord AVMs. There can also be acute non- hemorrhagic symptoms. Other manifestation include slowly progressive neurological deficit and rarely intracranial SAH[69]. In the series of Biondi et al 58% patients had a hemorrhagic onset. Hemorrhage was the initial manifestation in 70% patients of Corkill et al. Hemorrhage is more often seen in pediatric patients compared to the adults[62]. Rosenblum et al reported hemorrhage in 31% of their patients with motor and sensory deficits being commonest clinical deficits in patients with intradural AVM[35]. We found evidence of hemorrhage in 31% patients in our study. However if we include all the patients of intradural shunt then hemorrhagic manifestation were seen in 17 patients (38%). Other commonest clinical features seen in our study are motor weakness and sensory abnormalities. Sphincter disturbances were noted in 78% and erectile dysfunction in 17% patients.

In Rosenblum`s series intradural AVM occurred more diffusely along the spinal axis with lower thoracic being the commonest site. Multiple feeding arteries were noted in 72% lesions. Associated arterial or venous aneurysms were demonstrated in 24 of the 54 patients. More than 50% patients associated with arterial aneurysms have bled. Similar association between intramedullary AVM and feeding artery aneurysms was reported by Biondi et al. In Rodesch series thoracolumbar was the commonest site seen in 66% cases. There was statistically significant difference in bleed rates of cervical lesions and thoracolumbar lesions with former more likely to bleed. Arterial aneurysms and venous ectasia were not correlating with the bleed rates. However there was high significant association noted between pseudoaneurysms and hemorrhage. Multiple lesions were seen in 16 patients.

We observed multiple vascular lesions in 3 patients involving same as or the adjacent myelomers. In these cases possibility of SAMS was kept. In another case characteristic skin vascular lesion was seen involving same dermatomes as those involved by spinal vascular malformation. In another case large swelling was noted in the relation to the same myelomere and it was treated multiple times by surgery. These two cases may be incompletely expressed SAMS syndrome. Feeding artery aneurysm was seen in 4 cases, however no association was found between feeding artery aneurysm and clinical symptomatology of hemorrhage. Intranidal aneurysm was seen in 7 cases. In five patients harboring intranidal aneurysm hemorrhage was seen. Hugely dilated venous ectatic sac was seen in 3 patients. In 2 of these patients there was acute clinical presentation and hemorrhage was noted. Intracranial venous drainage was noted in 2 patients with one of them presenting with SAH. In two patients pseudoaneurysms were seen and hemorrhage was seen in both the cases. Our results regarding angioarchitecture are in line with the observations of Rodesch et al[62]. We have found weak association between venous pouch/ectasia and clinical event of hemorrhage. This association has not been found significant in some of the previous studies. In view of smaller number of patients in our study, statistical conclusions may not be correct in this regard. Higher bleed rates noted in the cervical lesions in the previous studies may be due to the increased chance of having intracranial venous drainage. However we can definitely conclude from our study that feeding arterial aneurysms are not associated with hemorrhage and presence of pseudoaneurysms has high association with hemorrhage.

Total cure of these lesions is difficult. Partial targeted treatment to obliterate weak portions like arterial or nidal aneurysms, size reduction, decreased flow, and decongestion of the venous drainage. Obliteration of a high-risk portion of the lesion, such as an associated aneurysm, the occlusion of a direct AVF within the nidus, or of a portion draining into a territory with outflow restriction, can result in a favorable long-term outcome[1,3,4,62,69].

Table 10:Comparison of results of embolization of AVM:

Authors & Year	No. of Patients treated	Embolization	Follow up	Angiographic result, (complete obliteration)	Clinical outcome
Da Costa et al, 2009	47	Glue	N/A	49%	I-78% Wor-22%
Rodesch et al 2003	69	Glue	Mean-5.6 yrs	16%	I-58% Unc-25% Wor-13% Failure-4%
Corkill et al, 2007	17	Onyx	Mean-2 yrs	37.5%	I-10(58%) Unc-4(23%) Wor-3(17%)
SCTIMST experience	16	Glue, PVA	Mean 4.2 yrs	20%	I-7(46%) Unc-2(16%) Worsening-6 (39%)

