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PROJECT REPORT

Name : **DR.MUTHU RETNAM.T**

Programme : **M.Ch.NEUROSURGERY**

Month and year of submission : **NOVEMBER -2002**

PROJECT REPORT

Title of the project:

A RETROSPECTIVE STUDY ON RARE CEREBELLOPONTINE ANGLE LESIONS

Name : **DR.MUTHU RETNAM .T**

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CERTIFICATE

I, **Dr. Muthu Retnam T** hereby declare that I have actually performed all the procedures listed/carried out in the project under report.

Place: Thiruvananthapuram
Date: 1st November 2002.

Signature

(MUTHU RETNAM. T)
Name in capital letters

Forwarded. He has carried out the minimum requirement of procedures / etc.

Signature

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INTRODUCTION

The cerebellopontine angle is a region of highly intricate anatomy. Surgical mastery in this zone requires great patience, vast experience and meticulous technique on the part of the surgeon. A Surgeon operating routinely on lesions of the Cerebellopontine angle encounters schwannoma, meningioma and epidermoids on a regular basis. Quite often, there are a lot of surprises awaiting. During a neurosurgical carrier, several times one may come across these unlikely lesions in this region confusing surgical decision-making. This is true even for the most accomplished and daring surgeons.

We have collected and analyzed the experience of our institute, a tertiary care center, while operating in this zone of vast surgical importance. It is hoped that this will be of interest to surgeons operating in the Cerebellopontine angle and might help in tiding over moments of surgical dilemma.

Cerebellopontine angle tumours account for about 5-10% of all intracranial tumours. The native structures of the cerebellopontine angle are the source of two expressions of neoplasia such as schwannoma and meningioma as well as few space-occupying lesions, such as the arachnoid cyst, giant aneurysms and AVMs. The extraneous products of the embryonic development contribute alien tissue to the Cerebellopontine angle, from where other rare tumours take origin such as dermoid and epidermoid cysts.

Anatomical structures that are contiguous to the cerebellopontine angle also spawn a variety of neoplasms that impinge upon, disfigure and occasionally occupy the angle. Notable among these neoplasms are the pontine and cerebellar gliomas, cerebellar

haemangioblastoma, choroid plexus papilloma and carcinoma, ependymoma and a variety of lesions nascent in the base of skull like glomus jugulare, metastasis, chordoma and nasopharyngeal carcinoma (Schmincke`s tumour).

Little is known about these rare lesions. This study aims at identifying the not so usual neoplasms of the cerebellopontine angle and analyze their clinical behavior, management and outcome.

AIMS AND OBJECTIVES

The aims and objectives are:

1. To study the incidence of rare lesions among the total cerebellopontine angle tumours.
2. To analyze the clinical presentation and the imaging characteristics of these cerebellopontine angle lesions.
3. To assess the outcome of surgical resection, adjuvant treatment and prognosis.

MATERIALS AND METHODS

A retrospective review of all cerebellopontine angle tumours operated at Sree Chitra Tirunal Institute for Medical sciences and Technology during a 10-year period between 1990-1999 were undertaken. The unusual neoplasms at this site (tumours other than acoustic schwannoma, meningioma and epidermoids) were identified . The clinical presentation, imaging characteristics, surgical approaches, post-operative complications, histopathological studies and outcome of 37 operated cases of these rare lesions were analysed.

All these patients underwent detailed clinical examination following admission. Computerised tomography with and without contrast were done in all cases. Eighteen patients underwent magnetic resonance imaging. A few patients suspected to harbour vascular lesions at the cerebellopontine angle were subjected to cerebral angiography.

The most common surgical approach adopted for accessing these lesions was the retromastoid route. A few cases were operated by combined approaches. Serial neurological assessments were performed post-operatively. Based on the histopathology reports, the patients were referred for adjuvant treatment if necessary. A post-operative neuroimaging was performed using computerised tomography to look for the extent of removal and other effects secondary to the surgery. All the patients were followed-up at 6 monthly intervals and neuraxial imaging was repeated in patients suspected to have local or distal recurrence of tumour.

REVIEW OF LITERATURE

Cerebellopontine angle is an area of difficult surgical anatomy and the tumours of this region are relatively common. Although the commonest tumour in this region is acoustic schwannoma, non-acoustic lesions form an interesting group of unusual cerebellopontine angle tumours. In the study by Moffat et al, among 305 cerebellopontine angle tumours treated at Addenbrooke`s hospital, 248 were acoustic schwannomas (80.7%) and there were 59 non acoustic tumours [1].

Unusual CP angle tumours (305 cases treated at Addenbrooke`s hospital)

S.no	Tumours	Incidence (%)
1	Meningioma	6.5%
2	Primary cholesteatoma	4.6%
3	Glomus jugulare	8.6%
4	Facial neuroma	5%
5	Trigeminal neuroma	5%
6	Arachnoid cyst	3.5%
7	Giant cell tumours	3.5%
8	Metastasis	3.5%
9	Other cranial Nerve neurinomas	3.5%

ARACHNOID CYST OF CEREBELLOPONTINE ANGLE:

Arachnoid cysts are rare congenital malformations that are thought to originate from splitting or duplication of the arachnoid membrane due to alteration of the CSF flow or pressure changes from trauma or infection.

Approximately 1% of all intracranial space occupying lesions seems to be arachnoid cysts. They are most often observed in the temporal fossa [2]. The cerebellopontine angle is the second most common location with approximately 10% frequency[3]. In most cases, these congenital lesions are found in adulthood. Males and females are equally affected. Rengachary and Watanabe analyzed 208 cases of arachnoid cysts reported in the literature from 1931-1980 and found 22 cysts have been located in the cerebellopontine angle.

The clinical syndrome produced by a cyst in the Cerebellopontine angle may closely mimic that of an acoustic neuroma with sensorineural hearing loss, cerebellar signs and less frequently fifth and seventh cranial nerve as well as pyramidal deficits [4]. Papilloedema is often observed even in patients without hydrocephalus. This has been explained by the possible impairment of CSF flow within the basal cisterns. Rarely paroxysmal shooting pain in the second and third divisions of fifth nerve and hemifacial spasm has been reported with Cerebellopontine angle arachnoid cyst [4]. Rarely arachnoid cysts in adults may remain confined within the internal auditory canal and causes widening of the canal and facial nerve palsy [5]. Although in most patients, the clinical history prior to diagnosis is relatively short, in some patients skull x-ray examination may show alteration of the overlying bone, which is indicative of long-standing lesion.

Prior to the introduction of the CT, diagnosis was nearly always made during surgery for suspected acoustic neurinoma. CT and MRI are currently the diagnostic techniques with which an arachnoid cyst of the CP angle can be readily recognized. The uncommon association of the CP angle arachnoid cyst with chiari type I malformation is explained by the gradual tonsillar displacement as a result of prolonged compression [3].

Operative management involves posterior fossa craniectomy by the paramedian approach and wide excision of the cyst wall. However, although cerebellar and pyramidal deficits usually regress after surgery, cochleovestibular dysfunction improves only rarely. In patients presenting with hemifacial spasm, it is necessary to decompress the nerve root exit zone from arterial compression for complete relief of hemifacial spasm [4].

of CP angle and late occurrence of intracranial hypertension with CP angle choroid plexus papilloma in contrast to early intracranial hypertension with intraventricular choroid plexus papilloma due to the obstruction of CSF pathways. CP angle choroid plexus papillomas are associated with early and insidious involvement of hearing mimicking acoustic neurinoma.

***Radiological features*:**

On CT scan, choroid plexus papillomas appear as an isodense homogenous enhancing tumour [8]. Macroscopic cysts have occasionally been observed in 20% of cases. Typically absence of bony changes in the region of internal auditory meatus (IAM) will favor the diagnosis of choroid plexus papilloma. However, some of the choroid plexus papilloma have been found to enlarge IAM and involve petrous bone [9]. Homogenous contrast enhancement will also be seen in CP angle meningioma and schwannoma.

