

Evaluation of Long Term Outcome in Surgically  
Treated Posterior Fossa Epidermoid:  
An Institutional Experience



*Submitted for MCh Neurosurgery*

By

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# ***CERTIFICATE***

This is to certify that the thesis entitled “Evaluation of Long Term Outcome in Surgically Treated Posterior Fossa Epidermoid: An Institutional Experience” is a bonafide work of Dr. Ansari Khursheed Ahmad E. and was conducted in the Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences & Technology, Thiruvananthapuram (SCTIMST), under my guidance and supervision.

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# ***DECLARATION***

This thesis titled “Evaluation of Long Term Outcome in Surgically Treated Posterior Fossa Epidermoid: An Institutional Experience” is a consolidated report based on a bonafide study of the period from January 1997 to December 2007, done by me under the Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences & Technology, Thiruvananthapuram.

This thesis is submitted to SCTIMST in partial fulfillment of rules and regulations of MCh Neurosurgery examination.

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## INTRODUCTION

Epidermoid cysts, also known as pearly tumors or cholesteatomas, are uncommon congenital lesions. Epidermoid tumors belong to the spectrum of “tumors of disordered embryogenesis” and are known to occur at diverse locations in the neuraxis. Epidermoids represent 0.2 to 1% of all primary intracranial tumors (107). These slow-growing benign tumors commonly occur in the parapontine or parapituitary regions. Uncommonly, their location can also be purely intracerebral, at the brain stem, or intraventricular. Cerebellopontine angle(CPA) epidermoids constitute 40% of all intracranial epidermoids (100) and are of considerable surgical interest. Epidermoids presents with a distinct growth pattern, i.e., they"flow" across the basal surface of the brain into the subarachnoid space and only in later stages of growth do they behave as expanding lesions, remaining clinically silent for many years (9,13,106,129). At diagnosis, epidermoids typically contain already infiltrated sulci and cisterns, displaced brain tissue, and engulfed cranial nerves and vessels (133). In particular, posterior fossa epidermoids are often the most troublesome to cure because of their insinuating growth into spaces difficult to reach in a single stage. It is often associated with significant morbidity in the postoperative period.

On the other hand, partial removal risks recurrence and subsequent surgery (8).

Even though neuroimaging allows reliable diagnostic evaluation and accurate surgical planning, it is still uncertain whether an aggressive attitude is better than conservative treatment in large, expansive, and widespread tumors (13,16,132,133). Moreover, the effect of tumor size on removal rates has not yet been determined. The epidermoid's slow growth poses a surgical dilemma in cases of elderly patients and tumor recurrences (8,16,132,133). Further insight into life expectancy may be revealed by long-term follow-up.

We summarize our single-institution experience of having treated patients with posterior fossa epidermoid tumors from January 1997 to December 2007 with an intention to contribute to the general body of knowledge of these tumors. The clinical profile, imaging characteristics, management options and long term outcome with the extent of surgical removal were retrospectively analyzed and compared with the available literature.

## REVIEW OF LITERATURE

Epidermoid tumors were first described in 1829 by the French pathologist Cruveilhier (29). Because of its pearly appearance, he described it as “La Tumeur Perlee”. Virchow and Bailey spoke “pearly tumors” (82). In 1836, Muller introduced the term “cholesteatoma” because of the presence of cholesterol crystals in these tumors. In 1854, Von Remak developed the theory of displaced germ cells occurring at an early embryonic stage (82)

Lepoivre and Pertuiset (74) introduced the first epidermoid classification, consisting of three groups, i.e., vertebrobasilar and carotid (both related to the main blood vessels at the base of the brain), and intraventricular (related to choroidal arteries). The primary criterion for this classification was the role of vessels in tumor migration. In 1969, Obrador and Lopez-Zafra (97) distinguished four groups at the base of the brain, according to the clinical and surgical data, as suprasellar-chiasmatic, parasellar-sylvian, retrosellar-cerebellopontine, and basilar-posterior fossa. These authors did, however, emphasize that such classification "does not in any way represent the true extension of the tumor." In the computed tomographic era, Yasargil et al. (133) added two other regions, anterosellar-frontobasal and mesencephalic-pineal, to

the same topographic distribution, according to anatomic criteria with reference to basal CSF cisterns. In 1996, Samii et al. (111) published another classification of posterior fossa epidermoids that introduced the concept of extension to better define the size of the tumor and the surgical implications. He classified CPA epidermoid in to five subgroups; as CPA alone, CPA with transtentorial extension, CPA with middle fossa extension, CPA with foramen magnum extension and CPA with transtentorial & foramen magnum extension.