Complications noted in our series included transient worsening in four cases which improved with conservative management and were possibly related to venous slowing following embolization. In 6 patients persistent clinical worsening was seen on follow up which included 5 patients with immediate post procedure worsening and one patient with worsening beginning after 1 month. In one patient embolization was done 14 yrs back and he had minimal disability in this period. Clinical worsening was noted and second sitting of embolization was planned. There was feeder perforation during second sitting of embolization, resulting in subarachnoid hemorrhage. Feeder was immediately embolized with glue. However patient developed severe neurological deficit. In two patients spinal cord abnormality was detected on post procedure MRI. In one, extensive signal changes were noted in the distal spinal cord which possibly occurred due to the occlusion of anterior spinal axis, while in the second case there was lesion noted in the territory unrelated to the primary site of abnormality. Cervical and posterior circulation infarct occurred, while embolized lesion happened to be in the mid dorsal spine. So possibility of thromboembolism occurring in the vertebral artery was considered. So in this case complication was unrelated to the primary embolization of the AVM. In remaining two patients, exact cause of clinical worsening could not be found. In one of the patients MRI after clinical deterioration did not show any new finding while in the other patient no follow up imaging was done. Possibility of venous slowing with thrombosis post embolization may be one of the causes contributing to deteriorating clinical status. One patient who was clinically unchanged after procedure developed gradual worsening during follow up showed enhancing intradural extramedullary lesion at the level of vascular lesion. Surgical resection was done and histology showed inflammatory granuloma of uncertain etiology.

Table 11: Disability in cases of clinical worsening following embolization

S.No	Pre-procedure disability	Post-procedure disability	Embolization material	Arterial axis embolized
1.	Gr 2/1/0	Gr 5/2/1	Glue 15%, PVA	ASA-glue, PSA-PVA
2.	Gr0/0/0	Gr4/2/0	Glue 20%	PSA
3.	Gr2/1/0	Gr5/3/0	PVA	ASA/PSA
4.	Gr2/1/0	Gr4/1/0	PVA	ASA
5.	Gr1/0/0	Gr4/0/0	PVA	ASA
6.	Gr2/0/1	Gr5/1/1	Glue/PVA	ASA

PARASPINAL AND VERTEBRAL ARTERIOVENOUS MALFORMATIONS:

One case of purely paraspinal AVM was seen in our study who has presented with features of myelopathy. Lesion was in paraspinal location and was showing bony destruction on CT. Myelography showed complete extradural block at D7 level. Patient underwent multiple sessions of embolization using glue and particles over a period of 6 yrs and disease was well controlled until he developed features of congestive heart failure. Patient was operated up on but died following surgery due to cardiac arrest. Because of rarity of these lesions no generalized management approach has been suggested and Goyal et al[51] has advocated a individualized case based approach. In authors series thoracic was the commonest site. Similarly, Kitagawa et al[109] reviewed all the cases of paraspinal AVM in pediatric age and found thoracic as the commonest site. These patients commonly present with myelopathic features. Congestive heart failure was seen in one of the patients of Hui et al[59]. Underlying pathophysiology is direct mass effect or congestive myelopathy due to the reflux in to perimedullary veins. In cases where reflux is present surgical interruption of the radicular vein is found useful. Embolization can cure the lesions in significant number of patients and control the symptoms in others. All types of embolic material have been used for treatment in these malformations including PVA, liquid agents and coils. In our case we were not able to cure the lesion however disease manifestations could be well controlled for 6 yrs till patient died due to CHF.

Vertebral arteriovenous malformations are a distinct entity; they can be acquired following trauma or can be spontaneous. Spontaneous can be seen in association with neurofibromatosis and fibromuscular dysplasia[52]. They are usually seen above C2 level or below C5 level and are commonly fed by vertebral artery. These lesions most commonly present with features of myelopathy. Upper cervical lesions can present with tinnitus or vertigo. Endovascular method is the treatment of choice for these lesions. Two patients in our series presented with myelopathic features and were both located in lower cervical spine with feeders from vertebral artery in both cases. In addition multiple feeders from the numerous muscular branches were seen in the other patient who initially underwent surgery. Following failure of surgery patient was sent for embolization. Possible cause for symptoms was dilated epidural veins causing compression of spinal cord. In view of very high flow and underlying neurofibromatosis, coils were used for closing the fistula. Following successful closure of fistula, patient recovered well. Second patient in our series is awaiting procedure.

