Imaging and Endovascular management of Cervicofacial vascular malformations

Thesis submitted in fulfilment of the rules and regulations for DM Degree Examination of Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram

By

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Resident in Neuroradiology

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Under the guidance of

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DECLARATION

I hereby declare that the dissertation titled **"Imaging and Endovascular management of Cervicofacial vascular malformations**" has been prepared by me under guidance **of Dr. A K Gupta,** Professor, Department of Imaging Sciences and Interventional Radiology, SCTIMST, Trivandrum and is submitted in partial fulfillment of the regulations for the award of DM Degree.

I have not submitted this work previously to any university for the award of any degree.

Place

Date Dr Swati Dayanand Chinchure

CERTIFICATE

This is to certify that the dissertation titled **"Imaging and Endovascular management of Cervicofacial vascular malformations"** is a record of work done by **Dr. Swati Dayanand Chinchure** , during the period January 2009- September 2011 at Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram under my guidance and supervision , in partial fulfillment of the regulations governing DM degree Examination of Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram to be held in December 2011.

Place

Date Dr A K Gupta.

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Dr. Swati Dayanand Chinchure

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Introduction

Peripheral vascular malformations are some of the most difficult lesions to diagnose and treat. Overall prevalence of vascular malformations is estimated to be 1.5% of the general population[1].Vascular malformations of the head and neck are rare lesions, thought to result from errors in vascular morphogenesis. They are present at birth and do not regress. However, they often present clinically, later in life[2].

Vascular malformations are believed to be the result of an inborn error of vascular morphogenesis between the 4th and10th weeks of intrauterine life[3]. The pathologic process that creates vascular malformations includes both arrest of normal vascular development and failure of resorption of the embryologic primitive vascular elements. Therefore, immature vascular structures that should have disappeared at birth remain as anomalies. These lesions can be subdivided either histologically based on the predominant vascular channel type, or functionally based on the flow characteristics, ie, high-flow versus low-flow lesions[1,4.]

Vascular malformations occur as a result of aberrant vessel angiogenesis. They are localized or generalized congenital vascular abnormalities comprising direct microscopic connections between arteries, veins, and lymphatic vessels without the normal capillary bed. Vascular malformations have a high recurrence rate because they originate from the mesenchymal cells at an early stage of embryogenesis. They retain the embryonic growth potential, which is often represented clinically as recurrence.

Clinical manifestations vary from none to life-threatening congestive heart failure[1]. Some vascular malformations become increasingly destructive as they continue to grow and progress.

Since the diagnosis of a vascular lesion relies mainly on medical history and clinical examination, diagnostic imaging can be focused on specific structural and functional information. Precise imaging evaluation is needed for treatment of the lesions, not only to evaluate the extent of lesions but also to confirm the suspected diagnoses. It plays major role in treatment planning[5].

Treatment options may depend on the site, size, and complexity of the lesion, as well as the experience and preference of the treating physicians. The available treatment modalities include percutaneous or endovascular embolization or sclerotherapy and surgical resection. Surgery has been the standard treatment, but functional or cosmetic problems sometimes follow surgical therapy. A multidisciplinary approach is essential [6].

AIMS AND OBJECTIVES:

1) To address diagnostic imaging features of cervicofacial vascular malformations.

2) To decide approach of diagnosis in cervicofacial vascular malformations.

3) To decide therapeutic approach focusing particularly on endovascular management for these lesions.

REVIEW OF LITERATURE

Peripheral vascular malformations are some of the most difficult lesions to diagnose and treat. Overall prevalence of vascular malformations is estimated to be 1.5% of the general population[1]. There may be a slightly higher prevalence in girls, with at least one series reporting a female-to-male ratio of 1.5:1.[7] Vascular malformations occur as a result of aberrant vessel angiogenesis. They are localized or generalized congenital vascular abnormalities comprising direct microscopic connections between arteries, veins, and lymphatic vessels without the normal capillary bed[7].

Clinical manifestations vary from none to life-threatening congestive heart failure. Vascular malformations are complex lesions with a variety of clinical manifestations. Vascular malformations are congenital, have an equal gender incidence, virtually always grow in size with the patient during childhood and virtually never involute spontaneously. Although vascular malformations are congenital, they may not be seen at birth. Approximately 40% of vascular malformations are seen in head and neck region[1]. Arteriovenous malformations, having no relation to endothelial proliferation, are caused by abnormal differentiation of the vascular system during embryogenesis[2]. These lesions may not be evident until additional growth or vascular engorgement manifests as a response to thrombosis, trauma, infection, or endocrine fluctuations; thus, these evolutive vascular malformations rarely appear before adolescence. Unlike hemangiomas, vascular malformations generally increase proportionally in size as the child grows. In contrast, more than half of hemangiomas are seen at birth. They have endothelial hyperplasia with increased endothelial turnover. They undergo an initial proliferative phase, and they finally involute with age[2].

Table1: Clinical features to differentiate between various vascular malformations

Clinical findings	capillary	arteriovenous	Capillary venous	venous	venolymphatic
Pulsatility	+/-	+	-	-	-
Bone					
Enlargement	-	-	+/-	-	+
Vascular space	-	+	-	-	-
Phleboliths	-	-	+/-	+	+/-
Compressible	-	-	-	+	+
Progression					
Progressive	+	+/-	+/-	-	+/-
Acute crisis	-	-	+/-	-	+
Valsalva swelling	-	-	+/-	+	-

Different classifications:

Vascular malformations are divided in to various sub-categories depending on the predominant anomalous channels—such as venous, lymphatic, capillary, and arterial malformations. Mixed vascular anomalies are common such as capillary–venous, lymphatic–venous or arteriovenous malformations.[8]

More simply, malformations can be categorized as either low-flow or high-flow lesions on the basis of their hemodynamic flow characteristics. Capillary, lymphatic, and venous malformations are classified as low-flow malformations, whereas any malformation with an arterial component is classified as a high-flow malformation (arteriovenous malformations and arteriovenous fistulas)[9]

Multiple classifications for vascular abnormalities have been established,

but the classification of Mulliken and Glowacki[2] is the most frequently used system. Treatment and prognosis of VMs are based on the type, subtype and architecture of the lesions.

Mulliken and Glowacki[2]

Currently accepted classification of congenital vascular malformations is based on number of histologic and clinical features described by Mulliken and Glowacki[1].

1. Hemangioma of infancy
   Tumors with an early proliferative and later involuting stage
2. Vascular malformations:
   Capillary, lymphatic, venous, arterial, or combined

This classification differentiates nontumorous vascular malformation from vascular tumors like true infantile hemangiomas on the basis of endothelial cell characteristics and number of mast cells. This classification has been useful clinically, has been correlated with angiography[10], and recently has been adopted for interventional radiology[9].

Jackson et a[11]

In 1993, Jackson et a11 proposed another system for classifying hemangiomas, vascular malformations, and lymphatic malformations on the basis of vascular dynamics.

Classification of Jackson et a1[11]

1. Hemangioma
2. Vascular malformation
   - Low-flow lesion
   - High-flow lesion
3. Lymphatic malformation

International Society for the Study of Vascular Anomalies[12]

A classification system for vascular anomalies based on cellular features, flow characteristics, and clinical behaviour was updated during the meeting of the International Society for the Study of Vascular Anomalies[12] (table 2)

Table 2: ISSVA classification of vascular anomalies.

Vascular tumor	Vascular malformations	
	simple	Combined
Hemangioma	Capillary malformation	Arteriovenous fistula, arteriovenous malformation, Capillary venous malformation, capillary-lymphatic venous malformation
other	Lymphatic malformation	Lymphatic –venous malformation, capillary- lymphatic-venous malformation
	Venous malformation	

Kawanabe et al[13]

In 1996, Kawanabe et al[13] reported a system for practical classification of vascular lesions in which vascular malformations are divided depending on its flow pattern and the treatment procedure is selected according to the characteristic flow within the lesion.

Flow related classification and recommended treatment by Kawanabe et al[13]

1. Slow-flow lesion: suggested management- sclerotherapy
2. Intermediate-flow lesion: suggested management- sclerotherapy (plus embolization)
3. High-flow lesion: suggested management- embolization (plus sclerotherapy)

Venous Malformations (VM) :

Venous malformations (VM) are congenital anomalies characterized by irregular endothelial-lined channels, with thin walls deficient in smooth muscle[1]. VM have variable clinical presentations, depending upon site, depth and extent. When superficial enough to be appreciated externally, they typically have a bluish purplish hue, and are soft and compressible. Head and neck venous malformations often expand when the patient is head-down, during valsalva, and during other maneuvers that reduce venous return. Episodic focal thrombosis is nearly ubiquitous and may be associated with swelling and pain. Permanent phleboliths resulting from such episodes are common. Pain associated with venous malformations is complex and multifactorial. Other than the above mentioned thrombosis, muscle fibrosis, and bone and muscle deformity resulting in premature arthropathy can all serve as etiologies for pain and discomfort. In the head and neck in particular, the extent of the lesion is often greater than appreciated on clinical examination[14]. Facial venous

malformations frequently demonstrate extension into the deeper musculature and oral mucosa, and can present with oral bleeding. Additional clinical manifestations typically relate to mass effect from growing lesion within or adjacent to an important anatomic space, such as the orbit or airway. Cosmesis is often an issue as well, with facial asymmetry inducing patients or their parents to seek treatment. Venous malformations are associated with several syndromes, including glomuvenous malformation (glomulin mutation), cutaneomucosal venous malformation (TIE2 mutation), and blue rubber bleb nevus syndrome[15,16].

As manifestations of dysplastic veins, venous malformations by definition drain into the regional normal venous system. Given the typically large caliber of the anomalous venous sac relative to its low inflow, as well as the typically diminutive connectors between the malformation and adjacent normal veins, venous drainage is often delayed; rapid drainage can, however, occur[16].