### **Embryogenesis and Pathology:**

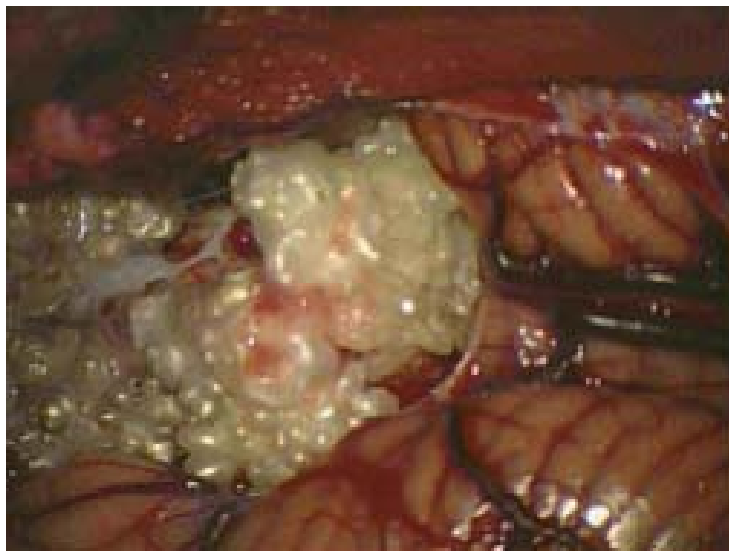
These rare lesions arise from displaced ectodermal cells during closure of the neural tube in the third to fifth weeks of embryonic life (27,29,47,61,69). Rengachary et al (104)suggested that epidermoid cysts occur because of an aberration of epithelial rests or sequestration of ectodermal elements. Congenital sequestration occurs between the third and fifth weeks of intrauterine life as the medullary groove closes. Sequestration usually occurs either in the midline or in relation to optic and otic vesicles. Secondary vesicles, such as the optic and otic vesicles, develop at about the 5th week, and inclusions in these regions may explain cerebellopontine lesions (124). Chandler et al (24) suggested that it seems reasonable to assume that when the neural tube closes and

divides from the cutaneous ectoderm, rests of cells are left on the inner or outer surface, or within the neural tube ectoderm. This would explain the occurrence of epidermoid tumors intraventricularly, on the surface, or within the brain (61). Pathogenesis of intraventricular epidermoids is however uncertain and probably relates to the development of choroidal vessels which explains the lateral migration through the choroidal fissure and subsequent entrapment of neuroepithelial cells (61). The tumours reported have a connection with median/paramedian structures, and the view that these lesions commence from quadrigeminal cisterns too needs serious consideration. Deep seated intracerebral epidermoids can project into the cavity of the lateral ventricles, and appear to be intraventricular (17).

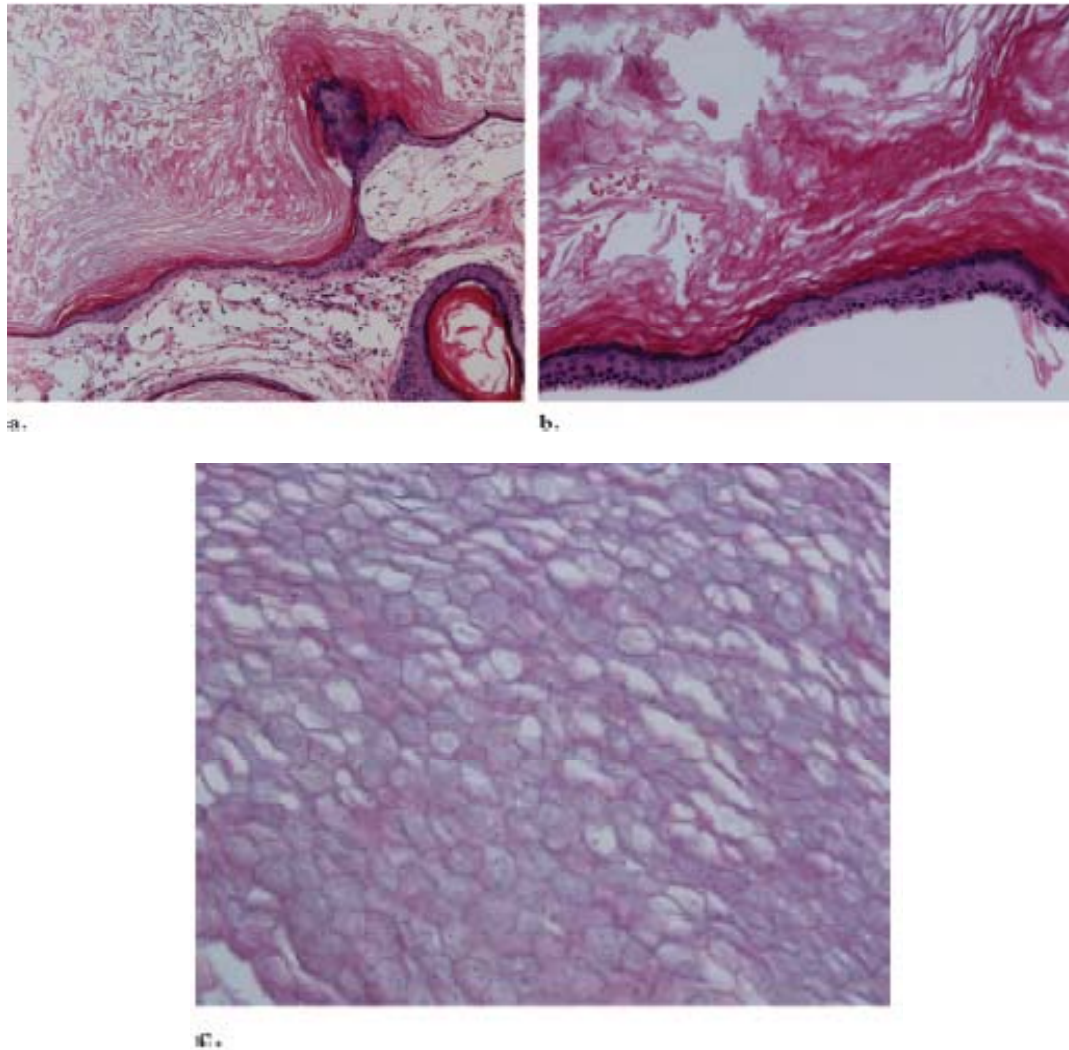
Chambers et al (23) assumed that dermoids are closely related tumors of similar origin, probably the result of more primitive cellular implants. Based on these assumptions, Kaido et al (61) summarize the possible pathogenesis of intraparenchymal epidermoid cysts as follows “they are composed of an outer capsule, an epithelial layer, and, in some cases, an inner cystic fluid. The outer layer is composed of connective tissue, which surrounds a layer of keratinized stratified squamous epithelium”. As the epithelial layer desquamates, the cells accumulate