In MRI scan, on TIWI, Choroid plexus papillomas are shown as hypointense or isointense with greymatter, whereas on T2WI they have variable signal. The presence of signal voids on T1WI and T2WI corresponds to rich vascular supply, small calcification or cysts. Because the signal characteristics are very similar, the differential diagnosis with meningioma is difficult, but the absence of broad attachment to the bone and the absence of `tail sign` are helpful in diagnosis. In cases of tumours with macroscopic cysts, differential diagnosis of haemangioblastoma can be considered. Thrombosed giant aneurysms have typical concentric calcification that do not enhance with Gd-DTPA [10]. Vertebral angiography has been used in the past and enlarged AICA with dilated branches has been considered to establish the diagnosis of Choroid plexus papilloma conclusively [11]. However at present angiography is not routinely used for diagnosis of CP angle masses.

Diagnosis of CP angle choroid plexus papilloma should be confirmed by histopathology. However, at times it may be confused with papillary ependymoma. Choroid plexus is distinguished by the delicate vascular stroma covered by a single layer of columnar epithelium in contrast to the frequent arrangement of cells in multiple layers, additional formation of tubules and presence of neuroglial stroma. Positivity for GFAP may be occasionally expressed by Choroid plexus papilloma as well, therefore immunochemistry may not be helpful in their differentiation (Russel and Rubenstein 1989).

Radical excision must be attempted except when the tumour is adherent to the brainstem. In such cases, radiotherapy is recommended (Chan etal 1983). On microsurgical excision, tumour will be found attached to the rootlets of the lower cranial nerves resembling neurofibroma [12]. The morbidity and mortality of surgical treatment of Choroid plexus papilloma has drastically decreased in thelast10-15 years to 8.6% and 4.3% respectively[13].

MEDULLOBLASTOMA OF CEREBELLOPONTINE ANGLE:

Medulloblastoma can have very variable clinicoradiological pattern and biological behaviour with occurrence at unusual sites. The Cerebellopontine angle is one such location and about 15 cases have been reported in the world literature[14]. Unfortunately, there are no definite clinical, neuro-otological or neuro-radiological findings peculiar to CP angle medulloblastoma which would help to differentiate this from other CP angle tumours[15].

The diminution of hearing or seventh nerve involvement is uncommon in medulloblastoma when compared to acoustic neurinoma of same size. Only 2 of the 15 reported cases presented with diminution of hearing as the initial symptom. Early onset of progressive cerebellar signs and gait ataxia may indicate an axial origin of the tumour, while positional nystagmus may be an early sign suggestive of medulloblastoma[16].

The site of origin of medulloblastoma in the CP angle is debatable, the possible sites of origin include,

1. Remnants of external granular layer of cerebellar hemisphere, mainly the flocculus that faces the CP angle[14].
2. Proliferating residue of the lateral medullary velum from where it may project into the CP angle[17].

Surgery via the retromastoid route followed by radiotherapy is the main treatment modality and 30% 5-year survival rate has been reported in CP angle medulloblastoma after surgery and radiotherapy[18]. Vincristine and Cisplatin based chemotherapy have also been combined with radiotherapy with improved survival [19]. In general, medulloblastomas of CP angle have a higher malignant potential than their vermian counterpart.

CEREBELLOPONTINE ANGLE PARAGANGLIOMA:

Paragangliomas are rare neoplasms arising from the extra-adrenal paraganglionic system cells of neural crest origin, which have chemoreceptor function. Typically intracranial paragangliomas are direct extension of tumour arising extracranially in the jugular fossa or

middle ear. In one of the large series about 15-17% of jugulotympanic paragangliomas grew intracranially. Tumours originating in other intracranial sites are exceptionally rare and so far only 4 cases have been reported from petrous ridge [20], pineal region [21] and sella turcica and parasellar region [22].

Paraganglioma presenting as a CP angle tumour may not be a very unusual occurrence, as jugulotympanic paraganglioma infrequently grow through the round window into the IAM and finally reach the CP angle cistern and is mistaken for acoustic neurinoma [23]. However, erosion of the floor of the IAM and inframeatal component of petrous bone, together with the presence of tumour in the jugular foramen excludes the diagnosis of acoustic neuroma. In addition, they possess intense vascularity with feeders from external carotid [24]. However, experience with angiography in paragangliomas of intracranial locations other than the jugulotympanic area is lacking and are probably less vascular than their jugulotympanic counterparts.

The unique origin in the CP angle is due to the presence of aberrant chemoreceptor cell nests present along the vessels and nerves within the IAM [25]. The tumour including the intrameatal part can be removed completely through suboccipital craniectomy and it is very difficult to isolate and preserve seventh and eighth nerves from the tumour capsule.

CAVERNOUS MALFORMATION OF THE CEREBELLOPONTINE ANGLE:

Cavernous malformations are well-circumscribed vascular malformations composed of dense cluster of honeycomb like, thin walled vascular channels without

intervening neural parenchyma. They comprise 10-15% of both intracranial and spinal vascular malformations [26]. Infratentorial location account for 25% of intracranial lesions and most of them arise from cerebellum and brainstem. Extra-axial location is noticed in 13% with the majority arising from the middle fossa and cavernous sinus [27]. About 16 cases of cavernoma involving IAM and 5 cases of CP angle lesions were reported in the literature so far [28,29]. Three of them had a cystic component and two were solid. Steiger etal reported that the intraparenchymal cystic form is particularly prone to growth and frequently present with progressive neurological deficit [30].

Precontrast CT may show a lesion which is hyperdense or of mixed density, with improved delineation and faint enhancement with contrast. Sensitivity of MRI to detect parenchymal cavernous malformation is very high, however in extraparenchymal locations, the findings are non-specific. The solid component was isointense in both T1WI and T2WI as reported by Kim etal. Variable enhancement with gadolinium depends on the degree of thrombosis. Brunori et al [77] found intense enhancement where as Kim et al did not find any enhancement. Lesional haemorhage was found in only one case.

Total excision if possible is the treatment of choice. Extra-axial cavernomas are associated with higher perioperative morbidity and mortality due to incomplete encapsulation and proximity to the sinuses eg; cavernous sinus. Total excision of the cyst wall may not be feasible in all cases due to the presence of adhesions to the vital structures in the CP angle. However, unless the cyst wall is carefully searched for and the solid component is totally excised, the treatment is likely to be incomplete [27].

CEREBELLOPONTINE ANGLE LIPOMA:

Intracranial lipomas are rare tumours that develop from the mesenchymatous cells and presumably are caused by aberrant differentiation of primitive meningeal matter instead of by a neoplastic process. The incidence of intracranial lipoma is 0.08% in autopsy series and is localized in the supratentorial part in 81% and on the midline in 82% of cases. The corpus callosum is the most frequent location and is rarely present in the CP angle and IAM (0.14% of tumours). According to the literature, about 98 cases have been reported since the first description by Kolb in 1859. In 1974, Budka performed the first surgical resection of a CP angle lipoma [31].

The tumour size ranged from 1 to 26 mm (average 11.2 mm). The sex ratio was a nearly 2 to 1 with predominance of men (64.5%) to women (35.5%). Tumour localization was 59.9% on the left side, 37% on the right side, and 3.1% bilateral. The patients age at discovery of tumours ranged from 7 months to 82 years (average age 40 years).

The evolution of CPA lipoma is quite different from that of other lipomas and they typically entrap cranial nerves and induce progressive focal neurological deficits. Patients with CP angle lipomas have multiple symptoms according to the involvement of neighboring structures. Clinical signs can last for years and include cranial nerve irritation, but not brain compression. Cochleovestibular symptoms are the most frequent, but facial and trigeminal signs can occur very rarely. Facial nerve symptoms such as hemifacial

spasm are the first signs in 9% of cases. Trigeminal signs such as paresthesia or more often neuralgia can be the first symptom in 14.4. % of cases.

Presenting symptoms of patients with CPA lipoma

Reported symptoms	*No.of pts (n=90)*	*%*
Hearing loss	56	62.2%
Dizziness	39	43.3%
Tinnitus	38	42.2%
Headache	14	15.5%
Trigeminal symptoms	13	14.4%
Facial palsy	8	8.9%
Ear pain	5	5.5%
Others*	9	10%

*- includes oculomotor symptoms, dysphagia and dyskinesia.