Conclusions

CONCLUSIONS

Spinal vascular malformations are rare lesions with spinal AVM and spinal dural arteriovenous malformation being the commonest one. Spinal dural arteriovenous malformations usually present with slowly progressive neurological deterioration while spinal AVM and perimedullary fistulas can have acute onset illness associated with spinal hemorrhage.

For initial evaluation, MRI is the investigation of choice followed by spinal angiography for identification of the type of malformation, feeders and angioarchitecture. Endovascular treatment is the first line of treatment, however if there is failure of endovascular treatment surgery may be the alternative. The goal of endovascular management should be obliteration of the fistula in cases of SDAVF and PMF with part of the liquid embolic agent percolating to venous side. Liquid embolic agent are the embolic agent of choice in these cases. In cases of AVM aim should be palliative i.e. to exclude high risk elements in a AVM nidus like intranidal aneurysm, pseudoaneurysm and venous ectasia/pouch, PVA should be used if there is high risk with the use of liquid embolic agents especially in the anterior spinal axis. For paraspinal and vertebral arteriovenous fistula treatment needs to be individualized.

Given the present day expertise, technology, machines and available materials endovascular management of spinal dural arteriovenous fistulas and perimedullary fistulas have extremely good results. In cases of AVM and epidural lesions treatment needs to be tailored as per the needs of the patient.

The mortality or morbidity associated with endovascular management is very low in the present day. Previously encountered morbidities were due to inexperience and technical limitations.

A detailed clinical evaluation, imaging work up, angiographic analysis and timely follow up for all patients with spinal vascular malformations is recommended.

References

REFERENCES

1. Krings T, Lasjaunias PL, Hans FJ, Mull M, Nijenhuis RJ, Alvarez H et al. Imaging in Spinal Vascular Disease Neuro imag Clin N Am 2007;17:57–72

2. Chaloupka JC: Future directions in the evaluation and management of spinal cord vascular malformations. Semin Cerebrovasc Dis Stroke 2002; 2:245–256.

3. Rodesch G, P. Lasjaunias. Spinal cord arteriovenous shunts: from imaging to management. European Journal of Radiology 2003;46:221-232.

4. Berenstein A, Lasjaunias P, ter Brugge K: Spinal cord arteriovenous malformations, in Berenstein A, Lasjaunias P, ter Brugge K (eds): Surgical Neuroangiography, Vol 2.2. New York: Springer-Verlag 2003, pp 737–843

5. Hebold O: Aneurysmen der kleisnten rucken marksgefasse. Arch Psychiatr Nervnkr 1885;16:813-823.

6. Gaupp J: Hamorrhoiden der pia matter spinalis im gebiet des lendenmarks. Beith Pathol 1888; 2:516-518.

7. Berenbruch K: Ein fall von multiplen Angio-Lipomen Lombiniert mit einem angiom des Rückenmarks [thesis]. Tubingin, Germany: Universität Tubingen, 1890. Ref Type: thesis/Dissertation.

8. Sargent P: Haemangioma of pia mater causing compression paraplegia. Brain 1925;48:259–267.

9. Elsberg CA: Diagnosis and Treatment of Surgical Diseases of the Spinal Cord and Its Membranes. Philadelphia: WB Saunders, 1916, pp 194–204

10. Foix C, Alajouanine T: La my61ite ndcrotique subaigue. Rev Neurol (Paris) 1926;33:1-42.

11. Lhermitte L, Fridbourg-Blanc A, Kyriaco N: La gliose ang6io-hypertrophique de la moelle 6pinidre (my61ite n6crotique de Foix-Alajouanine). Rev Neurol 1931; 2:37-53.

12. Wyburn-Mason R: The Vascular Abnormalities and Tumors of the Spinal Cord and Its Membranes. London, Saint Louis:CV Mosby, 1944

13. Perthes G: Uber des Rankenangiom der weichen haute des gehims and ruckenmarks. Dtsch Z Chir 1927;203/204:93-103.

14. Hook D, Lidvall H. Arteriovenous aneurysms of the spinal cord: A report of two cases investigated by vertebral angiography. J Neurosurg 1958;15:84-91.