and form a cholesterol-rich inner layer that gives the cyst its characteristic pearly white appearance (27,61,127,133).

Histopathologic evaluation shows typical features that were consistent with members of the family of pearly tumors (epidermoid tumors), such as attenuated squamous epithelium and abundant anucleate squamae that constituted the bulk of the mass (Fig 2a). The epithelial layer showed regular maturation with no evidence of atypia (Fig 2b). In many regions, cell ghosts (or “shadow cells”) resembling vegetable matter were noted (Fig 2c). No sweat glands or sebaceous glands (findings suggestive of a dermoid) were seen.



**Figure 1:** Intraoperative photograph shows an irregularly lobulated, cauliflower-like pearly tumor, an appearance characteristic of an epidermoid



**Figure 2:** Photomicrographs (hematoxylin-eosin stain) demonstrate an attenuated squamous epithelium with abundant anucleate squamulae (original magnification, x10; **a**), regular maturation of the epithelial layer (original magnification, x20; **b**), and many regions containing cell ghosts or “shadow cells” (original magnification, x20; **c**). Note the

absence of dermal appendages. These findings are typical features for a member of the family of pearly tumors (epidermoid tumor).

### **DISTRIBUTION AND CLASSIFICATION:**

CPA epidermoids constitute approximately 50% to 60% of all intracranial epidermoids and are the third most common tumors of the CPA after acoustic neuromas and meningiomas (8). CPA epidermoids of large extension are variously classified as lateral suboccipital (43), cerebellar (48,80), or CPA-clival (48). The second most prevalent location in the posterior fossa is the fourth ventricle (accounting for 5-18% of all intracranial epidermoids) (79). Other uncommon sites have been reported and grouped differently, as follows: 1) pineal (48,86), quadrigeminal (105); 2) prepontine (43,105), clivus (134), midline (50,66); and 3) paramedian (7), parapontine (54,98,52), petroclival (8), paratrigeminal (14), parasellar-CPA (134), and suprainfratentorial (109).

Intracranial epidermoid cysts can be primarily divided into four categories depending on their anatomic origin and primary location (76,134).

1. Retro sellar cerebellopontine angle
2. Parasellar sylvian fissure

3. Suprasellar chiasmatic and
4. Basilar posterior fossa

However, epidermoids are often seen in several other intracranial compartments like interhemispheric fissure, intraventricular and even intradiploic locations (6,24,27,36,64,67,76,130). In a landmark review, Lepoire and Pertuiset (74) offered an ingenious explanation regarding the seemingly haphazard anatomical distribution of the intradural tumors, considering that the epiblastic inclusions are carried to their final destinations by the developing network of cerebral arteries. Based on this hypothesis, the authors classified the location according to the major arterial territories: carotid(chiasmal and parasellar, frontal, supracallosal and sylvian), vertebrobasilar(cerebellopontine angle (83), prepontine, cerebellar), and choroidal(intraventricular). Most series have shown the infratentorial compartment as the most common location for these tumors with the cerebellopontine angle accounting for most of these cases (13,16,21,35,38,88,90,111,114,123,130). In Akar et al's (5) series of 28 patients, the most common location was the posterior fossa (71.4%). 17 of these 20 were located in the cerebellopontine angle, 2 in the fourth ventricle, and one in the cisterna magna. The other tumors were in the supratentorial location- 4 in the sylvian fissure, 2 in the occipital lobe, one in the lateral ventricle and one in a primary

intradiploic location. In Zhou's (134) series of 102 patients, the supratentorial and infratentorial tumors were of almost the same number with the site-wise distribution as given in Table 1.