Cranial nerve involvement in CPA lipoma

Cranial nerves	*No (n=62)*	*Percentage %*
VIII	60	96.7%
VII	51	82.2%
V	15	24.2%
IX	15	24.2%
X	13	21%
XI	7	11.3%
VI	4	6.4%
XII	1	1.6%
Brainstem	18	29%

Currently, diagnosis is made on the basis of imaging studies, particularly MRI scans. CT scanning with the fat density (-50 to –100 HU) shows a homogenous mass not modified by the contrast injection. Lipomas appear hyperintense on T1WI with progressive iso to hypointensity with increasing T2 weighting, and with a missing signal in fat suppression sequences.

Surgical treatment of CP angle and IAM lipoma is not easy, owing to the extreme frequency of cranial nerve involvement and its close relation leads to important post-operative sequelae even in patients in whom an incomplete resection or a simple biopsy is performed. According to the literature, translabyrinthine approach allows a complete resection to be performed with good control of facial nerve and CP angle in most patients with auditory sequelae. The middle and posterior fossa approaches allow for the preservation of hearing in 26% of patients even when the resection is incomplete [32]. Olson

et al reported the first case in which an IAM lipoma was dissected from the cochlear nerve with post-operative hearing preservation [33]. More recently, Zimmermann et al reported a case with conservation of hearing after complete tumour resection with a retrosigmoid approach [34]. Because of the severity and frequency of these sequelae, majority of the patients need regular follow-up [35]. The surgical indications should be restricted to,

1. Vertiginous syndromes that is refractory to medical treatment and vestibular rehabilitation. Bigelow et al[32] suggested vestibular nerve section as a treatment for vertiginous symptoms.
2. Patients with severe trigeminal neuralgia and hemifacial spasm where many authors described the efficacy of trigeminal and facial nerve decompression.

Total tumour resection is not the main target, because these tumours have a slow growth course (15% increase in tumour growth in 8 year period(Bigelow [32]) and important post-operative sequelae-22% of patients reported in the literature have experienced post-operative facial nerve palsy.

PRIMARY CEREBELLOPONTINE ANGLE MALIGNANT MELANOMA

Primary solitary intracranial melanoma is a rare tumour and its occurrence in the CP angle is extremely uncommon. Eight primary solitary CP angle malignant melanoma have been reported in the literature till 1992 [36]. One case has been reported from India [37].

Primary intracranial melanomas are either solitary or of diffuse variety. Diffuse leptomeningeal melanomas preferentially affect children and may be a part of

neurocutaneous melanosis complex or phakomas [38]. Such melanomas usually present with features of raised ICP, cranial nerve palsies and meningism. Focal melanomas present as leptomeningeal or dural based neoplasms and are more common in adults. Melanomas originate from melanocyte cell rests which are of neural crest origin, commonly distributed in the leptomeninges with a maximum density in the region of caudal medulla and high cervical cord [39]. The melanocytic tumours range between the relatively indolent melanocytomas to the aggressive malignant melanomas.

The prognosis of the patient with primary solitary intracranial melanomas depend upon the following factors:

- Degree of mitosis.
- Leptomeningeal dissemination.
- Extent of surgical excision.
- Location of tumour.

Yamane et al reported mean survival of 20.7 months in patients with primary solitary intracranial melanoma while the survival was only 6.7 months in patients with diffuse leptomeningeal spread [40]. Metastasis from primary solitary intracranial melanomas to lungs, spleen, pancreas and kidneys although rare, has been reported in the literature [41]. Metastasis from the extracranial primary melanoma is more prone to occur in the brain and is the third most common site of intracranial metastasis after carcinoma of breast and lung[36].

Intracranial melanoma is often confused with other pigmented lesions like pigmented meningioma, schwannoma, medulloblastoma, choroid plexus papilloma,

astrocytoma and pituitary tumours. Primary intracranial malignant melanoma is exceptionally rare and the other sites of primary melanoma in the body should be excluded by clinical and radiological examination. CSF examination can be positive in diffuse leptomeningeal variety and spinal melanomas [42].

Melanomas are hyperdense on CT and there is only marginal enhancement after contrast. MRI shows a hyperintense lesion on T1WI as a result of paramagnetic phenomenon exhibited by melanin due to the presence of free radicals with impaired electrons and is hypointense on T2WI [43].

Radical tumour resection and use of aggressive whole brain irradiation have been recommended. The prognosis and long term survival is longer (19.6+_ 2.3 months) in patients who had undergone a radical tumour resection and shorter (9.3+_2.4 months) in patients where only a biopsy or partial tumour excision was done [36].

TERATOMA OF THE CEREBELLOPONTINE ANGLE

Teratoma occurs most commonly in infancy and childhood (50% in newborns and infants). They account for 2% of intracranial tumours in patients under the age of 15 years [44]. Intracranial teratomas have a tendency to arise near the midline with the pineal region, suprasellar region, cerebellar vermis and lateral ventricles being the most common sites, in that order. In general infratentorial teratomas are rare. Wakai et al. found that only one of the 73 intracranial teratomas occurring in the neonatal period was infratentorial [45]. Most infratentorial teratomas occur in the cerebellar vermis and rarely in the

4th ventricle [46]. The CP angle is an unusual site of origin and only one immature teratoma has been reported in this location [44].

Teratomas are composed of tissues derived from all three germ layers. They can be mature, immature when they contain fetal tissue or malignant when they have sarcomatous or carcinomatous components or foci of malignant germ cells. They can metastasize by way of CSF.

On CT scan, mature teratomas have heterogenous density because they contain fat, soft tissue, cartilage or bone. Immature and malignant teratomas are usually homogenous in density and show marked contrast enhancement and are often confused with acoustic neurinoma or meningioma.

Total removal of the tumour is the treatment of choice for intracranial teratomas. For malignant teratomas, radiotherapy and chemotherapy are recommended.

HYDATID CYST OF CEREBELLOPONTINE ANGLE

Posterior fossa hydatid cyst is very rare and only a few case reports are available in the world literature [47]. The reported sites included pons, cerebellum [48]and 4th ventricle [49]. One case of CP angle hydatid cyst was reported [50].

The disease is more common in children and young adults. Infestation starts in childhood and manifests at the age of 15-30 years. Spinal and cerebral involvement occurs in 2-3% of cases.

Childern present with features of raised ICP and macrocrania. Papilloedema may occur and may progress to secondary optic atrophy. Blood counts may show eosinophilia and casoni`s test may be positive.

CT scan is the investigation of choice which shows a large hypodense cyst with a clearly defined margins.Cyst wall does not take much contrast enhancement.Significant ventricular compression or shift or hydrocephalus can be observed and there is no perilesional edema or mural nodule.

Treatment consists of surgical removal of the cyst intact. Accidental rupture poses possibilities of anaphylactic shock, meningitis and local recurrence. Cysts exposed by radical cortical incision is termed `dowling episiotomies`. Cyst can be delivered easily when the ICP is high. Head is lowered so that the cyst is dependent, its expulsion is facilitated by gentle saline irrigation of cleavage between brain and cyst. In the event of rupture, 3% hypertonic saline irrigation done and albendazole started at10mg/kg daily for 3 months which reaches 40% concentration in the cyst and 50% concentration in the CSF [50].

PRIMARY SQUAMOUS CELL CARCINOMA OF CP ANGLE

Cranial epithelial cysts occur in diverse locations such as the scalp, calvaria, jaw and base of brain. The most frequent sites of intracranial epithelial cysts are the CP angle and the parapontine region (constitutes 0.2-1% of all intracranial tumours). Malignant degeneration of these cysts may occur or primary squamous cell carcinomas arise de novo in the same location.

Strict selection criteria for inclusion as primary intracranial squamous cell carcinoma includes,

1) the tumour should be restricted to the intracranial, intradural compartment without invasion of or extension beyond dura or cranial bones,
2) there must be no extension or invasion through intracranial orifices,
3) no communication or connection with middle ear, air sinuses or sella turcica and
4) no evidence of a nasopharyngeal tumour.

Thirteen cases that compiled with this criteria have been described previously. In 10 cases, there was evidence of an epidermoid cyst with malignant changes and in other 3 cases the tumour was solid. Of the 13 cases, 10 were males and only 3 were females. The patients age ranged from 37 to 72 years. The duration of symptoms varied from 1 month to 13 years. In 5 patients, the symptoms lasted more than 10 years[51].

Symptoms depended on the location of the lesion and were predominantly those of brain stem compression and cranial nerve involvement. Signs of raised ICP were seen late in the course .