MRl is the imaging modality of choice; characteristic findings include focal or diffuse collections of high T2 signal, often containing spaces of variable size separated by septations[14]. Small fluid-fluid levels may be visible. Phleboliths may be evident as areas of signal void that are most prominent on gradient images. Flow sensitive images demonstrate no high flow vessels within or around the lesions[14]. Contrast administration results in variable enhancement and is important to distinguish VM from LM[14]. Angiography is not necessary for diagnosis but typically shows either no filling of the malformation, or delayed opacification of sinusoidal spaces with or without dysplastic draining veins. The rapidity, type, and particular pathway of venous drainage impact on management strategy[7].

Lymphatic Malformations (LM):

Lymphangiomas are composed of sequestered, noncommunicating lymphoid tissue lined by lymphatic endothelium and are thought to be caused by congenital obstruction of lymphatic drainage [17,18]. When lymphatic malformations occur in the neck and axilla, they are often called cystic hygromas. Lymphatic malformations consist of cysts that are classified as

1. Macrocystic (cystic components > 2 cm in diameter)
2. Microcystic (cystic components < 2 cm in diameter)
3. Combined.

Both the imaging characteristics and the response to treatment hinge crucially upon this distinction. Lymphatic malformations are particularly transspatial, crossing tissue planes and regional boundaries. The overlying skin or mucosal surfaces may demonstrate lymphatic vesicles (in microcystic cases). Macrocystic lymphatic malformations have a decided predilection for the head and neck region, manifesting there 70% to 80% of the time[18]. Although they are congenital lesions, lymphatic malformations may not present immediately after birth. Clinical manifestations typically appear before the second year of life. Sudden enlargement may follow infection or intralesional hemorrhage. Spontaneous involution, although unusual, has been reported. Numerous syndromes have been reported in association with LM, including Klippel-Trenaunay, Turner, Noonan, and trisomies 13 and 18[19]. The incidence of lymphatic malformations is approximately 2.8 per 100,000 hospital admissions[9], but there are no accepted figures for overall population incidence. Morbidity associated with lymphatic malformations in the head and neck is primarily through recurrent infection, tissue overgrowth, mass effect on functionally important structures, skeletal hypertrophy and via effects on cosmesis. In rare cases outside the head and neck, morbidity can be related to fluid depletion from massive chylous collections in the thorax or abdomen, or to functional impairment from diffuse lower extremity involvement.

On MR imaging, the lesions are generally bright on T2-weighted sequences and isointense to fluid on T1- weighted sequences[18]. Unlike VMs, they generally do not enhance, except for the rims or margins of the cysts. Occasionally, the microcystic lesions may show more generalized enhancement. Thrombi or fluid-fluid levels are common, but phleboliths are not. The macrocysts are seen clearly on ultrasound. Angiography is not needed for diagnosis, but often shows a vague blush or mildly increased vascularity without any arteriovenous shunting[19].

Combined and Syndromic Low-Flow Malformations

Although frequently diagnosed in clinical imaging reports, the true incidence of combined or syndromic vascular anomalies, while unknown, is almost certainly very low[17]. The frequent misdiagnosis results from atypical clinical and imaging appearance of a lesion (eg, amicrocystic LM mimicking a VM, although not manifesting the classic imaging findings). Klippel-Trenaunay patients have all three major types of malformation present in the affected limb; hence the name capillary-lymphatic-venous malformations (CLVM)[17].

High flow vascular malformations (HFVM) in head and neck:

Any lesion that has arterial components is considered a high flow malformation[6]. Compared with the low-flow vascular lesions of the head and neck the HFVMs are rare. The rarity of these lesions and the complexity of the pathophysiology pose a significant management challenge, and no standard treatment paradigm has been established.

These malformations can be of two types[6],

(1) Fistula or direct communication from an artery of visible caliber into a vein of visible caliber

(2) Nidus, a network of abnormal vascular channels bridging the feeding arteries and draining veins.
In either case, normal arterioles and the capillary bed are absent[5]

The high-flow lesions have arteriovenous shunting as an intrinsic feature, ie, shunting of blood under arterial pressure and arterial flow rates into the venous system; herein is the root of much of the pathophysiology of these lesions. The AV shunt presents a risk of hemorrhage, most commonly from rupture of venous structures not designed for arterial pressure, although arterial rupture, particularly at weak points such as flow related or intranidal aneurysms, certainly occurs as well[6]. Additionally, the AV shunt likely causes a localized steal phenomenon, with chronically ischemic tissue in the vicinity of the AVM leading to pain, infection, skin and mucosal breakdown, and so forth. Symptoms are usually referable to the anatomic location of the AVM. The larger and more anatomically central an AVM is, the greater is the likelihood of high output cardiac consequences. Other presenting symptoms can include pain, progressive nerve deterioration or palsy, disfiguring mass, tissue ulceration, hemorrhage as described[6].

Vertebrovertebral fistula:

Vertebrovertebral fistula is a term used to describe various types of fistulas that involve vertebral arteries and veins. Most of these (68%) are of traumatic or iatrogenic in origin, and 32% are spontaneous[20]. Traumatic fistulas are most commonly of iatrogenic cause, secondary to internal jugular vein puncture or to neck surgery. Spontaneous variants are less common and are associated with vascular dysplastic conditions such as neurofibromatosis,[21] fibromuscular dysplasia, and Ehlers-Danlos syndrome[22] .

Symptomatology differs according to the site of the fistula and the flow patterns. Sometimes a neck bruit may be the only presenting sign. In the case

of a proximal fistula, due to the effects on cardiac function, cardiac failure is the presentation[20]. In cases with central venous occlusion of the superior vena cava, reversal of increased internal jugular vein flow causes increase in cerebral venous pressure, which in turn, causes cerebral edema and headache[22]. Severe life-threatening neck hematoma is another important sequel, due to the rupture of a pseudoaneurysm. Finally, with the enlargement of the fistula, dilated epidural venous pouches cause neuronal compression syndrome, which in turn cause motor and sensory deficits[22].

Cirsoid aneurysms:

Cirsoid aneurysms are rare arteriovenous fistulas of the scalp. They are usually congenital in etiology. However, traumatic fistulas have also been described.[23,24,25] They are called "cirsoid" because of the characteristic variceal dilatation of the draining veins. Treatment options include surgical resection, endovascular occlusion, and direct percutaneous injection of sclerosing agents. The radiological findings are important for patient management. In 90% of patients, the superficial temporal artery is the main supply to the fistula with only one dominant feeding artery in 71% of patients[24]. Untreated patients can develop progressive scalp and facial cosmetic deformity from the markedly tortuous subcutaneous vessels. However, this condition is not life-threatening. Surgical resection of the fistula is usually successful. Endovascular and percutaneous occlusion of the fistulas have been described. However, the results have been mixed. The problem with an arterial approach is that there is recruitment of surrounding normal arteries following occlusion of the arterial feeder and draining venous structures. Arterial approaches may not often be successful in occluding the entire fistula[25]. The results of endovascular occlusion are dependent on the angioarchitecture of the fistula, the supplying arteries and draining venous structures. Arterial approaches may not often be

successful in occluding the entire fistula due to the problem of multiple feeding arteries being recruited to supply the fistula[24]. Occlusion of the venous pouch usually requires later surgical removal of the embolic material. A delayed venous phase angiogram is needed to identify all feeding arteries and draining veins, especially if there is drainage into the deep venous sinuses and cortical veins. If the draining sinus is isolated from the rest of the cerebral circulation, it may be possible to consider a transvenous approach using newer embolic material, such as Onyx[25].

Capillary hemangiomas:

Capillary hemangiomas are composed of small vessels lined by flattened endothelium. Capillary hemangiomas are five times more common in girls and are usually detected in first three months of life[17]. Though they are only identified at birth in about a third of patients, hemangiomas typically display a proliferative phase in first six months of life that is characterized by rapid growth of the lesion. During the proliferating stage, infantile hemangiomas may also be considered high-flow lesions[17]. Gradual involution is usually detected by first year and is the hallmark of capillary hemangioma. Involution is typically complete by age seven and will occur in over 95% of patients presenting in infancy[26].

During the proliferative phase, hemangiomas are high-flow lesions that are often revealed by bruit, pulsatility, and increased warmth[27]. Hemangiomas can have deep, superficial, or mixed components. The clinical appearance of hemangiomas varies with the degree of dermal involvement and the depth of the lesions. A characteristic straw- berry appearance is present when the lesions involve the skin. Deep hemangiomas, which do not involve the subcutaneous tissues, may have a blue appearance.

Since spontaneous involution is expected in over 95% of children presenting with capillary hemangiomas, conservative treatment is warranted in vast majority cases[17]. Some patients may require more urgent therapeutic

intervention for example mass effect compromising anatomically critical structures (air ways, vision), or causing functional impairment, congestive cardiac failure, consumptive coagulopathy.

Hemangiomas can be associated with a number of abnormalities. One cluster of abnormalities has been referred to as the PHACE syndrome: posterior fossa abnormalities, facial hemangiomas, arterial abnormalities, cardiovascular defects, and eye abnormalities. Sturge-Weber syndrome is a trigeminal nerve distribution capillary malformation with intracranial abnormalities[17].

Cavernous hemangiomas:

Cavernous hemangiomas are composed of dilated, blood-filled spaces lined by flattened endothelium and overall are less common than capillary lesions [28]. Cavernous hemangiomas frequently involve the deeper, soft tissues and manifest clinically as masses without other diagnostic features. Unlike capillary hemangiomas, they do not involute and may require surgical resection[28]. Young children are usually affected, and calcification may be present, typically in the form of dystrophic mineralization in an organizing thrombus (phlebolith).

Table 3: Differences between hemangiomas and vascular malformations

Haemangioma	**Malformations**
Exhibit cellular proliferation	Comprised of dysplastic vessels
Small or absent at birth	Present at birth
Rapid growth during infancy	Growth proportional to child
Involution during childhood	No regression

DIAGNOSIS: IMAGING CHARACTERISTICS

The primary goals of imaging vascular malformations or hemangiomas include characterizing the lesion and discovering the anatomic extent of disease. Knowing which tissues the vascular malformation involves and whether adjacent vital structures, such as neurovascular bundles, are involved by the lesion is important. Such information is vital to planning surgery or imaging-guided procedures. On physical examination, determining whether the subcutaneous tissue, the underlying deep muscular tissues, or both are involved is difficult[9].