15. Djindjian R, Faure C: Investigations neuro-radiologiques dans les maltbrmations vasculaires medullaraires. Neuroradiol Roentgenol Eur 1963;4:432-434.

16. Doppman J, DiChiro G: Subtraction angiography of spinal cord malformations. J Neurosurg 1965;23:440-443.

17. Lasjaunias P, Berenstein A. Surgical Neuroangiography. 3. Functional Vascular Anatomy of Brain, Spinal Cord and Spine. Springer-Verlag, Berlin, 1990.

18. Tatu L, Vuillier F, Moulin AT. Anatomy of the circulation of the brain and spinal cord Stroke,in Fisher M(ed). Handbook of Clinical Neurology, 2009;Vol. 92 (3rd series), Part I: pp247-281

19. Di Chiro G, Doppman JL, Ommaya AK: Radiology of spinal cord arteriovenous malformations. Progr Neurol Surg 1971;4:329–354.

20. Djindjian M, Djindjian R, Rey A, Hurth M, Houdart R. Intradural extrameduallary spinal arterio-venous malformations fed by the anterior spinal artery. Surg Neurol 1977; 8: 85-93.

21. Merland JJ, Riche MC, Chiras J: Intratraspinal extramedullary arteriovenous fistula draining into medullary veins. J Neuroradiol 1980;7:271-320.

22. Guegen B, Merland JJ, Riche MC, et al: Vascular malformations of the spinal cord. Intrathecal perimedullary arteriovenous fistulas fed by medullary arteries. Neurology 1987;37:969-979.

23. Heros RC, Debrun GM, Ojemann RG, Lasjaunius PJ, Naessens PJ. Direct spinal arteriovenous fistula: A new type of spinal AVM. J Neurosurg 1986; 63:134-139,

24. Aminoff MJ, Logue V: Clinical features of spinal vascular malformations. Brain 1974;97:197–210.

25. Kendall BE, Logue V: Spinal epidural angiomatous malformations draining into intrathecal veins. Neuroradiology 1977;13:181-189.

26. Oldfield EH, Di Chiro G, Quindlen EA, Rieth KG, Doppman JL: Successful treatment of a group of arteriovenous malformations by interruption of dural fistula. J Neurosurg 1983; 59:1019–1030.

27. Symon L, Kuyama H, Kendall B: Dural arteriovenous malformations of the spine. Clinical features and surgical results in 55 cases. J Neurosurg 1984; 60:238–247.

28. Anson JA, Spetzler RF: Classification of spinal arteriovenous malformations and implications for treatment. BNI Q 1992;8:2–8.

29. Spetzler RF, Detwiler PW, Riina HA, Porter RW: Modified classification of spinal cord vascular lesions. J Neurosurg 2002; 96:145–156.

30. Rodesch G, Hurthe M, Alvarez H, Tadie M, Lasjaunias P. Classification of spinal cord arteriovenous shunts: proposal for a reappraisal—the bicêtre experience with 155 consecutive patients treated between 1981 and 1999. Neurosurgery 2002; 51:374-380.

31. Krings T, Mull M, Gilsbach JM, Thron A. Spinal vascular malformations. Eur Radiol 2005; 15:267–78.

32. Jellema K, Tijssen CC, Sluzewski M, van Asbeck FW, Koudstaal PJ, van Gijn J: Spinal dural arteriovenous fistulas–an underdiagnosed disease. A review of patients admitted to the spinal unit of a rehabilitation center. J Neurol 2006; 253:159–162.

33. Grandin C, Duprez T, Stroobandt G, Laterre EC, Mathurin P. Spinal dural arterio-venous fistula: an underdiagnosed disease? Acta Neurol Belg 1997; 97: 17–21.

34. Jellema K, Tijssen CC, van Gijn J Spinal dural arteriovenous fistulas: a congestive myelopathy that initially mimics a peripheral nerve disorder Brain 2006; 129(Pt 12):3150-64.

35. Rosenblum B, Oldfield EH, Doppman JL, Di Chiro G. Spinal arteriovenous malformations: a comparison of dural arteriovenous fistulas and intradural AVM's in 81 patients. J Neurosurg 1987; 67: 795–802.