In two cases, the first biopsy showed epidermoid cysts, but malignant changes were seen on subsequent biopsies. Although the earlier cases were diagnosed only at autopsy, the last eight cases were diagnosed on surgical exploration.

Lewis etal[52]discovered irregular nodular rim enhancement associated with an area of malignancy on post contrast CT. The generally accepted MRI pattern of low signal intensity on TiWI and high intensity on T2WI with 2 subtypes – black and white has

been reported by Horowit etal [53]. The clinical outcome in patients with intracranial squamous carcinoma is bleaker than that of benign epidermoids which has a 20 year survival rate of 93%.Yamakawa etal reported 24% recurrence in 9 years for epidermoids and if a second recurrence was discovered, the average interval was another 12 1/2 years.Only one case of squamous carcinoma has been reported to be lived 3 years after documentation of malignancy.

DEVELOPMENTAL VENOUS ANAMALY OF CP ANGLE

DVAs, so called venous angiomas, are anatomical variants of cerebral veins, which drain normal cerebral tissue into an extra-parenchymal collector and are associated with absence of a venous pathway which would normally drain the territory [79].

They occur above or below the tentorium, with either superficial or deep drainage and are often associated with cavernous angiomas. Those located in the posterior fossa occasionally drain through the brainstem.

Apart from the majority found incidentally on neuro-radiological studies, non-specific symptoms like headache, gait ataxia, diplopia, dysphagia and symptoms caused by haemorrhage can be found. In sporadic cases, trigeminal neuralgia is caused by DVA and in 1999 Konan etal published one case of a posterior fossa DVA,which became symptomatic by spontaneous thrombosis and subsequent non-haemorhagic cerebellar infarct.

Axial MR images may show a large venous anomaly passing through the cerebellar peduncle and cerebellopontine cistern in contact with the trigeminal nerve. DSA may show a typical `caput medussae` in the cerebellar hemisphere and large collecting vein draining into the lateral mesencephalic vein.

The management not only of asymptomatic, but even of symptomatic DVAs remain controversial. Mori etal [54]compared conservative versus operative treatment among 6 patients with posterior fossa DVAs and found a satisfactory outcome in 2 operated cases. Goulao etal and Kuker etal [54] each had one case with trigeminal neuralgia among their 16 and 3 patients ,where no operation was performed.

Thus the occurrence of DVAs in the CP angle should be added to the possible causes of trigeminal neuralgia, and microvascular decompression instead of hazardous coagulation and removal of the vein is the correct treatment [79].

NEUROSARCOIDOSIS OF CP ANGLE

Sarcoidosis is a chronic multisystemic disease with the prevalence of 10-40 per 1 lakh persons in United states and a black/white ratio ranging from 10:1 to 17:1 [55].

Cranial nerve palsies and meningitis are the most common manifestations, with the seventh nerve palsy occurring in 20-50% of patients. Only one case of neurosarcoidosis presenting as a CP angle tumour in a 38 year old man has been reported [56]. Neither CT nor MRI is specific for neurosarcoidosis. Treatment is medical and aggressive surgery should be avoided. The clinical course is difficult to assess. In some patients, it may be chronic, relapsing , or progressive, eventually leading to death despite treatment.

PINEOBLASTOMA OF CP ANGLE

An extremely rare case of a CP angle pineoblastoma has been reported in a 32 year old male who presented with features of raised ICP. Computerised tomography showed a large enhancing mass with hydrocephalus and no abnormalities in the pineal region. Intraoperatively the tumour was easily separable from the adjacent cerebellum and was totally resected.In a few months, he presented with a massive recurrence in the spinal cord and subarachnoid space and died 6 months after first surgery [57].

CRANIOPHARYNGIOMA OF CP ANGLE

Craniopharyngiomas are relatively uncommon lesions accounting for 1.2 to 4% of all intracranial tumours. Rare ectopic craniopharyngiomas located in the optic nerves, pineal region, pharynx and cerebellopontine angle have been reported [58]. Craniopharyngiomas have been noted to extend into the posterior fossa in 4 to 5.9% of cases. Twenty two case reports described patients who had craniopharyngiomas with large posterior fossa components either at initial presentation [59] or at recurrence [60]. In only two of these cases was the tumour thought to arise primarily in the posterior fossa and in others, the posterior fossa component was an extension of a suprasellar tumour [61].

EPITHELOID HAEMANGIOENDOTHELIOMA OF CP ANGLE

Tammam etal reported a rare case of epitheloid haemangioendothelioma in a 4 year old boy who presented with a CP angle tumour thought to be an invasive acoustic schwannoma. Subtotal resection followed by radiotherapy was given [62].

LYMPHOMA OF CP ANGLE

Primary CNS lymphomas are rare tumours. Jellinger et al reported 68 primary CNS lymphomas in a series of 8000 intracranial neoplasms(0.85%) [63]. Most tumours occur in the supratentorial areas including the basal ganglia,thalamus,corpus callosum and paraventricular white matter; however 25% of primary CNS lymphomas occur in an infratentorial location[64], of which the majority are found in the cerebellum. There are only 14 reported cases of malignant CP angle lymphoma in the literature[65] , of which 10 cases of primary lymphoma and 4 cases secondary to systemic malignant lymphoma. The age at onset of initial symptoms was between 24 and 82 years in primary cases and between 28 and 76 years in secondary cases. There was a preponderance of left sided lesions in both primary and secondary types. Bilateral CP angle lesions were noted in 2 patients-one primary and the other secondary category. There was a preponderance of females in primary CP angle lymphoma(a male to female ratio of 1: 2.3) , in contrast to those with primary CNS lymphoma(a male to female ratio of 1.5-2.7 : 1).

The most frequent symptoms are hearing disturbance and cerebellar signs. Facial palsy was recognized in only one case.CP angle lymphomas were isodense or hyperdense on unenhanced CT and exhibit homogenous contrast enhancement. However in contradistinction to most acoustic neurinomas , there is less bony erosion of internal auditory canal. Compared to meningioma , the lymphoma is totally avascular. MRI showed CP angle lymphomas as either low or high intensity on T1-weighted images , iso-intensity on T2-weighted images and homogenous enhancement with Gadolinium.The results of immunohistochemical studies described in 6 cases indicate that they all were B-cell type lymphomas.

There are three distinct patterns in which lymphomas occupy the CP angle,

1) an Extra-axial CP angle lymphoma,
2) an intraaxial lymphoma extending to the CP angle and ,
3) a leptomeningeal lymphoma presenting as a CP angle lesion

It is desirable to obtain a frozen section in all CP angle tumours during surgery to identify the tumour, because aggressive removal is not necessary, but radiation therapy should be additionally performed for malignant lymphoma.

CARTILAGINOUS TUMOURS OF CP ANGLE

Intracranial chondromas and chondrosarcoams of the base of skull are rare neoplasms and their incidence is about 0.1 to 0.2% of all intracranial tumours[66]. They develop from

embryological cartilaginous residues with potential neoplastic properties in sutures or in areas between centers of ossification. A slight female preponderance is reported.

Krayenbuhl and Yasargil describe three types of localization:

1) sinus chondromas , 2) chondromas of the convexity and intracerebral structures and 3) basal chondromas in the region of synchondrosis. The occurrence of these tumors in the CP angle is considered a rarity.

Angiographically chondromas are avascular lesions. On CT scan there may be lytic bony destruction and variable enhancement with contrast.On MRI T2-weighted images, chondrosarcomas are reported to have high signal intensity as a result of increased water in contrast to the meningioma. Close proximity of these tumours to the petro-occipital synchondrosis, lack of typical vascularisation on the angiogram and lytic changes of the skull base strongly argue against meningioma[66].

Radical surgical removal of the tumour is the treatment of choice and should be performed wherever possible.

GIANT ANEURYSMS SIMULATING A CP ANGLE TUMOUR

Giant aneurysms developing in the posterior fossa are very rare. However an analysis of large series of cases of giant aneurysms show that the frequency of them in the vertebrobasilar system is somewhat higher than that of normal sized aneurysms. In Drake`s series giant aneurysms account for 62%[67] and in Sarwar series 25%[68].

The most common sites from which these lesions arise are the bifurcation of basilar artery and the origin of posterior inferior cerebellar artery and very rarely in anterior inferior cerebellar artery[69].