In general, evaluation of vascular malformations requires delineation of its components:

(1) Location, size, and tissue involvement
(2) Origin, orientation, and course of feeding arteries
(3) Origin, size, and course of the draining veins.

USG:

Superficial and small lesions are well examined by ultrasound, with gray scale studies defining the extent and compressibility of the lesion, and spectral and color doppler interrogation used to identify the flow characteristics[9]. Very superficial lesions may be remarkably inconspicuous on MR imaging and yet well defined on ultrasound. Additionally, ultrasound has significant utility in providing image guidance for sclerotherapy[5,7,9].

Venous malformations typically show compressible, confluent anechoic-hypoechoic channels on ultrasound (which are demonstrably venous on color Doppler imaging), separated by more solid regions of variable echogenicity. Sonography can also play a crucial role in differentiating macrocystic and microcystic lymphatic malformation[17]. Macrocystic lymphatic malformations

show anechoic cysts, often containing internal debris or fluid-fluid levels resulting from episodes of hemorrhage. Internal septations are common and best visualized with sonography. Microcystic lymphatic malformations are ill-defined echogenic masses, showing diffuse involvement of surrounding tissue[18].

In HFVM, Color Doppler sonography indicates a direct connection between the arterial and the venous systems and resistive indexes indicate low-resistance flow[5]

CT AND CT ANGIOGRAPHY:

The advent of CT in the late 1970s enabled direct visualization of the soft tissues and its pathology for the first time because of its high contrast resolution. With further development, spiral CT scanners offered faster imaging algorithms and the possibility to image entire anatomic regions relatively quickly. MDCT systems have been a revolutionary advancement in CT technology. They provide greater anatomic coverage, higher spatial resolution, faster acquisition times, and improved image quality[5,9]. These faster scanning techniques improve temporal resolution, resulting in dynamic images of the arterial and venous systems. The ability to obtain isotropic volumetric data with superior spatial resolution enables high-quality three dimensional picture displays of the curved vascular structures. In cases of complex arteriovenous malformations and fistulas, dynamic CT angiogram allows identification of feeding arteries, nidus, and draining vessels and enhances the preoperative anatomic assessment of such vascular lesions. CT is crucial in delineating skeletal involvement, in particular the mandible, skull base, orbits, and calvarium[9].

MRI and MR ANGIOGRAPHY:

The unparalleled contrast resolution of MRI reliably determines full extent of the lesion than any other modality. Pre- and post-contrast T1-weighted images as well as T2-weighted images, and consider fat suppression to be critical in all MR sequences in the workup of vascular anomalies[30].

Venous malformations often display irregular intervening venous walls within the hyperintense areas, and occasionally adjacent enhancing serpentine vascular channels can be seen. Thrombi can have variable appearances, based on their age. Phleboliths are hypointense on all sequences on MR, are dense on CT, and are typically mobile (although confined to the venous space). Although phleboliths have been thought to be virtually pathognomonic of venous malformations, they can rarely occur in lymphatic malformations as well [7].

Untreated, both venous and lymphatic malformations typically appear hyperintense on T2 imaging, and the scope of the abnormal T2 signal can be used to define the overall extent of the lesion. After treatment by sclerotherapy, with the resultant progressive conversion of lesion to scar tissue, the T2 signal characteristically becomes less hyperintense. The enhancement pattern after contrast administration is a crucial differentiating feature, with venous malformations most characteristically enhancing avidly but in a patchy, heterogeneous pattern. In contrast, enhancement of lymphatic malformations is variable. Mild rim enhancement may be seen with macrocystic lymphatic malformations, with only minimal enhancement identified in microcystic LMs; however, in the setting of superinfection or inflammation, LMs may enhance avidly as well[9,31].

MR imaging of proliferating hemangiomas often shows a discrete lobulated mass that is hyperintense to muscle on T2-weighted images and isointense to muscle on T1-weighted images[30]. Typically, prominent draining veins will be identified as both central and peripheral high-flow vessels [30]. Hemangiomas

usually enhance diffusely with gadolinium. Involuting hemangiomas can indicate areas of fibrofatty tissue with associated high signal intensity on T1-weighted images and less contrast enhancement than that of proliferating hemangiomas[30]

On MR imaging, the HFVM appear as a tangle of multiple flow voids that indicate high flow on gradient-echo images. Although the lesions can be associated with surrounding edema or fibrofatty stroma, usually no focal discrete soft-tissue mass is found. Newer sequences, such as time-resolved MR angiography and MR perfusion imaging, may further characterize the flow characteristics and hemodynamics [30,31].

DSA:

Catheter angiography with digital subtraction still remains the gold standard for the evaluation and characterization of the cervical and cranial vascular malformations[7]. However, catheter angiography remains an invasive procedure and requires significant experience to perform it safely. The disadvantage of DSA is

1. Invasive procedure
2. Exposure to ionizing radiation
3. Iodinated contrast.

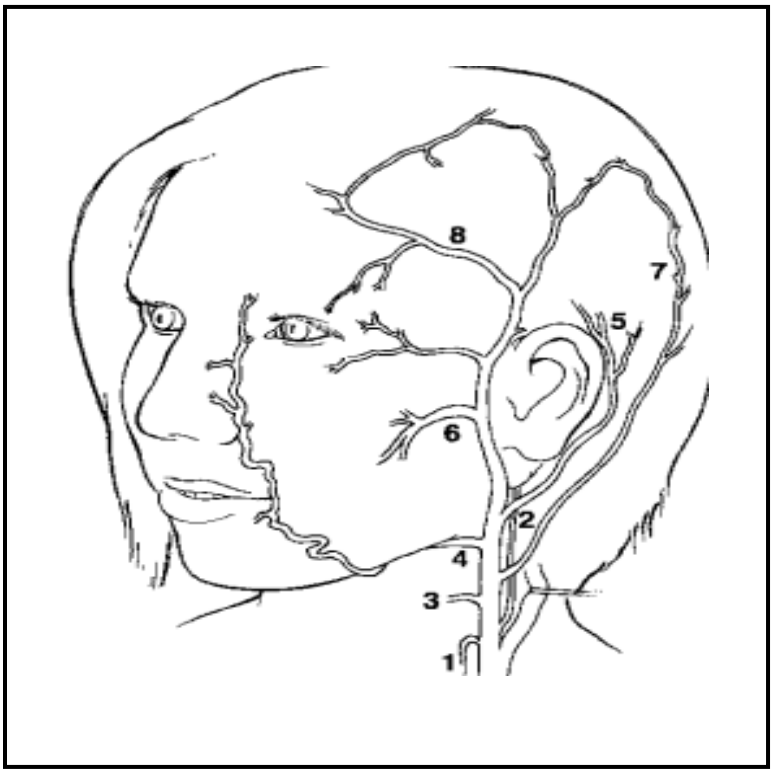
Inherent to the invasive nature there is risk of arterial dissection and puncture site complications. In recent years, advances in CTA and MRA imaging have replaced DSA in many situations for diagnostic information[9]. DSA has also undergone technologic advances with enhanced machine capabilities and newer postprocessing softwares. Current systems are equipped with flat panel detectors and offer significant improvement in low-contrast resolution which is of great importance in neuro-interventional procedures.

ECA ANATOMY

An understanding of the anatomy of the ECA is essential for safe and effective endovascular therapy. The ECA originates from the bifurcation of the common carotid artery and lies anterior to the internal carotid artery (ICA) in 94% of patients. The short trunk of the common ECA progressively decreases in size as it gives rise to eight branches, terminating in the largest of those, internal maxillary artery

External carotid artery.

(1) Superior thyroid artery;
(2) Ascending pharyngeal artery;
(3) Lingual artery;
(4) Facial artery;
(5) Posterior auricular artery;
(6) Internal maxillary artery;
(7) Occipital artery;
(8) Superficial temporal artery

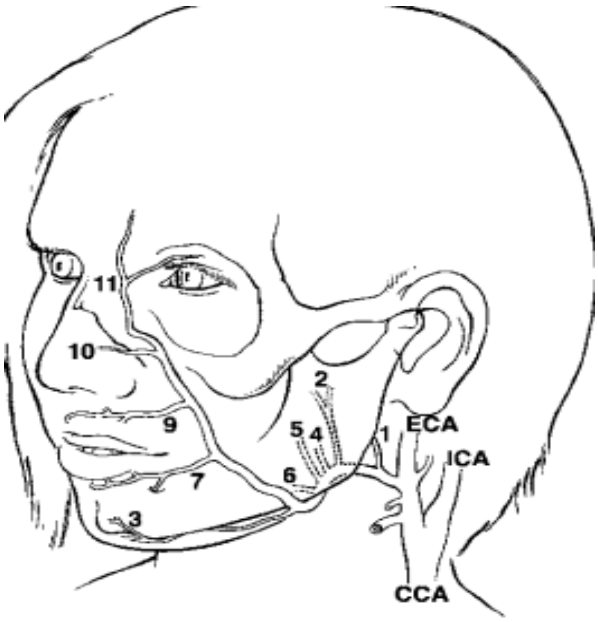


The **superior thyroid artery** is typically the first branch from the ECA. It supplies the thyroid gland, larynx, and submandibular gland. The ascending pharyngeal artery is usually the second overall branch and the first posterior branch of the ECA. It supplies the pharynx, dura, lower cranial nerves, and the middle ear. It divides into pharyngeal (anterior) and neuromeningeal (posterior) trunks. The neuromeningeal trunk gives rise to the inferior tympanic artery, which supplies the middle ear and the hypoglossal artery. The hypoglossal artery supplies the posterior meninges; vasa nervosum of the XII cranial nerve; and the jugular branch of the odontoid system, which supplies the vasa nervosum of cranial nerves IX, X, and XI, and cranial nerve VI that is proximal to Dorello's canal.

The **lingual artery** supplies the tongue and floor of mouth and frequently has anastomoses with terminal divisions of the facial artery, although there are no contralateral collaterals within the tongue musculature end divisions.

A large branch of the ECA supplies much of the face.