36. Bedersen JB, Spetzler RF. Pathophysiology of Type I spinal dural arteriovenous malformations. BNI Quarterly 1996; 12: 23–32.

37. Westphal M, Koch C. Management of spinal dural arteriovenous fistulae using an interdisciplinary neuroradiological/neurosurgical approach: experience with 47 cases. Neurosurgery 1999; 45: 451–457.

38. Van Dijk JM, TerBrugge KG, Willinsky RA, Farb RI, Wallace MC. Multidisciplinary management of spinal dural arteriovenous fistulas: clinical presentation and long-term follow-up in 49 patients. Stroke 2002; 33: 1578–1583.

39. Yaşargil MG, Symon L, Teddy PJ: Arteriovenous malformations of the spinal cord. Adv Tech Stand Neurosurg 1984; 11:61– 102.

40. Barrow DL, Colohan ART, Dawson R. Intradural perimedullary arteriovenous fistulas (type IV spinal cord arteriovenous malformations). J Neurosurg 1994; 81:221–229

41. Halbach VV, Higashida RT, Dowd CF, Fraser KW, Edwards MS, Barnwell SL () Treatment of giant intradural (perimedullary) arteriovenous fistulas. Neurosurgery 1993; 33:972–979.

42. Mourier KL, Gobin YP, George B, Lot G, Merland JJ. Intradural perimedullary arteriovenous fistulae: results of surgical and endovascular treatment of 35 cases. Neurosurgery1993; 32:885–891.

43. Riche MC, Scialfa G, Gueguen B, Merland JJ Giant extramedullary arteriovenous fistula supplied by the anterior spinal artery: treatment by detachable balloons. AJNR Am J Neuroradiol 1983; 4:391–394.

44. Rodesch G, Pongpech S, Alvarez H, Zerah M, Hurth M, Sebire G, et al. Spinal cord arteriovenous malformations in a pediatric population. Intervent Neuroradiol 1995; 1:29–42.

45. Cho KT Lee DY, Chung CK, Han MH, Kim HJ. Treatment of spinal cord perimedullary arteriovenous fistula: embolization versus surgery. Neurosurgery 2005; 56:232-241.

46. Berenstein A, Lasjaunias P. Spinal cord arteriovenous malformations. In: Berenstein A, Lasjaunias P, editors. Surgical neurorangiography: endovascular treatment of spine and spinal cord lesions, vol. 5. New York: Springer Verlag, 1992:24-76.

47. Sala F, Nimii Y, Krzan MJ, Berenstein A, Deletis V. Embolization of spinal cord arteriovenous malformations: correlation between motor evoked potentials and angiographic findings: technical case report. Neurosurgery 1999; 45:932-938.

48. Bandhyopadhyay S, Sheth RD. Acute spinal cord infarction: vascular steal in arteriovenous malformation. J Child Neurol 1999; 14:685-687.

49. Lasjaunias P. Parachordal arteriovenous fistulas (extracranial and extraspinal arteriovenous fistulas). In: Lasjaunias P, Terbrugge K, editors. Vascular diseases in neonates, infants and children. Interventional neuroradiology management. Berlin/Heidelberg/ New York: Springer Verlag 1997:501-530.

50. Rodesch G, Alvarez H, Chaskis C, Peeters J, Lasjaunias P. Clinical manifestations in paraspinal arteriovenous malformations: spinal cord symptoms, pathophysiology and treatment objectives. Int J Neuroradiol 1996;2:430-436.

51. Goyal M, Willinsky R, Montanera W, terBrugge K. Paravertebral Arteriovenous Malformations with Epidural Drainage: Clinical Spectrum, Imaging Features, and Results of Treatment. AJNR Am J Neuroradiol 1999; 20:749–755.

52. Beaujeux RL, Reizine DC, Casasco A, Aymard A, Rufenacht D, Khayata MH et al. Endovascular treatment of Vertebral arteriovenous fistulas. Radiology 1992; 183:362-367.

53. Krayenbuhl H, Yasargil MG. Die varicosis spinalisund ihreBehandlung. Schweizer Arch Neurol Psychiat 1963; 92: 74-92.

54. Pia HW, Vogelsang H. Diagnose und Therapie spinaler Angiome. Dt Z NervHeilk 1965; 187: 38-41.

55. Odom GL. Vascular lesions of the spinal cord: malformations, spinal subarachnoid, and extradural hemorrhage. Clin Neurosurg 1962; 8:196-236.