They characteristically present as a space occupying lesion which is more common than subarachnoid haemorrhage. Depending on the site, they present clinically as cerebellar tumours, foramen magnum tumours, intrinsic tumours of the brainstem or cerebellopontine angle tumours.

CT scan may not clarify the diagnosis of the nature of the lesion as reported by many authors[70] because they do not permit differentiation from the tumour. Angiography is the most important diagnostic tool because it reveals the true nature of the lesion.

EPENDYMOMA OF THE CP ANGLE

Ependymomas are relatively rare gliomas arising from the differentiated cell layer lining the ventricular system and central canal of the spinal cord. They primarily affect children with the mean age at diagnosis at 3 years. They comprise between 6 to 15% of childhood brain tumours and account for 1.1 to 5% of all adult intracranial tumours. Roughly 2/3rds occur in the posterior fossa and the remaining 3rd in the supratentorial compartment. They are slightly more prevalent in males (male : female = 1.1 to 1.3 : 1).

Infratentorial ependymomas frequently arise from the ependyma of the 4th ventricle but may also originate in the foramina of Luschka, posterior cerebral aqueduct or the

lateral medullary velum. They can extend inferiorly into the cervical spinal canal through the foramen of Magendie, into the subarachnoid spaces within the cerebellopontine angle through the foramina of Luschka presenting with the cerebellopontine angle syndrome. In some cases, higher grade ependymomas can pread to the other sites via CSF seedlings (3 to 17% of cases).

Prognosis is generally poor and the 5year survival rate is about 50%.[71].

CHOLESTEATOMA OF CP ANGLE

Cholesteatoma constitutes 4.6% of cerebellopontine angle tumours [1] . They occur in two settings i.e: congenital and acquired forms. The former arise from congenital epithelial inclusion rests in contrast to the acquired form secondary to chronic suppurative otitis media. They occur in the subarachnoid space, although they may be present extradurally. Macroscopically they appear white, shiny and lobulated (pearly tumour). They are avascular, lined with squamous epithelium and filled with keratin.

Cholesteatoma compresses and irritates cranial nerves and vessels rather than displacing them. Facial twitching is an early sign with progeressive facial palsy more common than acoustic schwannoma.

CT scan shows a hypodense lesion usually with irregular margins eccentric to the IAM and mild or no enhancement owing to its avascularity. The T1-weighted is usually hypointense and T2WI hyperintense. The clinically important distinction between this and the cholesterol cyst is that the latter is hyperintense on both T1 and T2 weighted images.

The interior of the tumour is caseous and removed with simple suction or curettage. The capsule however, may be adherent to the surrounding neural structures and vessels and may be very difficult to remove totally. Recurrence may occur warranting repeat surgery [1].

RESULTS AND ANALYSIS

In this study, fifteen different rare cerebellopontine angle lesions involving thirty seven patients were reported. The most common lesion found among this group was arachnoid cyst-seen in 11 patients. Other lesions noted in this study were brainstem glioma in 5 patients, medulloblastoma in 4 patients , choroid plexus papilloma in 3 patients. Cavernoma, paraganglioma and giant PICA aneurysms each constituted 2 patients and one each of chordoma, chondrosarcoma , haemangioblastoma , ependymoma , teratoma , cholesteatoma , craniopharyngioma and pontine capillary telangiectasia were found.(table 1).

The commonest age group affected with rare cerebllopontine angle lesions in the present study was 21-30 years(9 cases).Arachnoid cyst was uniformly distributed in all age groups. The youngest patient with arachnoid cyst was 9 years and oldest was 64 years. Brainstem glioma extending to CP angle was commonly seen in second decade in 2 patients(table 2).

Medulloblastoma of the CP angle was found in first 2 decades in 3 patients and at the age of 47 in one patient. Choroid plexus papilloma was uniformly seen in older patients between 4^{th} to 6^{th} decade. Two cases of giant PICA aneurysms presenting as CP angle

lesion were found in the 5th and 6th decades. Cavernoma was seen in 2 patients in the 3rd decade and paragangliomas in the 4th and 5th decades.Teratoma was seen at the age of 2 1/2 years. Bony tumours like chordoma and chondrosarcoma were found in the 3rd decade. Craniopharyngioma and haemangioblastoma were seen in the 5th and 6th decade respectively(table 2).

The male, female distribution in the present study was 18:19. Arachnoid cyst was slightly more common in males than in females(6:5). Brainstem glioma and choroid plexus papilloma were commoner in females(2:3 and 1:2). Medulloblastoma, cavernoma and giant PICA aneurysms had equal sex distribution. Both the paragangliomas involved female patients in the present study(table 3).

Patients with arachnoid cyst had variable duration of symptoms from 2 months to 8 years though most of the cases presented within a year of onset(8 cases). Malignant tumours like brainstem glioma and medulloblastoma presented early within 6 moths of onset of symptoms. Paraganglioma , cavernoma and choroid plexus papilloma had a long interval varying from 1 to 7 years. Among the other rare tumours, craniopharyngioma had the longest interval of 7 years , cholesteatoma 5 years and chondrosarcoma 2years.(table 4).

In the present study, left side was more involved than right side(23:14). Arachnoid cyst, brainstem glioma , medulloblastoma , choroid plexus papilloma and giant PICA aneurysms were the lesions which had definite left side preponderance. Cavernoma,

paraganglioma and other rare tumours had equal distribution on both sides(table 5).

In the present study, the commonest symptom was headache seen in 30 patients. Other commoner symptoms were ataxia(25 cases), vomiting (23 cases) and facial deviation(21 cases). Chewing difficulty was noticed in 4 cases.(2 of arachnoid cyst, 1 craniopharyngioma and 1 pontine capillary telengiectasia). Bloody ear discharge was noted in both the cases of paragangliomas and purulent discharge in cholesteatoma. Decreased tear secretion was noted in 1 case of arachnoid cyst and reduced taste in 1 arachnoid cyst and in cholesteatoma. One of the PICA aneurysms presented with features of acute subarachnoid haemorrhage(table 6).

In the present study, the commonest finding noted was the cerebellar sign in 27 patients. Other common signs were papilloedema(22 cases) , nystagmus(22 cases) and facial nerve involvement in 21 patients. Pyramidal signs were noted in 3 cases of arachnoid cysts, 4 cases of brainstem gliomas and in craniopharyngioma. Both the cases of cavernoma did not have papilloedema, nystagmus , hearing loss or lower cranial nerve symptoms(table 7).

In the present study, 19 patients had undergone only computerized tomography and three patients had only magnetic resonance imaging. Fifteen had both CT and MRI. Cerebral angiography was done in both PICA aneurysms (one thrombosed), both the cases of paragangliomas, 1 case of cavernoma and 1 each of chordoma, chondrosarcoma, haemangioblastoma and pontine capillary telengiectasia respectively. Both the

paragngliomas had feeders from ascending pharyngeal branch of external carotid artery and underwent pre-operative embolisation. One PICA aneurysm inaddition had opposite side superior cerebellar artery aneurysm. Cavernoma and pontine capillary telengiectasia did not reveal any angiographic abnormality(table 8). Calcification in CT scan was noted in chordoma, chondrosarcoma,ependymoma and craniopharyngioma. Of the 37 cases, extension into IAM was noted in only one case of choroid plexus papilloma. Arachnoid cyst and chordoma were the lesions which did not show any enhancement. All cases of medulloblastoma , cholesteatoma , ependymoma , pontine capillary telengiectasia and teratoma showed mild enhancement.Chondrosarcoma showed moderate enhancement. Bright enhancement was noted in choroid plexus papilloma , paraganglioma and with mural nodule in haemangioblastoma . Cavernoma and craniopharyngioma were the tumours which had heterogenous enhancement. All the cases of choroid plexus papilloma , medulloblastoma and 3 of the brainstem glioma had significant hydrocephalus. Cavernoma did not have any hydrocephalus(table 9).