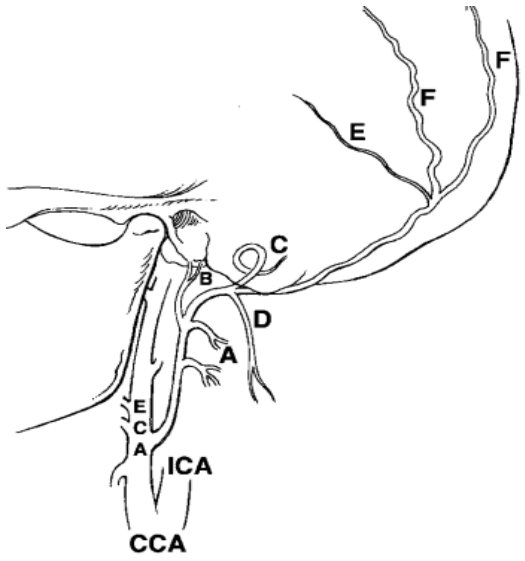
The **facial artery** divides into multiple terminal branches—most notably the superior and inferior labial arteries, lateral nasal artery, tonsillar artery, submental artery, buccal and masseter arteries, ascending palatine artery—and terminates in the angular artery. The last branch courses along the medial orbit and has variable anastomoses with orbital branches of the ophthalmic artery, namely the dorsal nasal artery.



Facial artery. (1) Ascending palatine artery; (2) Tonsilar artery; (3) Submental artery; (4) Inferior masseteric artery; (5) Jugular trunk; (6) Middle mental artery; (7) Inferior labial artery; (8) Anterior jugal artery; (9) Superior labial artery; (10) Lateral nasal artery; (11) Angular artery

The largest posterior ECA branch, the **occipital artery**, forms multiple communications with the adjacent posterior auricular, ascending pharyngeal, superficial temporal, and cervical muscular branches of the vertebral arteries. The posterior auricular artery is a small, posterior branch of the ECA that provides superficial supply to the scalp and pinna; it often communicating with the occipital artery. Originating variably from the terminal divisions of the ECA or the ECA itself, the transverse facial artery often communicates with divisions of both the facial and the internal maxillary arteries (IMAs). The ECA terminates into the IMA and the superficial temporal artery (STA). The STA supplies the scalp through frontal and parietal branches. The IMA is the larger terminal division of the ECA and is the primary supplier to the deep, midline facial structures, forming multiple anastomoses with facial artery and SFA, as well as potential collaterals with the internal circulation.

Occipital artery. (A) Sternocleidomastoid branches; (B) Stylomastoid artery; (C) Mastoid branch; (D) Descending branch; (E) Lateral meningeal branch; (F) Occipital branches.



The **internal maxillary artery** course is divided into mandibular, pterygoid, and pterygopalatine segments. The most proximal, mandibular segment gives off the inferior alveolar and middle and accessory meningeal arteries, the last of which can form anastomoses with the inferolateral trunk of the ICA as it feeds the trigeminal ganglion. The pterygoid segment gives rise to the masseter and buccal arteries and anterior and posterior temporal arteries. The pterygopalatine segment yields multiple terminal divisions that supply the mid face, including the superior alveolar artery, the infraorbital artery, the descending palatine artery, and the sphenopalatine artery (the primary supply to the nose). There are smaller, posteriorly directed branches from the pterygopalatine including the Vidian artery.

Dangerous Anastomoses

An operator must be cognizant of the multiple and variable potentially **dangerous anastomoses** represented in the head and neck arterial anatomy. Failure to recognize such anastomoses can cause stroke, cranial nerve injury, blindness and permanent neurologic deficit. The cavernous (C4) and petrous (C2) portions of the ICA have multiple potential sites of anastomoses primarily from divisions of the IMA and the ascending pharyngeal artery. The middle meningeal, accessory meningeal, and foramen rotundum arteries communicate with the inferolateral trunk in a cavernous sinus anastomosis. The IMA also gives off pterygovaginal and Vidian artery divisions that communicate with the mandibular and vidian arteries from the petrous ICA in a eustachian anastomosis. The anterior tympanic artery, a branch of the neuromeningeal trunk of the ascending pharyngeal artery, communicates with the corticotympanic artery from the petrous ICA in a middle ear anastomosis. Stylomastoid and petrosal branches from the occipital and middle meningeal arteries contribute as well to the middle ear anastomosis. Meningeal branches of the ascending pharyngeal neuromeningeal trunk communicate with clival divisions of the meningohypophyseal trunk in a clival anastomosis. Finally, the superior branch of the pharyngeal trunk of the ascending pharyngeal may communicate with the inferolateral trunk of the ICA.

Table4: Danger zones: Common anastamoses: Anterior circulation[32]

Territory at risk	Anastamosis from	Anastamosis to
Brain: Anterior circulation	Ascending pharyngeal, neuromeningeal trunk	Cavernous carotid via meningo-hypophyseal trunk
	Ascending pharyngeal, inferior tympanic branch	Petrous carotid via caroticotym-panic
	Ascending pharyngeal, superior pharyngeal	Cavernous carotid via inferolateral trunk
	Ascending pharyngeal, superior pharyngeal	Petrous carotid via mandibular branch
	Accessory meningeal (cavernous branch) Middle meningeal (cavernous branch)	Cavernous carotid via inferolateral trunk, posterior branch Cavernous carotid via inferolateral trunk, posterior branch
	Middle meningeal (cavernous branch)	Cavernous carotid via meningo-hypophyseal trunk
	Distal internal maxil-lary (artery of foramen rotundum)	Cavernous carotid via inferolateral trunk, anterolateral branch

The ECA exhibits multiple potential connections to the ophthalmic artery primarily from the IMA but also from the facial artery and the SFA. The IMA anastomoses arise from the (1) infraorbital to the recurrent ophthalmic arteries, (2) the anterior deep temporal and meningo-ophthalmic (via the middle meningeal artery) arteries to the lacrimal artery, and (3) the sphenopalatine and ethmoidal (via the middle meningeal artery) arteries to the ethmoidal arteries (via the ophthalmic). The angular artery via the facial artery communicates with the dorsal nasal branch of the ophthalmic artery, and the transverse facial artery communicates with the ophthalmic artery via zygomatico orbital branches.

Table 5: Danger zones: Common anastamoses: Ophthalmic artery

Territory at risk	Anastamosis from	Anastamosis to
Eye (and secondarily brain)	Middle meningeal, sphenoidal branch	Ophthalmic
	Middle meningeal, frontal branch	Ophthalmic via anterior falx artery
	Inferolateral trunk, anteromedial branch	Ophthalmic
	Distal internal maxillary, anterior deep temporal	Ophthalmic
	Distal internal maxillary, infraorbital	Ophthalmic
	Distal internal maxillary, sphenopalatine	Ophthalmic via ethmoidal branches
	Distal facial	Ophthalmic
	Transverse facial	Ophthalmic
	Superficial temporal, frontal branch	Ophthalmic
	Cavernous carotid, inferolateral trunk	Ophthalmic via recurrent meningeal branch

The ECA has multiple connections to the C1-C4 vertebral artery divisions via the (1) musculospinal and neuromeningeal trunk (hypoglossal artery) from the ascending pharyngeal artery and (2) the radicular and cervical muscular branches from the occipital artery.

Table 6: Danger zones: Common anastamoses: Posterior circulation

Territory at risk	Anastamosis from	Anastamosis to
Brain: Posterior circulation	Ascending cervical	Vertebral segmental branches
	Deep cervical	Vertebral segmental branches
	Occipital, muscular branches	Vertebral segmental branches
	Ascending pharyngeal, muscular branches	Vertebral segmental branches
	Ascending pharyngeal, neuromeningeal trunk	C3 segmental vertebral via odontoid arch

Table 7: Cranial nerve blood supply[32]

Cranial nerve	Arterial supply
I: Olfactory	Anterior cerebral
II: Optic	Supraclinoid carotid, ophthalmic
III: Occulomotor	Basilar, superior cerebellar, posterior cerebral, inferolateral trunk, ophthalmic
IV: Trochlear	Inferolateral trunk, meningohypophyseal trunk
V: Trigeminal	Inferolateral trunk, meningohypophyseal trunk, middle meningeal, accessory meningeal, artery of foramen rotundum, infraorbital
VI: Abducens	Inferolateral trunk, meningohypophyseal trunk, middle meningeal, accessory meningeal, ascending pharyngeal(jugular branch)
VII: Facial	Stylomastoid (from post auricular or occipital), middle meningeal (petrous branch), ascending pharyngeal (inferior tympanic and odontoid arcade)
VIII: Auditory	Basilar, AICA, ascending pharyngeal jugular branch
IX: Glossopharyngeal	Ascending pharyngeal jugular branch
X: Vagus	Ascending pharyngeal jugular branch, superior and inferior thyroid, laryngeal branches
XI: Spinal Accessory	Ascending pharyngeal (jugular, inferior tympanic and musculospinal branches)
XII: Hypoglossal	Ascending pharyngeal, hypoglossal branch and proximal trunk, occipital, directly from external carotid, lingual

ENDOVASCULAR MANAGEMENT

Vascular malformations were initially treated by surgeons alone. The early rationale of proximal arterial ligation of AVMs proved totally futile as the phenomenon of neovascular recruitment reconstituted arterial inflow to the AVM nidus. Microfistulous connections became macro fistulous feeders. Complete extirpation of a vascular malformation can be very difficult and, at times, even hazardous necessitating suboptimal partial resections. Very often the patient's presenting symptoms recur are worsen on follow-up[33]. Because of the significant blood loss that frequently accompanies surgery, the skills of interventional radiologists were eventually employed to embolize these vascular lesions preoperatively.

There is strong evidence in the literature [33] that
1. External carotid artery ligation inevitably leads to recruitment of collateral circulation from the internal carotid, vertebral, and the contralateral external carotid circulations, making subsequent treatments more difficult, and must be avoided at all cost.
2. Proximal vessel ligations, or embolization do little to decrease blood flow since recruitment of new feeding vessels quickly re-establishes blood supply to the lesion, and the reactive nonsprouting angiogenesis may become indistinguishable from the nidus and should be avoided at all costs.
3. Only complete removal, either by surgical resection or embolic devascularization of the vascular malformation, results in a cure.