56. Logue V. Angiomas of the spinal cord:review of the of the pathogenesis, clinical features, and results of the surgery. J Neurol Neurosurg Psychiatrry 1979; 42:1-11.

57. Badejo L, Sengalang VE. Vascular malformations of the spinal cord: A critique of its pathogenesis. Surg Neurol 1979;11:101-106.

58. Aminoff MJ. Spinal angiomas. Blackwell, Oxford, 1976

59. Hui F, Trosselo MP, Meisel HJ, Alvarez H, Sequeira E, Lasjaunias P: Paraspinal arteriovenous shunts in children. Neuroradiology 1994; 36:69–73.

60. Hurst RW, Kenyon LC, Lavi E, Raps EC, Marcotte P. Spinal dural arteriovenous fistula: the pathology of venous hypertensive myelopathy. Neurology 1995; 45: 1309–13.

61. Asakawa H, Yanaka K, Fujita K, Marushima A, Anno I, Nose T. Intracranial dural arteriovenous fistula showing diffuse MR enhancement of the spinal cord: case report and review of the literature. Surg Neurol 2002; 58: 251–257.

62. Rodesch G, Hurth M, Alvarez H, Ducot B, Tadie M, Lasjaunias P: Angio-architecture of spinal cord arteriovenous shunts at presentation. Clinical correlations in adults and children. The Bicetre experience on 155 consecutive patients seen between 1981-1999. Acta Neurochir (Wien) 2004; 146:217–226.

63. Hurth M, Houdart R, Djindjina R, Rey A, Djindjian M. Arteriovenous malformations of the spinal cord. Clinical, anatomical and therapeutic considerations: a series of 150 cases. Progr Neurol Surg 1978; 9:238 - 266.

64. Aminoff MJ, Logue V. The prognosis of patients with spinal vascular malformations. Brain 1974; 97:211-218.

65. Kataoke H, Miyamoto S, Nagata I, Ueba T, Hashimoto N. Venous Congestion Is a Major Cause of Neurological Deterioration in Spinal Arteriovenous Malformations. Neurosurgery 2001; 48:1224–1230.

66. Niimi Y, Berenstein A, Setton A, Neophytides A: Embolization of spinal dural arteriovenous fistulae: results and followup. Neurosurgery 1997; 40:675–683.

67. Jellema K, Canta LR, Tijssen CC, van Rooij WJ, Koudstaal PJ, van Gijn J. Spinal dural arteriovenous fistulas: clinical features in 80 patients. J Neurol Neurosurg Psychiatry 2003; 74: 1438–1440.

68. Koch C, Kucinski T, Eckert B, Rother J, Zeumer H. Spinal dural arteriovenous fistula: clinical and radiological findings in 54 patients. Rofo 2003; 175: 1071–1078.

69. Leodante da Costa, Dehdashti AR, terBrugge K. Spinal cord vascular shunts: spinal cord vascular malformations and dural arteriovenous fistulas. Neurosurg Focus 2009; 26 (1):E6.

70. Rodesch G, Hurth M, Alvarez H, Ducot B, Tadie M, Lasjaunias P. Spinal cord intradural arteriovenous fistulae: anatomic, clinical, and therapeutic considerations in a series of 32 consecutive patients seen between 1981 and 2000 with emphasis on endovascular therapy. Neurosurgery 2005; 57:973-983.

71. Roccatagliata L, Centanaro F, Castellan L. Venous congestive myelopathy in spinal dural arteriovenous fistula mimicking neoplasia. Neurol Sci 2007; 28:212–215.

72. Bostroem A, Thron A, Hans FJ, Krings T. Spinal vascular malformations–typical and atypical findings. Zentralbl Neurochir 2007; 68:205–213.

73. Eddleman CS, Jeong H, Cashen TA, Walker M, Bendok BR, Batjer HH et al. Advanced noninvasive imaging of spinal vascular malformations. Neurosurg Focus 2009; 26 (1):E9.