In the present study, 7 cases of arachnoid cysts underwent deroofing. Two cases inaddition had associated chiari malformation and were subjected to foramen magnum decompression. One deroofed arachnoid cyst underwent cystoperitoneal shunt(table 11). Total excision was done in 4 cases. Malignant tumours like brainstem gliomas and medulloblastomas were treated by decompression followed by radiotherapy. Total excision of choroid plexus tumors were done in 2 cases and subtotal excision in 1 patient. Both the cases of giant PICA aneurysms were clipped by retromastoid approach. Both the

cases of cavernoma and one paraganglioma,haemangioblastoma and teratoma could be removed totally.The patient with craniopharyngioma underwent pterional craniotomy at first stage followed by retromatoid craniectomy for the removal of posterior fossa component. Both chordoma and chondrosarcoma were treated by combined subtemporal and retromastoid approach with the addition of posterior petrosectomy in chondrosarcoma because of the tumour involvement. Cholesteatoma was treated by atticoantral suprameatal approach combined with labyrinthectomy and petrosectomy(table 10). Ten out of 37 cases underwent ventriculoperitoneal shunt for hydrocephalus. This included 2 cases of brainstem glioma, all 4 cases of medulloblastoma and one case each of choroid plexus papilloma,chordoma, chondrosarcoma and haemangioblastoma. Three patients underwent shunting along with the tumour surgery and three cases post-operatively because of persistent hydrocephalus(table 11).

Among the operated cases of arachnoid cyst, 5 had facial palsy and 4 had lower cranial nerve palsy in the immediate post-operative period. Four cases of brainstem glioma had both facial and lower cranial nerve paralysis in the immediate post-operative period. All operated cases of medulloblastoma had immediate seventh nerve and 2 had lower nerve involvement. All cases of paraganglioma and giant PICA aneurysm had 7^{th}, 8^{th} and lower cranial nerve paralysis post-operatively. One patient of cavernoma had transient facial palsy. Ependymoma had intact 7^{th}, 8^{th} and lower cranial nerve function in the post-operative period(table 12).

Ten among the 37 patients required post-operative ventilation and 8 patients needed tracheostomy in the post-operative period. Five patients had CSF leak in the post-operative period including one patient with CSF otorrhoea in teratoma patient. None of the patients with CSF leakage required any surgical intervention. Seven patients in the present series had post-operative meningitis and required longterm antibiotic therapy. There were 4 deaths in this series. One case of arachnoid cyst expired on the 10^{th} post-operative day because of aspiration pneumonia and multiorgan dysfunction. The cause of death was brainstem infarct in one case of medolloblastoma and massive operative site haematoma in haemangioblastoma. Paraganglioma operated patient succumbed after 2 weeks because of septicemia(table 13).

Among the eleven operated cases of arachnoid cysts, 8 patients were normal and continuing to work at followup periods ranging from 1 to 8 years. One patient had persisting lower cranial nerve palsy and the other case had lost for follow-up.

The histopathological diagnosis in 5 operated cases of brainstem glioma included one high grade , three cases of grade 2 glioma and the last one astrocytoma with varying differentiation. All the brainstem glioma patients were subjected to adjuvant radiotherapy. The high grade glioma patient was readmitted 8 months later with spinal metastasis and was sent for radiotherapy. One patient had residual hemiparesis at 1 year follow-up and the other had residual 6^{th},7^{th},$8^{th}$and lower cranial nerve paralysis at 6 months follow-up.

Among the 4 cases of medulloblastoma operated, the histological diagnosis was the classical type in 3 cases and medulloblastoma with glial differentiation in one patient. All of them had adjuvant radiotherapy to the local site and whole brain. Two patients were independent at 5 years follow-up, one had improvement of immediate post-operative facial nerve palsy and the other patient had mild cerebellar signs not interfering with his daily activities. One patient was readmitted 2 years later with quadriparesis. MR imaging revealed fresh brainstem lesions and was subjected for chemotherapy.

In the present series, one case of operated choroid plexus papilloma had acinar differentiation. At 2 years follw-up, two patients were independent but with compensated lower cranial nerve palsy and with mild cerebellar signs inaddition in one patient. The other subtotally resected patient had no increase in the size of the residual tumour on 2 years follow-up imaging.

One case of PICA aneurysm had residual facial paralysis at 5 years follow-up. The other case had his tracheostomy closed at 3 months but with persisting facial paresis and was not willing for the surgery of superior cerebellar aneurysm of the opposite side.

Both the operated cases of cavernoma were normal at 7 years without deficits. One case of paraganglioma had residual residual 7^{th}, 8^{th} and compensated lower cranial paresis at 2 years follow-up. Residual tumour in the chordoma patient was re-explored after 6 months. Chondrosarcoma patient was readmitted 1 year later with quadriparesis and total

ophthalmoplegia on the operated side. MR imaging revealed massive tumour recurrence at local site with extension to the middle fossa and cavernous sinus. He was referred for palliative care.

At 3 years follow-up, cholestetoma patient had residual facial paralysis. Pontine capillary telengiectasia patient had residual 7^{th} , compensated lower cranial nerve palsy and mild cerebellar signs at 5 years. Histopathological examination of ependymoma showed degenerative changes and the patient was well preserved at 10 years after surgery.

Histopathological examination of teratoma showed only mature elements and the patient was independent at 11 years and repeat imaging was normal. At 3 years follow-up, craniopharyngioma patient had residual 7^{th} and compensated lower cranial nerve paralysis. She met a road traffic accident 6years after surgery and became hemiplegic.

Table 1 . Tumour Distribution

Serial No.	Tumour	Number	%
1	Arachnoid cyst	11	29.7
2	Brainstem glioma	5	13.5
3	Medulloblastoma	4	10.8
4	Choroid plexus papilloma	3	8.1
5	Giant PICA aneurysm	2	5.4
6	Cavernoma	2	5.4
7	Paraganglioma	2	5.4
8	Chordoma	1	2.7
9	Chondrosarcoma	1	2.7
10	Haemangioblastoma	1	
11	Cholesteatoma	1	2.7
12	Ependymoma	1	2.7
13	Teratoma	1	2.7
14	Craniopharyngioma	1	2.7
15	Pontine capillary telangiectasia	1	2.7
Total	-	37	100%

Table 2. Age Distribution

Age	Arachnoid cyst	Brain stem glioma	CPP	Medulla	PICA	Cavernoma	Paraganglioma	Others
0-10	2	1	-	2	-	-	-	1
11-20	2	2	-	1	-	-	-	-
21-30	2	1	-	-	-	2	-	4
31-40	1	1	2	-	-	-	1	1
41-50	2	-	-	1	1	-	1	1
51-60	1	-	1	-	1	-	-	1
61-70	1	-	-	-	-	-	-	-
Total	11	5	3	4	2	2	2	8

Table 3. Sex distribution

Sex	Arachnoid cyst	Brain stem glioma	CPP	Medulla	PICA	Cavernoma	Paraganglioma	Others	Total
Male	6	2		2	1	1	-	5	18
Female	5	3	2	2	1	1	2	3	19

Table 4. Duration of symptoms

Duration	Arachnoid cyst	Brainstem glioma	CPP	medullo	PICA	Cavernoma	Paragangliom a	Others
< 3 months	2	3		2	1	-	-	1
3-6 months	3	1	1	1	-	-	-	2
6-12 months	3	1	1	1	-	1	1	2
1-2 years	1	-	-	-	1	-	-	1
> 2 years	2	-	1	-	-	1	1	2
Total	11	5	3	4	2	2	2	8

Table 5 . Side

Side	Arachnoid cyst	Brain stem glioma	CPP	Medullo	PICA	Cavernoma	Paragangliom a	Others	Total
Right	4	2	1	1	-	1	1	4	14
Left	7	3	2	3	2	1	1	4	23
Total	11	5	3	4	2	2	2	8	37

Table 6 . Symptoms

Symptoms	Arachnoid cyst(11 cases)	Brain stem glioma	CPP	Medulla	PICA	Cavernoma	Paraganglioma	Others
Headache	6	4	3	4	2	2	2	7
Vomiting	4	4	3	4	1	1	1	5
Double vision	2	3	-	2	1	-	1	3
Facial numbness	5	1	-	-	-	1	-	4
Chewing difficulty	2	-	-	-	-	-	-	2
Hearing loss	3	3	1	2	1	-	2	3
Tinnitus	4	1	1	-	1	1	2	3
Ear discharge	-	-	-	-	-	-	2	1
Decreased tear	1	-	-	-	-	-	-	-
Decreased taste	1	-	-	-	-	-	-	1
Facial deviation	4	4	2	3	1	1	2	4
Vertigo	7	1	1	1	1	2	-	1
LCN symptoms	2	3	1	2	1	-	1	2
Ataxia	7	4	3	3	1	1	1	5