The differentiation of vascular malformation into high and low flow is utmost clinical importance, as treatment options are drastically different. The cornerstone treatment of low flow vascular malformations is percutaneous sclerotherapy[34,35]. Percutaneous sclerotherapy is not effective for high flow lesions since the infused agents are rapidly washed away from the endothelial

lining. The most effective treatment for high flow lesions is transarterial embolization, with occasional subsequent surgical resection[36,37].

MANAGEMENT OF VENOUS MALFORMATION:

Direct injection of a sclerosing agent (98% ethanol, sodium tetradecal, or sodium ducal) results in thrombosis and gradual shrinkage of the malformation and is the preferred treatment [17]. Angioarchitecture of venous malformations divide it in three forms: focal, multifocal, and diffuse forms. Focal lesions may be intramuscular, cutaneous or mucosal and usually consist of collections of abnormal interconnecting channels or spaces that are "sequestered" or drain through fairly small channels to normal adjacent conducting veins. This type of lesions are easily treated by sclerosant injection[17,38]. Diffuse venous malformations involve multiple tissue layers and usually include muscle, subcutaneous fat, skin, and sometimes bone. In diffuse lesions, the malformed veins are not sequestered but communicate directly with the main conducting veins, which frequently are also abnormal[17]. Diffuse VMs are difficult to treat effectively, because injected sclerosant can directly enter the circulation, potentially causing deep venous thrombosis, pulmonary embolism, or systemic effects of ethanol. Recanalization also is more likely than after sclerotherapy of sequestered lesions. Sclerotherapy may still be beneficial, because the fibrosis that results gradually decreases the amount of swelling the patient experiences. Some patients with diffuse venous malformations have focal eccentric varices, which can exert considerable mass effect on adjacent structures. These varices can be obliterated nicely with endovascular treatment[17].

The technique of sclerotherapy involves the percutaneous catheterization of the malformation using a needle or Teflon-sheathed needle

cannula. After confirming free blood return, contrast is injected, recorded with serial angiographic imaging or road-mapping to document the cannula position within the malformation and the presence or absence of venous outflow. In the presence of significant venous outflow, local compression is applied and contrast injections repeated until the venous outflow no longer fills.

Percutaneous Ethanol embolization for venous malformations

Alcohol injection is extremely painful, and should be done under general anesthesia; the subsequent swelling is pain-free if the agent has been injected within the malformation itself. Ethanol has the potential for severe complications, which the anaesthesiologist should be prepared to treat. Absolute ethanol is the most commonly used sclerosing agent due to its superior ability to cause endothelial damage and induce thrombosis and sclerosis. Disadvantages of ethanol include its neurolytic and cardiovascular effects, CNS depression, and pain on injection. Ethanol should not be used in lesions adjacent to major nerves, such as the facial nerve in the parotid region, or in cutaneous lesions[38].

An estimation of the volume of the cannulated compartment within the malformation is calculated from the contrast material injection, as the venous drainage is seen, and the walls of the compartment are convex outward. Contrast medium is then aspirated or pressed out of the compartment and a similar quantity (usually one-third less of the calculated volume) of sclerosing agents is injected; 98% ethanol is usually opacified by mixing it in a 1:3 Ethiodol or Pantopaque: ethanol ratio. Ethanol denatures the blood cells within the malformation and dehydrates and scleroses the vessel wall if sufficient contact between the sclerosing agent and the endothelium is attained. The injected part of the malformation becomes firm and noncompressible because of thrombus formation within approximately 10 min[38]. If the lesion does not become firm, and if there is persistent blood return from the cannula, additional ethanol may be injected, as swelling correlates with outcome. The

total volume of injected ethanol should not exceed 0.3 ml/kg in children below the age of 2, and 0.5ml/kg in older children [17,38].

Percutaneous detergent sclerosing agent:

This group includes sodium tetradecyl sulphate (STS), polydocanol, sodium morrhuate, and ethanolamine. Like ethanol, these drugs all damage the endothelial cells, resulting in thrombosis and fibrosis. Thrombosis occurs more slowly than with ethanol and there is probably a greater tendency for recanalization. These drugs are not neurolytic and there has been only one report of a cardiovascular complication (from polydocanol) [39]. As with ethanol, the cannula position should be confirmed with contrast medium injection or sonography before injection of sclerosant, as arterial injection of any of these drugs causes severe tissue damage. Foaming the sclerosant by mixing the drug with air (or air and oily contrast medium) has become popular[40]. It is felt that the use of foam is more effective than the use of liquid sclerosant alone. Possibly, the foam results in better contact between the drug and the circumference of the vein wall, and more prolonged displacement of the intralesional blood. One method of making foam is to mix 10 ml of detergent sclerosant with 3 ml of Ethiodol and 5–10 ml of air, through a three-way stopcock[39]. Patients receiving large amounts of detergent sclerosants must be aggressively hydrated to treat the associated hemoglobinuria[39].

In the treatment of benign conditions, it is our preference to stage the sclerotherapy to diminish the risk of complication. Treatment of large malformations is therefore invariably staged.

In general, the more localized deep venous malformations respond well to direct injection of a sclerosing agent. Likewise, small cutaneous lesions, such as those in patients with multiple venous malformations, often respond very favorably. In venous malformation involving the gingival mucosa, the treatment can be very effective, but one should be cautious not to use large

amounts of sclerosing agent in order to avoid mucosal necrosis[38]. Diffuse lesions are much more resistant to sclerotherapy.

Lymphatic Malformations

Previously, it was felt that macrocystic LMs were best treated by excision. However, in the past decade, it has become clear that macro-cystic lesions often respond very well to treatment by sclerotherapy, which has a much lower risk of morbidity[17]. Microcystic LMs, in general, respond less well to sclerotherapy, although in some instances, such as orbital lymphatic malformations, cysts or channels measuring as little as 5 mm in diameter can be accessed and injected with good results[17].

Sclerosant Drugs for Lymphatic Malformations-

Sclerotherapy agents useful for treating lymphatic malformations (LM) include ethanol, doxycycline, bleomycin, Ethibloc and OK-432.

OK-432 is undergoing clinical evaluation in a multicenter head and neck trial under the supervision of the University of Iowa. It is a solution of killed group A streptococci in a suspension that contains penicillin. It causes an inflammatory reaction and subsequent shrinkage of the cysts [41]. Side effects, aside from postprocedure swelling, appear to be minimal and this agent has been shown to be effective in 80%– 90% of macrocystic lesions[42]

Ethibloc (a suspension of ethanol,zein protein, and contrast medium) has been used extensively for macrocystic LM in Europe and Canada[43]. It is effective in a high percentage of macrocystic LM, but has an unfortunate side affect of skin breakdown and extrusion. Ethanol is useful for treating relatively localized macrocystic lymphatic malformations. It does have a risk of nerve injury and skin necrosis. Doxycycline can be used more effectively in large or extensive lymphatic malformations, because large volumes administered in one session are well tolerated. It is generally used as a solution of 10 mg/ mL, in

volumes up to 100 mL. STS generally is not used for sclerosing LM, except intraorbital lesions[44]. Bleomycin is an antibiotic derivative with cytostatic properties. It has antimitotic activity with antiangiogenic properties. Bleomycin is effective in treatment of macrocystic LM as well as in hemangiomas; however, because of potential systemic toxicity (pulmonary fibrosis, alopecia, pigmentation), it must be used in small doses [45].

The technique of sclerotherapy of LM differs from that of VM. Individual cysts are cannulated with a needle with US guidance[17,44]. For large cysts, a standard 20- guage angiocatheter with a side hole cut in the plastic outer sheath can be used. Fluid is aspirated as completely as possible. The cysts can be outlined with a small amount of contrast medium, which is then aspirated, or the needle position can be confirmed with US alone. The sclerosing agent can be injected with US or fluoroscopic guidance. For lesions consisting of smaller cysts, it is not practical to inject each cyst with contrast medium. Rather, US guidance is more practical and effective. Because LMs have a high rate of spontaneous infection, patients should be treated immediately before and for approximately 10 days after the procedure with prophylactic antibiotics post procedure.

Patients do not require excessive hydration; however, especially if doxycycline is used, adequate analgesia should be provided. Regression of the macrocystic lesions occurs slowly and the results should be assessed at approximately 6 weeks after injection. If additional treatment is required, 6–8 weeks is an appropriate interval [17, 44].

ENDOVASCULAR MANAGEMENT OF HIGH FLOW VASCULAR MALFORMATIONS

The ultimate target for endovascular embolization is occlusion of the nidus and initial segment of the venous outflow. Embolization may be curative, palliative (for symptomatic control of unresectable lesions), or preoperative, with the choice of agent depending on the intended outcome. Treatment options may depend on the site, size, and complexity of the lesion, as well as the experience and preference of the treating physicians. The available treatment modalities include percutaneous or endovascular embolization or sclerotherapy and surgical resection. A multidisciplinary approach is essential. The best chance for a complete cure of AVMs of the head and neck seems to be via a combination of preoperative embolization and surgical resection[46,47]

Vascular malformations at different locations:

Orbits:

Orbital AVMs are best considered to be congenital hamartomas, with trauma possibly precipitating hemodynamic changes, leading to symptoms. Based on location, they may be classified into 3 types: purely orbital, orbital and periorbital, and orbital with retinal or cerebral AVMs (Wyburn-Mason syndrome) [6,48]. Diagnosis of orbital AVMs is based on angiographic findings highlighting an engorged, rapidly filling proximal arterial system, a malformation, and distal venous outflow. The diagnosis can be aided by clinical history and noninvasive tests, such as flow Doppler studies, and by computed tomography and magnetic resonance imaging to highlight the extent of the lesions. Because these lesions are rare, they must be considered in the differential diagnosis of orbital vascular lesions with similar clinical features, such as carotid-cavernous fistulas, dural cavernous sinus fistulas, orbital arteriovenous fistulas, and cerebral AVMs with drainage into orbital veins. The

management of orbital AVMs is based on a multidisciplinary approach [6]. As described earlier, the slow growth and the low incidence of hemorrhage permit observation in many cases. Regression is well documented in cerebral AVMs but has not been reported in orbital lesions. Indications for intervention include visual compromise, patient discomfort related to symptoms, and aesthetic concerns when the lesions extend outside the orbit. The primary treatment for orbital AVMs is surgical excision with or without preoperative embolization. Visual compromise and persistent or progressive patient discomfort are the main indicators for intervention. Recurrences were reported when incomplete excision or partial embolization alone was performed, highlighting the tendency of these lesions to recruit new feeder vessels when their supply is partially reduced[6]. The risk-benefit ratio must be evaluated on a case-by-case basis before interventional management is undertaken in orbital AVMs. Their natural history must be understood and considered, alongside the risks of neuroradiologic and surgical interventions. [6]