74. Si-jia G, Meng-wei Z, Xi-ping L, Yu-shen Z, Jing-hong L, Zhong-hui W , et al. The clinical application studies of CT spinal angiography with 64-detector row spiral CT in diagnosing spinal vascular malformations. Eur J Radiol. 2009; 71(1):22-28.

75. Lai P, Pan H, Yang C: Multi-detector row computed tomography angiography in diagnosing spinal dural arteriovenous fistula: initial experience. Stroke 2005; 36:1562–1564.

76. Lai P, Weng M, Lee K: Multidetector CT angiography in diagnosing type I and type IVA spinal vascular malformations. AJNR Am J Neuroradiol 2006; **27:**813–817.

77. Mull M, Nijenhuis R, Backes W, Krings T, Wilmink JT, Thron A: Value and limitations of contrast-enhanced MR angiography in spinal arteriovenous malformations and dural arteriovenous fistulas. AJNR Am J Neuroradiol 2007; 28:1249–1258.

78. Ali S, Cashen T, Carroll T, McComb E, Musaffar M, Shaibani A, et al: Time-resolved spinal MR angiography: initial clinical experience in the evaluation of spinal arteriovenous shunts. AJNR Am J Neuroradiol 2007; 28:1806–1810.

79. Lasjaunias P, Maillot C, Terbrugge K. Pial relations with spinal cord veins explain MRI occult spinal AV shunts. Interv Neuroradiol 2000; 6:333-336.

80. Djindjian R: Arteriography of the spinal cord. AJR Am J Roentgenol 1969; 107:461–478.

81. Doppman JL, Krudy AG, Miller DL, Oldfield E, Di Chiro G. Intraarterial digital subtraction angiography of spinal arteriovenous malformations. AJNR Am J Neuroradiol 1983; 4:1081–1085.

82. Doppman JL, DiChiro G, Ommaya G: Obliteration of spinal cord arteriovenous malformations by percutaneous embolization. Lancet 1968; 1:477 (Letter).

83. Hall WA, Oldfield EH, Doppman JL: Recanalization of spinal arteriovenous malformations following embolization. J Neurosurg 1989; 70:714–720.

84. Chaloupka JC, Huddle DC, Alderman J, et al: A reexamination of the augiotoxicity of superselective injection of DMSO in the swine rete embolization model. AJNR Am J Neuroradiol 1999; 20:401-410.

85. Molyneux AJ, Coley SC: Embolization of spinal cord arteriovenous malformations with an ethylene vinyl alcohol copolymer dissolved in dimethyl sulfoxide (Onyx liquid embolic system). Report of two cases. J Neurosurg 2000; 93:304-308.

86. Chaloupka JC, Vinuela F, Vinters HV, Robert J. Technical feasibility and histopathologic studies of ethylene vinyl copolymer (EVAL) using the swine endovascular embolization model. AJNR Am J Neuroradiol 1994; 15:1107-1115.

87. Morgan MK, Marsh WR: Management of spinal dural arteriovenous malformations. J Neurosurg 1989; 70:832–836.

88. Song JK, Gobin YP, Duckwiler GR, Murayama Y, Frazee JG, Martin NA, et al. N-butyl 2-cyanoacrylate embolization of spinal dural arteriovenous fistulae. AJNR Am J Neuroradiol 2000; 22:40–47.

89. Steinmetz MP, Chow MM, Krishnaney AA, Andrews-Hinders D, Benzel EC, Masaryk TJ, et al. Outcome after the treatment of spinal dural arteriovenous fistulae: a contemporary single institution series and meta-analysis. Neurosurgery 2004; 55:77–78.

90. Nogueira RG, Dabus G, Rabinov JD, Ogilvy CS, Hirsch JA, Pryor JC. Onyx embolization for the treatment of spinal dural arteriovenous fistulae: initial experience with long-term follow-up. Technical case report. Neurosurgery. 2009 Jan;64(1):E197-8

91. American Society for Interventional and Therapeutic Neuroradiology. Embolization of spinal arteriovenous fistulae: spinal arteriovenous malformations, and tumors of the spinal axis. AJNR Am J Neuroradiol 2001; 22:28–30.