Table 7 .Signs

Signs	Arachnoid cyst	Brain stem glioma	CPP	Medulla	PICA	Cavernoma	Paraganglioma	Others
Papilloedema	5	4	3	4	1	-	1	4
Nystagmus	7	3	3	3	2	-	1	3
V sensory	5	2	-	4	1	1	1	4
V motor	3	-	-	-	-	-	-	2
VI	1	4	-	4	-	-	1(UL)	4
VII	5	3	1	3	1	1	2	5
VIII	4	4	1	2	1	-	2	3
LCN	4	4	1	2	2	-	1	2
Long tract	3	4	-	-	-	-	-	1
Cerebellar	7(4BL,3UL)	4(1BL,3UL)	3UL	4UL	2UL	1UL	1UL	5

Table 8 Imaging

Imaging	Arachnoid cyst	Brain stem glioma	CPP	Medulla	PICA	Cavernoma	Paraganglioma	Others
CT only	5	3	1	3	1	-	1	5
MRI only	3	-	-	-	-	-	-	-
CT& MRI	3	2	2	1	1	2	1	3
Total	11	5	3	4	2	2	2	8

Table 9. COMPUTERISED TOMOGRAPHY FINDINGS

Serial No.	Tumour	calci	IAM extension	enhance	hydroce
1	Arachnoid cyst	-	-	No	4+
2	Brainstem glioma	-	-	4hetero, 1ring	3+
3	Medulloblastoma	-	-	3mild, 1irregular	4+
4	Choroid plexus papilloma	-	1 Y	3bright	3+
5	Giant PICA aneurysm	-	-	1rim	1+
6	Cavernoma	-	-	2hetero	No
7	Paraganglioma	-	-	2bright	1+
8	Chordoma	Y	-	No	+
9	Chondrosarcoma	Y	-	Moderate	+
10	Haemangioblastoma	-	-	Bright+mural Nodule	+
11	Cholesteatoma	-	-	Mild	No
12	Ependymoma	Y	-	Mild	No
13	Teratoma	-	-	Mild	No
14	Craniopharyngioma	Y	-	Hetero	No
15	Pontine capillary telengiectasia	-	-	Mild	No

Table 10. TYPE OF SURGERY

Tumour	deroof	deroof +FMD	decomp	subtotal	total	combined	clip	total
Arachnoid cyst	5	2			4			11
Brainstem glioma			5					5
Medulloblastoma			4					4
Choroid plexus papilloma				1	2			3
Giant PICA aneurysm							2	2
Cavernoma					2			2
Paraganglioma				1	1			2
Chordoma						1		1
Chondrosarcoma						1		1
Haemangioblastoma					1			1
Cholesteatoma						1		1
Ependymoma				1				1
Teratoma						1		1
Craniopharyngioma						1		1
Pontine capillary telangiectasia				1				1

Table 11. SHUNT PROCEDURES

Tumour	Pre-op	Introp	Post-op	No shunt	Total
Arachnoid cyst		1(cystoperit)		10	11
Brainstem glioma		1	1	3	5
Medulloblastoma	1	2	1		4
Choroid plexus papilloma			1	2	3
Giant PICA aneurysm				2	2
Cavernoma				2	2
Paraganglioma				2	2
Chordoma	1				1
Chondrosarcoma	1				1
Haemangioblastoma	1				1
Cholesteatoma				1	1
Ependymoma				1	1
Teratoma				1	1
Craniopharyngioma				1	1
Pontine capillary telangiectasia				1	1

Table 12. Immediate post-operative cranial nerve status

Tumour	7th palsy	8th palsy	LCN palsy
Arachnoid cyst	5	4	4
Brainstem glioma	4	3	4
Medulloblastoma	4	1, 3 CNBT	2
Choroid plexus papilloma	3	3	2
Giant PICA aneurysm	2	2	2
Cavernoma	1	nil	Nil
Paraganglioma	2	2	2
Chordoma	1	Nil	1
Chondrosarcoma	1	CNBT	Nil
Haemangioblastoma	1	CNBT	1
Cholesteatoma	1	1	Nil
Ependymoma	Nil	Nil	Nil
Teratoma	Nil	1	Nil
Craniopharyngioma	1	CNBT	1
Pontine capillary telangiectasia	1	1	1

Table 13. COMPLICATIONS

Tumour	Ventilation	Trachostomy	CSF leak	Meningitis	Aspiration	Death
Arachnoid cyst	1	nil	-	2	1	1
Brainstem glioma	2	2	1	1	2	-
Medulloblastoma	1	1	2	2	1	1
Choroid plexus papilloma	1	1	-	-	1	-
Giant PICA aneurysm	1	1	-	-	1	-
Cavernoma	-	-	-	-	-	-
Paraganglioma	2	2	1	1	-	1
Chordoma	-	-	-	-	-	-
Chondrosarcoma	1	1	-	-	-	-
Haemangioblastoma	1	-	-	-	-	1
Cholesteatoma	-	-	-	-	-	-
Ependymoma	-	-	-	-	-	-
Teratoma	-	-	1(otorrhea	1	-	-
Craniopharyngioma	-	-	-	-	-	-
Pontine capillary telangiectasia	-	-	-	-	-	-

DISCUSSION

According to Moffat etal [1], arachnoid cysts constituted 3.5% of rare cerebellopontine angle lesions in contrast to 29.7% in the present series. Ucar etal [3] found that these lesions are usually found in adults with equal sex incidence. But in the present study, there was uniform distribution between first upto fifth decade and a male-female ratio of 6:5. Little etal [72]noted an average of 4 years from the onset of symptoms to the correct diagnosis in their 20 patients. This was in contrast to our present study where 75% of our patients with cerebellopontine angle arachnoid cyst had symptom duration of less than a year. Pappas and Brackmann[72] described that the signs and symptoms of cerebellopontine angle arachnoid cysts are indistinguishable from an acoustic schwannoma, even after an extensive otolaryngological evaluation. In the present study also, the signs and symptoms simulated an acoustic schwannoma.Higashi etal [4] reported trigeminal neuralgia and hemifacial spasm in association with arachnoid cysts, which were not seen in any of our cases. As reported by many authors, computerized tomography and magnetic resonance imaging established the diagnosis in the present series. Similar to Ucar`s [3] report , we had two patients with Chiari type 1 associated with arachnoid cyst. Most surgeons advocate suboccipital craniectomy and wide excision of the cyst wall but we had more number of deroofing compared to total excision. As reported by most authors, there was regression of cerebellar and pyramidal deficits with no change in hearing status in all our patients.

Brainstem glioma with exophytic extension into the cerebellopontine angle constituted 13.5% of lesions in our series. Tokuriki etal[73] reported 85 cases of brainstem glioma with 81% of patients younger than 16 years at the time of diagnosis which is slightly different from our series with 60% of cases which occurred before 20 years. Langmoen et al reported that the duration of symptoms was shorter in children than in adults(2 to 3 months and 10.6 months respectively).This is slightly different from our series with wide variation in the duration of symptoms from 2-9 months in children and 3-6 months in adults. They did not present with any peculiar features. Tsuchida etal described irregular and peripheral enhancement of the lesion which has been ascribed to coagulation necrosis. This finding was noted in all our cases with irregular or ring enhancement. Decompression followed by radiotherapy was done in all our cases as described by most authors. Brainstem gliomas often lack any surrounding plane and resection may sacrifice the patients ability to swallow and to protect the airway. This complication was noted in 2 of our patients who needed prolonged post-operative ventilation and trachaeostomy.