Orbital cavernous hemangiomas are most common vascular lesions in adults. Cavernous malformations usually are solitary and most often occur in the lateral aspect of the retrobulbar intraconal space. On imaging, cavernous malformations typically are well circumscribed, round or ovoid, homogeneously hyperattenuating, intraconal lesions. They occasionally contain microcalcifications (phleboliths) and may produce expansion of the orbital walls. The lesions may displace adjacent structures but do not invade them. At multiphase dynamic contrast enhanced CT, poor enhancement can be seen in the early arterial phase because of the low-flow arterial supply ;contrast material does not fill the central part of the lesion until the late venous phase. The MR appearance of cavernous hemangiomas is that of a smooth or slightly lobulated well-circumscribed mass of low intensity on T1-weighted images, that is, isointense to muscle. Occasionally, these lesions contain regions of high signal intensity on T1-weighted images, corresponding to thrombosed vascular spaces

They are usually markedly hyperintense on T2-weighted images. If an intralesional flow void is seen, another diagnosis should be considered, such as vascular hemangiopericytoma, venous varix, or an arteriovenous malformation. Cavernous malformations enhance with IV contrast; however, the enhancement pattern is variable. Because the contrast agent enters these lesions slowly, images obtained in a dynamic fashion or otherwise quickly after injection usually show minimal or heterogeneous enhancement. Enhancement is more uniform on images obtained after more delay. The major role of MR in the evaluation of cavernous hemangiomas is to provide the precise anatomic delineation of the mass and its relationships to the optic nerve and other orbital structures[49].

These lesions are usually managed conservatively, and surgical excision is reserved for those that cause severe proptosis or optic nerve compression[37]. These lesions are surgically curable because they are encapsulated and do not recur if completely removed. Because of the inaccessibility of the small feeding arteries and the multiple collateral pathways available for recanalization, embolization therapy is not often performed[48,49].

Venous lymphatic malformations in orbit They may be evident at birth, but they generally manifest in infancy or childhood. Although venous lymphatic malformations may enlarge slowly, producing progressive proptosis, restriction of eye movements, or vertical globe displacement, many manifest abruptly because of hemorrhage. Hemorrhages within these malformations often occur after minor trauma or infection and occasionally develop spontaneously. Lymphatic malformations are unencapsulated, diffuse, and multicompartmental, often including both intraconal and extraconal components that are insinuated between normal orbital structures.
The lesions may extend across tissue planes to infiltrate the eyelid and orbit, and they may cause bone remodelling. Orbital venous lymphatic malformations are isolated from the normal orbital vasculature and, unlike varices, are not

affected by postural changes. However, they may be associated with intracranial vascular malformations, especially developmental venous anomalies. The signal intensity of the lesions depends on the type of fluid within the cystic components, whether hemorrhage has occurred, and the age of the hemorrhage.T1-weighted images best depict lymphatic or proteinaceous fluid, and T1Weighted fat-suppressed images are best for detecting blood or blood products.T2- weighted fat suppressed images provide improved visibility of components that contain hemorrhagic fluid. The use of contrast material does not typically provide significant additional information, but an absence of enhancement is indicative of a lymphatic component[48]. Fluid-fluid levels produced by hemorrhages of various ages within the lesion. Because the natural history of these lesions is variable and unpredictable, their treatment is controversial and problematic; the best method of treatment depends on the growth, size, location, and morphology of the particular lesion. Observation and conservative management, when possible, are recommended. Surgery has been suggested for lesions that cause marked stretching or compression of the optic nerve; however, complete surgical excision of diffuse lesions is usually impossible, and recurrence is common. Surgery is therefore reserved for dire situations such as optic nerve dysfunction, corneal compromise, severe discomfort, or impending amblyopia [48]. Various alternative therapies have proved successful or promising, including the intralesional injection of sclerosing agents [49] fractionated beta irradiation ,carbon dioxide laser ablation, and intralesional injection of steroids[17].

Pinna

Arteriovenous malformations are rare in the auricular region, however it is the second most common site for AVMs of the head and neck[6]. The most common Feeding arteries in this region are the posterior auricular, superficial temporal, and occipital arteries. If the AVM is small and asymptomatic, no treatment is

required, especially in children. For a symptomatic AVM, complete excision with prior embolisation is the treatment of choice. Embolisation alone can be used for palliation of lesions located in difficult-to-approach areas or very close to vital structures. Total resection requires a wide-field resection of all the involved tissue is necessary to prevent recurrence, however, cosmetic and functional issues might limit the extent to which tissue can be removed. Partial excision usually leads to rapid recurrence so in these cases the remaining AVM tissue must be obliterated using intravascular embolisation[50]. Although ultimately most auricular AMV patients are likely to face amputation, selective palliative embolization is an important part of the therapeutic armamentarium. The margin of resection is not altered with preoperative embolization[50].

Tongue:

Lingual hemangiomas may remain indolent or may produce obstructive symptoms or alarming hemorrhage. Most lingual tumors present as mucosal changes and tongue being superficially located and easily accessed, these can be diagnosed without imaging analysis. However, the characteristic and extent of lesions situated at deep portion of tongue, such as its base or submucosal lesions can be recognized only on cross-sectional CT scan or MRI. A number of options exist for lesions that require therapy, including medical and surgical interventions. Medical management includes systemic and intralesional administration of corticosteroids. However, only 30% respond to corticosteroids and they are not free from complications[51]. Systemic corticosteroids carry well-documented risk, such as disseminated varicella, herpes infection, growth retardation and cushingoid habitus. For lesions, which do not respond to steroids, surgical therapy is often necessary. Surgery may be complicated by extreme blood loss. Surgical resection may be facilitated by pre-operative embolisation in selected cases although embolisation has also been used as the

sole form of treatment for unresectable lesion[51]. Laser photocoagulation is the other modality of surgical treatment. Both surface and intralesional delivery of laser phototherapy are used for treatment of hemangiomas and vascular malformation, but recent interest has centered on the latter. Although laser therapy has fewer complications, the frequent numbers of treatment, variable response and regrowth of lesion are the disadvantages of this technique.

Lymphatic malformations presents as giant masses of tongue at birth. When these lesions are diagnosed prenatally, airway compromise can be anticipated at birth. Prenatal MR imaging has been advocated in the evaluation of giant neck masses to determine the anatomic location of the lesion and the relationship of the lesion to the airway. Two treatment options are available for such large mass: intralesional embolization with OK-432 and surgical excision[52]. Ok 432 is a lyophilized mixture of group A streptococci and streptococcus pyogens. It produces inflammatory reaction followed by gradual regression of the lesion. Sometimes surgical management needs staged resection[52].

Endovascular management of vertebrovertebral arteriovenous fistula:

The goal of treatment should be occlusion of the fistula site, and preservation of the patency of the vertebral artery[53]. These lesions are difficult to treat by surgical means, because of the anatomic location, the critical condition of the patient especially in the cases with hematoma, and the difficulty in localizing the exact site of the fistula. Endovascular intervention has been increasingly used to treat AV fistulas. If the contralateral vertebral artery can supply sufficient vertebrobasiler circulation despite the steal effect, transarterial occlusion of the affected vertebral artery with detachable balloons or coils can be an effective way of treatment. For the complete elimination of the fistula, the embolic material should be placed both proximal and distal to the fistula site,

in order to prevent the retrograde filling of the fistula[54]. To preserve the vertebral artery perfusion, not the parent artery, but the fistula site itself can also be selectively embolized with coils or balloons. On the other hand, both coils and balloons have some disadvantages. Coils may not produce occlusion of the fistula, because of their poor thrombogenicity and the difficulty in achieving dens coil packing. Also they may migrate intracranially causing inadvertent arterial occlusion, or flow through the draining veins, because of the high flow fistula. In such cases, balloon aided coil embolization can be applied, in order to prevent coil migration and achieve a dense coil packing. Sometimes it is impossible to pass a balloon through the narrow orifice of the fistula. The balloon, on the other hand, is a flow guided device and, in the case of a large bore high flow fistula, it is hard to pass the balloon distal to the fistula site for the parent artery occlusion. Sealing of the fistula with a stent graft is the treatment of choice to preserve the vertebral artery[55].
Surgical treatment consists of resection or trapping of the fistula. It is difficult and dangerous due to the proximity of surrounding neurovascular structures, which often renders a surgical approach impossible. However, at surgery the fistulous communication should be correctly localized and excluded to avoid blockage of the endovascular route for future interventions in case of recurrence[55].

CHOICE OF EMBOLIZATION MATERIAL:

Choice of embolization material depends on lesion

1. Angioarchitecture,
2. Catheter positioning,
3. Adjoining dangerous anastomosis and last but not the least the
4. Experience of neurointerventional radiologist.

The most effective treatment for high flow lesions is transarterial embolization, with occasional subsequent surgical resection[36,37]. For preoperative embolization, temporary occlusive agents such as gelfoam powder, PVA particles, and embospheres can be used. These agents very effectively reduce vascular inflow to AVMs, allowing for significantly lower rates of intraoperative blood loss. PVA and embospheres are available with different particulate diameters, allowing for initial targeting of distal arteriolar branches with small sizes, followed by occlusion of larger, more proximal branches with larger sizes. However, these agents are removed within weeks by phagocytosis, resulting in short term revascularization[46]. Thus, for embolization that is not to be followed by resection, permanent liquid agents capable of permeating the AVM nidus, such as absolute ethanol, n-BCA, or the more recently available Onyx, may be used[47].