92. Jellema K, Sluzewski M, van Rooij WJ, Tijssen CC, Beute GN. Embolization of spinal dural arteriovenous fistulas: importance of occlusion of the draining vein. J Neurosurg Spine 2005; 2(5):580-583.

93. McCutcheon IE, Doppman JL, Oldfield EH. Microvascular anatomy of dural arteriovenous abnormalities of the spine: a microangiographic study. J Neurosurg 1996; 84: 215–220.

94. Huffmann BC, Gilsbach JM, Thron A. Spinal dural arteriovenous fistulas: a plea for neurosurgical treatment. Acta Neurochir (Wien) 1995; 135:44–51.

95. Lee TT, Gromelski EB, Bowen BC, Green BA. Diagnostic and surgical management of spinal dural arteriovenous fistulas. Neurosurgery 1998; 43:242–246.

96. Medel R, Crowley RW, Dumont AS. Endovascular management of spinal vascular malformations: history and literature review. Neurosurg Focus 2009 Jan;26(1):E7.

97. Biondi A, Merland JJ, Hodes JE, Aymard A, Reizine D.: Aneurysms of spinal arteries associated with intramedullary arteriovenous malformations. II. Results of AVM endovascular treatment and hemodynamic considerations. AJNR Am J Neuoradiol 1992; 13:923-93I.

98. Rodesch G, Hurth M, Alvarez H, David P, Tadie M, Lasjaunias PL. Embolization of spinal cord arteriovenous shunts: morphological and clinical follow-up and results – review of 69 consecutive cases. Neurosurgery 2003; 53:40–49.

99. Corkill RA, Mitsos AP, Molyneux AJ. Embolization of spinal intramedullary arteriovenous malformations using the liquid embolic agent, Onyx: a single-center experience in a series of 17 patients. J Neurosurg Spine 2007; **7**:478–485.

100. Atkinson JL, Piepgras DL. Surgical treatment of spinal cord arteriovenous malformations and arteriovenous fistulas. Seminars in Cerebrovascular Diseases and Stroke 2002; 2(3):201-208.

101. Rodesch G, Hurth M, Alvarez H, Tadie M, Lasjaunias P. Spinal cord intradural arteriovenous fistulae: anatomic, clinical, and therapeutic considerations in a series of 32 consecutive patients seen between 1981 and 2000 with emphasis on endovascular therapy. Neurosurgery 2005; 57:973–983.

102. Oran I Parildar M, Derbent A. Treatment of slow-flow (type I) perimedullary spinal arteriovenous fistulas with special reference to embolization. AJNR Am J Neuoradiol 2005;26(10):2582-2586.

103. Eskandar EN, Borges LF, Budzik RFJ, Putman CM, Ogilvy CS. Spinal dural arteriovenous fistulas: experience with endovascular and surgical therapy. J Neurosurg 2002; 96: 162–167.

104. Narvid J, Hetts SW, Larsen D, Neuhaus J, Singh TP, McSwain, et al: Spinal dural arteriovenous fistulae: clinical features and long-term results. Neurosurgery 2008; 62:159–167.

105. Hurst RW, Grossman RI. Peripheral spinal cord hypointensity on T2-weighted MR images: a reliable imaging sign of venous hypertensive myelopathy. AJNR Am J Neuroradiol 2000; 21: 781–786.

106. Gilbertson JR, Miller GM, Goldman MS, Marsh WR. Spinal dural arteriovenous fistulas: MR and myelographic findings. AJNR Am J Neuroradiol 1995; 16: 2049–2057.

107. Guillevin R, Vallee JN, Cormier E, Lo D, Dormont D, Chiras J. N-butyl 2-cyanoacrylate embolization of spinal dural arteriovenous fistulae: CT evaluation, technical features, and outcome prognosis in 26 cases. AJNR Am J Neuroradiol 2005; 26: 929–35.

108. Cenzato M, Versari P, Righi C, Simionato F, Casali C, Giovanelli M. Spinal dural arteriovenous fistulae: analysis of outcome in relation to pretreatment indicators. Neurosurgery 2004; 55: 815–22.

109. Kitagawa RS, Mawad ME, Whitehead WE, Curry DJ, Luersen TG, Jea A. Paraspinal arteriovenous malformations in children. J Neurosurg Pediatr. 2009 May;3(5):425-428.