Rajkumar etal[15] reported that there were no clinical, neuro-otological or neuroradiological findings peculiar to cerebellopntine angle medulloblastoma. Three out of 4 cases in the present series were below 20 years in contrast to all 15 reported cases in adults. Our series had equal sex distribution. All our cases had less than one year of symptoms comparable to Kumar etal`s series. Two of our cases presented with hearing loss similar to Kumar`s series and acute onset hearing loss was not seen in any of our patients. Positional nystagmus, an early sign suggestive of medulloblastoma was noted in 3 of our

patients. The histopathological examination showed the classical pattern in all our 4 cases. At surgery in our patients, tumour had been very vascular and was encircling the 7^{th}, 8^{th} and lower cranial nerves similar to other reports. In the present series, 2 patients were and symptom free at 5years followup, resembling Mehta et al [18]series which showed 30% 5year survival but in contrast to Kumar`s series where 3 of his 4 patients succumbed within 30 months. One of our patient presented 2 years later with brainstem recurrence.

Choroid plexus papillomas constituted 8.1% of lesions in our series. All the patients in our series were adults similar to Talacchi`s series [6]. Talacchi et al reported that the tumours originating in the fourth ventricle and extending into the cerebellopontine angle produce early intrcranial hypertension, and those developing primarily in the CP angle cause symptoms of slow growing tumours of that region. Similar to this series , we had two patients where the tumour originated from the foramen of Luschka and present within 6 –12 months of the onset of symptoms and one patient where the tumour originated from the cerebellopontine angle and extended into the internal acoustic meatus resulted in progressive unilateral hearing loss. Singh etal [7] found that radiologically,absence of bony changes in the region of IAM favour the diagnosis of choroid plexus papilloma. However, Naguib etal[9] found that some papillomas can enlarge the IAM and involve the petrous bone. One of our case had similar presentation with extension into IAM. Similar to Picard et al series, lobulated lesion with irregular borders and uniform bright contrast enhancement were noticed in all our patients. Zhang [11]et al described enlarged AICA with

dilated branches can conclusively establish the diagnosis in vertebral angiography. Similar finding was noted in one of our patients.Similar to Talacchi`s [6]series, all the patients were operated by retromastoid route in our series, which has the advantage of encountering the 7th and 8th cranial nerves on the way ti the tumour. Singh[7] et al advised radical excision except when the tumour is adherent to the brainstem. Chan et al described the main surgical difficulties viz. the tumour bleeding and firm adhesion to the brainstem and lower cranial nerves which might lead to incomplete removal. This finding was noted in one of our patients which resulted in subtotal excision.

Cavernoma represented 5.4% of lesions in the present series. Both of our patients were in the fourth decade similar to the 4 of 5 previously reported cases. We had equal sex distribution in contrast to the previously reported cases , all of whom were males. One of our case had the symptom duration of 6 months similar to the Vajramani`s series [75]and the other had long duration of symptom before presentation. One of our case presented with 5th and 7th cranial nerve palsy similar to Iplikcioglu[76] and Brunori`s[77] series. Both the patients had normal hearing in our series in contrast to all the other reported cases. Both the cases had the solid and cystic components with heterogenous enhancement similar to the other series.Iplikcioglu etal found calcification in the lesion and Kim et al [29]did not find any calcification in his series. Both our cases did not show aby calcification or bleed radiologically or histopathologically.Vajramani etal described that the tumour was beefy red, highly vascular and advocated staged resection in order to preserve the cranial nerve function. In Brunori`s series, the lesion was reddish blue, mulberry like lesion adherent to

brainstem and 7th and 8th nerve complex. Contrary to these reports, we were able to do single staged total excision with preservation of 8th nerve in both our cases and 7th nerve in one case.

Paraganglioma constituted 5.4% of lesions in the present series. Both our patients were middle aged adults similar to other reported cases. Brown etal[74] reported that there is a definite female preponderance , with the male-to-female ratio of 1:6. Similar to this , both were females in our series. Both our cases presented with hearing loss, tinnitus and bloody ear discharge similar to Jamjoom etal[78]series. Computerised tomography showed hyperdense lesion with erosion of petrous bone and brilliant contrast enhancement in both the cases similar to the other reported series. Angiography showed prominent ascending pharyngeal and external carotid feeders in both the cases in contrast to Jamjoom`s series which showed moderate vascularity and no tumour blush. One was operated by retromastoid craniectomy with total excision and the other by Fisch type B approach with subtotal excision. Both the patients had post-operative 7th,8th and lower cranial nerve palsy similar to other case reports and both needed ventilation and trachaeostomy. One patient in our series had CSF leak which resolved on conservative therapy similar to Jamjoom`s series. One patient had improvement of cerebellar function but persistent 7th and 8th cranial nerve palsy on followup similar to other case reports.

We had one mature teratoma in our series in a 21/2 year old child similar to other teraoma case reports but in contrast to Iplikcioglu`s series [44]of cerebellopontine angle teratoma in an adult. In contrast to the reported case with symptom duration of 3 months, our case had 1 year history . Computerised tomography showed a mixed density lesion with calcification and totally removed by retromastoid craniectomy similar to other series. We had one case of pontine capillary telengiectasia in our series in a 34 year old man. In contrast to Korinth`s series[79] who presented with trigeminal neuralgia, our patient presented with headache , facial numbness and cerebellar symptoms. Angiography was normal in our case in contrast to Korinth`s reported case. Subtotal excision of the lesion was done and patient had improvement of fifth nerve function but persistent facial palsy and cerebellar signs at follow-up.

We had one case of ependymoma in our series in a 30 year man in contrast to previous reported cases which show a high preponderance in children. Our patient presented with hearing loss and ataxia simulating an acoustic neurinoma. Computerised tomography revealed a hyperdense lesion with calcification and mild enhancement similar to other reported cases. He underwent subtotal excision of the lesion and had good post-operative outcome.

We had one case of cholesteatoma in a 27 year old man in our series who presented with ear discharge, hearing loss and facial deviation similar to other reported series. Computerised tomography revealed a hypodense lesion with irregular margins with petrous extension in our case similar to other reported cases. Per-operatively the lesion was

adherent to the 7^{th},8^{th} cranial nerves and vessels resulted in subtotal excision as described by many authors. He had persistent 7^{th} and 8^{th} nerve palsy at follow-up similar to other series.

We had one case of craniopharyngioma in our series in a 48 year old female in contrast to previously reported cases which showed maximum incidence in the first and second decades. Our case presented with visual blurring, hearing loss and cerebellar symptoms similar to other case reports. Computerised tomography revealed mixed density lesion with calcification similar to other series. Connolly etal [59]advised removal of parasellar component first when visual symptoms are predominant and removal of cerebellopontine angle component first when brainstem compressive symptoms are prominent. Similar to this, our patient underwent pteional craniotomy at first stage followed by retromastoid craniectomy at second stage for the removal of posterior fossa component.

We had one each of chordoma, chondrosarcoma , haemangioblastoma and pontine Capillary telengiectasia which all resembled acoustis neurinoma and the diagnosis was confirmed only after histopathological examination.

CONCLUSION

Cerebellopontine angle lesions remain a challenging problem for the neurosurgeon. Although schwannoma , meningioma and epidermoids comprise the majority of lesios at this site , this report brings out a plethora of other pathologies like arachnoid cyst,brainstem glioma, choroid plexus papilloma and medulloblastoma .Arachnoid cyst is the commonest of the rare leisons occurring at the cerebellopontine angle in the present series. As is frequently the case in clinical medicine, a well taken historty and a meticulous neuro-otological examination will alert the astute clinician to the fact that all is not what it initially appears to be. Patients with symptoms others deafness,tinnitus and disequilibrium and those with additional neurological findings and involvement of ninth,tenth,eleventh and twelvth cranial nerves deserve careful consideration particularly when internal acoustic meatus is normal.Though, high resolution computerised tomography by demonstrating the bony anatomy of the temporal bone and skull base in great detail and magnetic resonance imaging which reveals the soft tissues with such amazing accuracy has made it more likely that the correct diagnosis of these interesting lesions can be made, most of these lesions are operative surprises and will be confirmed only by histopathological examination . Angiography is required in vascular lesions like paraganglioma , haemangioblastoma and choroid plexus papilloma for demonstrating the feeders and pre-operative

embolisation.Though the retromastoid route offers the best exposure and optimal access , some lesions extending into the middle fossa like craniopharyngioma, to the clivus like chordoma and chondrosarcoma , to the jugular foramen like paraganglioma may require combined approaches. Although total excision is possible in many of these lesions, malignant tumours like medulloblastoma and brainstem glioma might need decompression and adjuvant therapy. Patients with arachnoid cysts and cavernoma have excellent surgical outcome.

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