The cornerstone treatment of low flow vascular malformations is percutaneous sclerotherapy[34,35]. Percutaneous sclerotherapy is not effective for high flow lesions since the infused agents are rapidly washed away from the endothelial lining.

MATERIALS AND METHODS

We retrospectively reviewed 32 patients with clinical diagnosis and imaging evidence of cervicofacial vascular malformations from Jan 2009 to July 2011. Youngest patient was 1 year old girl and oldest was 57 year old man, mean age was 29.4 . The male to female ratio was 1.6 :1. Patients suspected to have vascular malformation in head, neck and face region on clinical examination were referred from plastic surgery, otorhinolaryngology, ophthalmology and neurosurgery departments.

Inclusion criteria:

This study included only patients who had been diagnosed with vascular lesions based on clinical and radiological evaluations, i.e. clinical presentation and a physical examination in addition to selective arteriogram and/or direct puncture venogram findings and/or from a pathology specimen.

Criteria of exclusion:

Patients who had a head or neck mass who were referred for evaluation of the vascularity of the mass and found to have neoplastic lesion rather than vascular malformation.

All 32 patients were evaluated with either CT or MRI scan along with DSA. Twenty one patients underwent endovascular therapy either transarterial or percutaneous embolization. In rest 11 patients endovascular intervention was deferred either due to very low flow flow angioarchitecture (eliminating need for embolization)or due to deep location not accessible for direct /percutaneous approach.

Clinical history in detail was obtained from all the patients with special reference to chronology of lesion growth. All patients were well evaluated clinically. Lesion size, shape, consistency, tenderness, compressibility, pulsatility were noted. Any palpable or audible bruit was noted. Overlying skin was properly inspected for any e/o ulceration/ signs of inflammation. Fixity of the lesion to overlying skin/ underlying bone was evaluated.

All patients were evaluated with either CT or MR imaging with contrast before they were taken for diagnostic angiogram. Additional imaging was performed when the clinical scenario dictated; including MR angiography. For superficial lesions ultrasound Doppler was done. Imaging characteristics like site, size, extension, presence of phleboliths, calcification, pattern of contrast enhancement, AV shunting, feeding artery/ draining vein anatomy and aneurysms were noted. On Doppler any peak systolic velocities, low resistance /high resistance waveforms, AV shunting, AV fistulas were noted.

Informed consent was obtained from the patient/ close relatives or family members before any intervention. The procedure and the risks of all the possible complications were well explained. All patients received premedication by intramuscular injection of Tramadol 50mg and and Phenargan 25 mg, 30 min before the procedure. Once the puncture site was chosen and marked, the skin was prepared with Povidone-Iodine solution and draped. All procedures were performed in dedicated angiography suite (Advantx LCV, GE Medical Systems, USA- single plane and Innova 3131, GE medical system, USA- biplane). Local anaesthesia was administered by means of superficial administration of 1% lignocaine. All patients received intra-arterial heparin approx. 50-80 IU/kg body weight after arterial puncture followed by 20-30 IU/kg hourly. Complete 6 vessel cerebral angiogram was done for all patients which included bilateral ICA, ECA and vertebral injections. Whenever needed superselective catheterization of ECA feeders were done using either 4F vertebral glide catheter (Terumo, Terumo Medical Corporation, Somerset, NJ,

USA) or microcatheter and angiograms obtained. Angiograms were carefully reviewed with special reference to angiographic architecture, vascularity, AV shunting, dilatation and tortuosity of feeders. Dangerous ECA –ICA anatosmotic channels were carefully looked for before taking decision regarding embolization.

Choice of embolization material was mainly dependent on angioarchitecture of the lesion, degree of AV shunting, the proximity of catheter tip to the nidus i.e superselelctive cannulation and clinical presentation. Whenever needed additional imaging modality like ultrasound and Doppler was used as guidance during the procedure.

After the procedure patients were referred back to either plastic surgery or neurovascular surgery for definitive surgical treatment. Follow up was taken on clinical as well as by Doppler imaging and occasionally by CT/MRI. Angiographic follow up was not obtained in any of the patients.

RESULTS

This is a prospective and retrospective imaging analysis of 32 patients of cervicofacial vascular malformation and endovascular management.

Clinical presentations:

Seventeen patients presented with cosmetic deformity while 6 patients presented with local pain and 9 patients presented with functional impairment due to mass effect of the lesion. One child with orbital capillary hemangioma presented with congestive cardiac failure and one patient with cirsoid aneurysm presented with active severe bleeding.

Imaging analysis:

In this study 14 patients had low flow vascular malformation while 17 patients were diagnosed with high flow malformation and one patient had scalp aneurysm.

Three cases are of cirsoid aneurysm, two were post-traumatic while one was spontaneous. All the cases show hypertrophied and tortuous feeders from branches of superficial temporal artery with AV shunting and venous dilatations (Figure 1, 2).

One case of capillary orbital hemangioma in a one year old girl presented with congestive cardiac failure shows densely enhancing lesion involving intraconal as well as extraconal regions with extension in preseptal space. No obvious phleboliths were noted within this lesion (Figure 3).

All the orbital cavernous hemangiomas in this series (five cases) are intraconal in location in middle aged adults, four were females. All patients presented with gradual painless proptosis and diplopia. On imaging these lesions were encapsulated, limited to intraconal space and all lesions showed progressive accumulation of contrast material in the lesion on delayed images. Two cases showed tiny calcific specks probably phleboliths.

Three patients had vascular malformations involving tongue (Figure 5, 6); two had mixed low flow- lymphovenous malformation (cases 13,19) and one had high flow hemangioma (case 2). All the three patients presented with dysarthria due to bulky tongue.Two patients with high flow arteriovenous malformations presented with cosmetic deformity.

Three cases of vertebrovertebral arteriovenous fistulas were seen (cases 15, 29 and 32). Two patients were of Neurofibromatosis I and had spontaneous fistulas while one had iatrogenic fistula probably created during attempted central venous puncture for procedure of permanent pacemaker implantation. All the three patients presented with neck pain and palpable bruit in neck. Visible pulsatile swelling was present in two patients. One patient also had radiating neck pain radiating to ipsilateral upper limb, probably due to enlarged venous pouches causing neuronal compression. Fistulous communication of main vertebral artery with epidural venous plexus noted in all three patients making it 'intersegmental' type (Figure 7, 8). In spontaneous V-V arteriovenous fistulas, fistula was also fed by ipsilateral ECA branches- occipital and posterior auricular branches and proximal vertebral artery showed dysplasia.

Endovascular management:

In this study of 32 patients, 21 patients underwent endovascular management while in 11 patients no intervention was done. Five among these 11 patients were advised surgery, only three underwent surgery while two patients were lost to follow up.

Transarterial embolization:

Among 21 patients who underwent endovascular management, 13 patients underwent transarterial embolization, and six among these underwent surgery after embolization, while 7 patients were managed with transarterial embolization alone. One patient received both transarterial PVA followed by percutaneous bleomyecin injection. One patient in this group was lost to follow up after transarterial embolization.

Two patients of high flow arteriovenous malformation involving pinna were embolised with PVA and gel foam followed by surgical excision of the lesion. One patient underwent partial excision, he was advised for another sitting of embolization for residual AVM, however he is lost to follow up.

Total 11 patients were treated with transarterial PVA with or without gelfoam, while 5 patients glue was injected.

Two patients with spontaneous vertebrovertebral (VV) arteriovenous fistula was embolized with coils while one patient with iatrogenic vertebrovertebral fistula was treated with two overlapping stents- wallstent prosthesis. Balloon occlusion test was not done in any of these three cases as the other side vertebral artery was providing good intracranial supply.

Three cases of tongue malformation were found; two were venolymphatic malformations and one was a case of high flow hemangioma. Presurgical embolization with particles was done in case of tongue hemangioma, and patient underwent surgical excision afterwards. One case of capillary lymphovenous malformation showed very minimal blush, which was embolized with PVA particles.

Percutaneous embolization:

Total seven patients underwent percutaneous embolization, one child of capillary hemangioma with percutaneous bleomyecin. One child with orbital and periorbital capillary hemangioma was treated with transarterial PVA embolization followed by percutaneous bleomyecin injection, indication for repeat embolization with bleomyecine was refractory congestive cardiac failure. Two patients underwent STS injection and four, percutaneous glue injection. Indication for percutaneous STS injections was superficial venolymphatic malformation in both the cases. The rate of blood flow (flow velocity) in the nidus was checked before injection of sclerosant. Direct puncture in the nidus with 18 to 22 gauge needle was done. Care was taken to inject the agent gradually so as to overflow or overdose into the draining vein.

Three patients with cirsoid aneurysm and one case of scalp aneurysm were treated with percutaneous glue injection. Tourniquet was used in all the cases of cirsoid aneurysm, which was tied just below the aneurysm along the forehead. Aim was to prevent passage of embolizing material on venous side. All the glue injections were done under fluoroscopy guidance with 18 G scalp vein needle. Initially approximately 2cc of 5% Dextrose was injected through the scalp vein needle followed by controlled injection of NBCA under fluoroscopic guidance were done.

Postprocedure care and Complications:

All patients with percutaneous embolization using STS were kept on analgesics postprocedure for 24 hours. Injectable antibiotic was not routinely given. Two patients had immediate postprocedure fever which was managed symptomatically with injectable antibiotics and antipyretics. One patient with cirsoid aneurysm which was treated on emergency basis with percutaneous

glue injection had moderate local pain postprocedure, which managed conservatively with analgesics.
No major complications were noted in immediate postprocedure period. No patient developed puncture site/ procedure related vascular complications.

Follow up and recurrence:

Two patients required repeat endovascular management because of recurrence of symptoms, one was large neck- occipital region AVM, which was managed twice with transarterial particle embolization and other was one year old female child with orbital and preseptal capillary hemangioma who was initially treated with transarterial particle embolization but because or refractory congestive cardiac failure, was again taken for tranarterial PVA + percutaneous bleomyecine injection. Her cardiac function was improved postsecond embolization. Four patients were lost to follow up.

DISCUSSION

Peripheral vascular malformations are some of the most difficult lesions to diagnose and treat. Overall prevalence of vascular malformations is estimated to be 1.5% of the general population[1]. The head and neck are quite rare and their true incidence is unknown. There may be a slightly higher prevalence in girls, with at least one series reporting a female-to-male ratio of 1.5:1.[7] Vascular malformations occur as a result of aberrant vessel angiogenesis. They are localized or generalized congenital vascular abnormalities comprising direct microscopic connections between arteries, veins, and lymphatic vessels without the normal capillary bed[7]. Vascular malformations are divided in to various sub-categories depending on the predominant anomalous channels—such as venous, lymphatic, capillary, and arterial malformations. Mixed vascular anomalies are common such as capillary–venous, lymphatic–venous or arteriovenous malformations[8].More simply, malformations can be categorized as either low-flow or high-flow lesions on the basis of their hemodynamic flow characteristics. Capillary, lymphatic, and venous malformations are classified as low-flow malformations, whereas any malformation with an arterial component is classified as a high-flow malformation (arteriovenous malformations and arteriovenous fistulas)[9]

The differentiation of vascular malformation into high and low flow is utmost clinical importance, as treatment options are drastically different. The cornerstone treatment of low flow vascular malformations is percutaneous sclerotherapy[34,35]. Percutaneous sclerotherapy is not effective for high flow lesions since the infused agents are rapidly washed away from the endothelial lining. The most effective treatment for high flow lesions is transarterial embolization, with occasional subsequent surgical resection[36,37].

We had nine cases of **orbital vascular malformation**; two had low flow-venolymphatic malformation, one had orbital AVM while four were hemangiomas. All the patients presented with painless gradual proptosis except one child with orbital capillary hemangioma presented with congestive cardiac failure. Cavernous hemangiomas were presurgically embolised with PVA particles. Capillary hemangioma usually do not need intervention as majority of these lesions resolve spontaneously as suggested by natural course. However, sometimes symptomatic lesions may need intervention. One case of orbital capillary hemangioma in our study was treated with percutaneous bleomycin injection. The potential beneficial effects of intralesional bleomycin injection (IBI) in the treatment of haemangiomas were initially reported by Kullendorf and Sarihan et al.[56,57] then the effectiveness of IBI was evaluated in 37 patients with haemangiomas in a study conducted by the Pretoria Vascular Malformation Study Group. Complete resolution or significant improvement was seen in 87% of the patients[58].

Excessive angiogenesis is considered a central event underlying haemangioma development. In addition, proangiogenic growth factors VEGF and bFGF were previously detected in proliferating lesions.[59] Bleomycin inhibits haemangioma growth by inhibiting angiogenesis.

One patient with orbital AVM presented with mono ocular diplopia and proptosis. ECA feeders were embolised with PVA particles and then patient underwent surgical excision. Regression is well documented in cerebral AVMs but has not been reported in orbital lesions. The primary treatment for orbital AVMs is surgical excision with or without preoperative embolization. Review of the literature shows that recurrences were reported when incomplete excision or partial embolization alone was performed highlighting the tendency of these lesions to recruit new feeder vessels when their supply is partially reduced. In our case complete surgical excision was done with no clinical recurrence on 8 months follow up.

Among 9 cases of orbital vascular malformation five were cavernous hemangiomas, It most frequently occurs in young to middle-aged females and

is characterized by slowly progressive and painless proptosis. These lesions are almost always intraconal, with extraconal malformations being uncommon. All the five orbital cavernous hemagioma cases in this study are intraconal in location in middle aged patients. Two patients underwent surgical excision of intraconal hemangioma due to visual disturbances; one of these patients underwent presurgical embolization with PVA. No endovascular intervention was done in rest three cases and were managed conservatively. There was no worsening in clinical symptoms in two cases on follow up, while one patient was lost to follow up.

Table 8 : Orbital vascular malformations

Age	Sex	Type	Diagnosis	Presentation	Management	Embolisation agent
1	F	high flow	capillary malformation	congestive cardiac failure	transarterial embolization+ Percutaneous bleomyecin	PVA and bleomyecin
9	M	High flow	Cavernous hemangioma	proptosis	transarterial embolization	PVA
13	F	high flow	AV malformation	proptosis and diplopia	transarterial embolization	PVA
25	M	low flow	venolymphatic malformation	proptosis and diplopia	no intervention- referred for surgery	Embolization not done
29	M	low flow	Cavernous hemangioma	proptosis and diplopia	managed conservatively	Embolization not done
38	M	low flow	Cavernous hemangioma	proptosis	no intervention- referred for surgery	Embolization not done
39	M	low flow	venolymphatic malformation	diplopia	managed conservatively	Embolization not done
33	F	low flow	Cavernous hemangioma	painless proptosis	no intervention- referred for surgery	Embolization not done
51	F	low flow	Cavernous hemangioma	painless proptosis	managed conservatively	Embolization not done

Cirsoid aneurysms are rare arteriovenous fistulas of the scalp. They are usually congenital in aetiology. However, traumatic fistulas have also been described. They are called “cirsoid” because of the characteristic variceal

dilatation of the draining veins. The cirsoid aneurysm comes to be composed essentially of two parts, the fistula itself and the dilated afferent and efferent vessels.We found two cases of cirsoid aneurysm, two of them presented with prior history of trauma, and other with spontaneous swelling. On patient was taken on emergency basis with indication of severe acute bleeding from the prediagnosed cirsoid aneurysm. In 90% of patients, the superficial temporal artery is the main supply to the fistula with only one dominant feeding artery in 71% of patient[25]. In all three cases the feeding artery was superficial temporal branch. Transarterial approach may not be successful always because of distal location of fistula and multiple tiny distal feeders. Another problem with an arterial approach is that there is recruitment of surrounding normal arteries following occlusion of the arterial feeder and draining venous structures[60]. Both the cases were treated with percutaneous liquid embolization agents.

We found three cases of **tongue vascular malformation**; two were venolymphatic malfornationms and one was a case of high flow hemangioma. Presurgical embolization with particles was done in case of tongue hemangioma, and patient underwent surgical excision afterwards. One case of capillary lymphovenous malformation shows very minimal blush, which was embolized with two bolus of 250-355 micron PVA particles and later patient underwent surgical excision in department of plastic surgery.

Two cases of **pinna vascular malformation** was seen, both had high flow arteriovenous malformation and underwent presurgical particle + gelfoam embolization. One of them had partial excision of AVM as lesion was extending in adjoining soft tissue, he was adviced repeat embolization for residual lesion, as there is high rate of recurrence in these partially excised lesions as suggested by natural history.

Three cases of **vertebrovertebral (VV) arteriovenous fistulas** were seen. Beaujeux et al[53] reported that, most of the traumatic fistulas affect the lower portion of the vertebral artery (below C5), while spontaneous ones involve the upper portion (at or above C2). Among our three cases two had spontaneous V-V arteriovenous fistulas both at C3, C4 level while one patient who had iatrogenic V-V arteriovenous fistula was located at C7 level.

V V fistulas can be classified either as segmental and intersegmental types or as upper and lower cervical groups. In the segmental type, the fistula involves the VA branch, whereas in the intersegmental type, the VA itself is involved. In our study, fistulous communication of main vertebral artery with epidural venous plexus was noted in all three patients making it 'intersegmental' type. The upper cervical group usually seen in the paediatric population includes a fistula that has developed at the C1–2 level from the proatlantal system. Lower cervical fistulas involve cervical arteries and can be seen in adults with underlying dysplastic vessel wall disease. All three patients here showed lower cervical fistulas.

Table 9: Vertebrovertebral arteriovenous fistulas

No	Age	Sex	Presentation	Feeders	Type	Management	Follow up
	34	F	neck pain, k/c/o NF1	Vertebral artery, ECA branches	Lower cervical , intersegmental	transarterial embolization with coils	partial resolution
29	29	M	neck pain and swelling, k/c/o NF1	Vertebral artery, ECA branches	Lower cervical , intersegmental	transarterial embolization with coils	partial resolution
32	45	M	postsurgical swelling	vertebral artery	Lower cervical , intersegmental	transarterial stent placement	partial resolution

Various endovascular techniques are available for the management of VVFs. Commonly described techniques include transarterial use of detachable balloons,[20,53]transarterial placement of detachable coils at the fistulous site with or without trapping of the parent vessel,[54,55] and placement of a covered stent in the feeding artery[22]. Other than these aforementioned transarterial approaches, adjunctive transvenous2 and direct percutaneous embolization has been used to close VVFs. Both the spontaneous V-V fistulas were occluded by transarterial detachable coil embolization with trapping of parent vessel. Balloon occlusion was not done as contra lateral vertebral artery was adequate for posterior circulation and involved vertebral artery showed dysplastic changes. Stent grafts are said to be superior as parent artery is preserved. In third case of iatrogenic V-V arteriovenous fistula where vertebral artery was normal and not dysplastic, stent was placed to preserve antegrade flow through vertebral artery.

CONCLUSIONS:

Vascular malformations represent some of the most difficult challenges in the field of interventional radiology. To assess the extent of vascular malformations and flow velocity, CT, Doppler US, and MR imaging are useful. Angiography is useful to confirm the diagnosis, evaluate the extent and dynamics of the lesion. Patients present because of pain, cosmetic concerns, or because of symptoms related to mass effect of the malformation on vital structures. The latter is particularly true in the head and neck, given the concentration of vital structures in very contained anatomic regions. There is an ever-expanding role of interventional radiology in the wide range of pathology in the head and neck. There are potentially dangerous anastomoses between the extracranial and intracranial circulations, which should be sought for actively on prembolisation angiograms to minimize the risk of cranial nerve palsies, blindness, or stroke.

Our approach to workup and treatment is intrinsically multidisciplinary, with interventional and surgical. Despite the complexity and rarity of these types of cases, most patients can be significantly helped through this kind of multifaceted intervention.

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