|                             |       |
|-----------------------------|-------|
| Intradural                  | 89.2% |
| Extradural                  | 10.8% |
| Intraventricular            | 5.9%  |
| Intracerebral 11 cases      | 10.8% |
| Supratentorial 4 cases      | 3.9%  |
| Infratentorial 7 cases      | 6.8%  |
| Cisternal 74 cases          | 72.5% |
| Suprasellar 8 cases         | 7.8%  |
| Callosal 2 cases            | 1.9%  |
| Parasellar-sylvian 10 cases | 9.8%  |
| CP angle 31 cases           | 30.4% |
| Parasello-CP angle 8 cases  | 7.8%  |
| Clivus 9 cases              | 8.8%  |

**Table 1:** Site-wise distribution of intracranial epidermoid (Zhou et al (134))

In Yasargil's series (133) of 35 cases, 25(71%) were in the infratentorial compartment; of these, 22 were found to be primarily in the cerebellopontine angle and 3 within the 4<sup>th</sup> ventricle. There were 10 supratentorial tumors (29%) of which seven were primarily situated along the medial base of the middle cranial fossa. One was suprasellar and the two remaining tumors were in the mesencephalic/pineal region. In Yamakawa's (132) series, the distribution of tumors according to location was as given in Table 2;

|                  |    |
|------------------|----|
| Cranial vault    | 12 |
| Intracranial     | 33 |
| CP angle         | 15 |
| Middlefosa       | 5  |
| Hemisphere       | 5  |
| Suprasellar      | 3  |
| Third Ventricle  | 3  |
| Fourth ventricle | 2  |

**Table 2:** Distribution of tumors according to location (Yamakawa et al (132))

In Samii Madjid series of 40 patient with Cerebellopontine epidermoid following were the distribution (111) Table 3.

| Location of Lesion                              | No. of Cases |
|-------------------------------------------------|--------------|
| CPA alone                                       | 15           |
| CPA + transtentorial extension                  | 3            |
| CPA + middle fossa extension                    | 5            |
| CPA + foramen magnum extension                  | 9            |
| CPA + transtentorial & foramen magnum extension | 8            |

**Table 3:** Distribution of CPA epidermoid according to location (Samii M. (111))

Talacchi et al (120) in their series of 28 patients with posterior fossa epidermoid had epidermoid tumor had distributions of tumor given in Table 4:

|          |               |   |
|----------|---------------|---|
| CPA (20) | Pure CPA      | 6 |
|          | Suprasellar   | 5 |
|          | Parasellar    | 3 |
|          | Mesencephalic | 6 |

|                           |               |   |
|---------------------------|---------------|---|
| Posterior fossa Basal (3) | Parasellar    | 2 |
|                           | Mesencephalic | 1 |
| Fourth ventricular        |               | 5 |

**Table 4:** Distribution of posterior foss tumors according to location (Talachchi et al)

### **Clinical Aspects:**

Epidermoids have an extremely slow linear growth rate (9). Hence, the duration of symptoms is often prolonged and the patient presents late in the course of the illness. The symptomatic onset of epidermoid cysts is usually lasting 2 or more years, although some patients with remitting signs and symptoms (28) or with rapid onset (80,132) have been reported. Onset of Symptoms and clinical features may vary to some extent for CPA, posterior fossa basal, and fourth-ventricle tumors. Epidermoid cysts of the CPA cause the symptoms and signs of a slowly expanding mass in that region, (16) including ataxia, nystagmus, facial pain, paresthesias, and weakness (8,35,79,81,89).

The involvement of the facial nerve (34) or unilateral hearing loss (8,80,110,132) has been reported as the most common sign. All investigators agree, however, that acoustic neurinomas involve the facial nerve much later than CPA epidermoid cysts. This is because the

epidermoid tends to “strangle” the seventh cranial nerve and reduce its blood supply (11). The acoustic tumor may stretch the nerve but impulses can still be conducted.

Epidermoid cysts of the CPA have been known to cause trigeminal neuralgia, atypical facial pain, glossopharyngeal neuralgia, and hemifacial spasm (8,16,34,35,41,58,79,81,89,110,132).

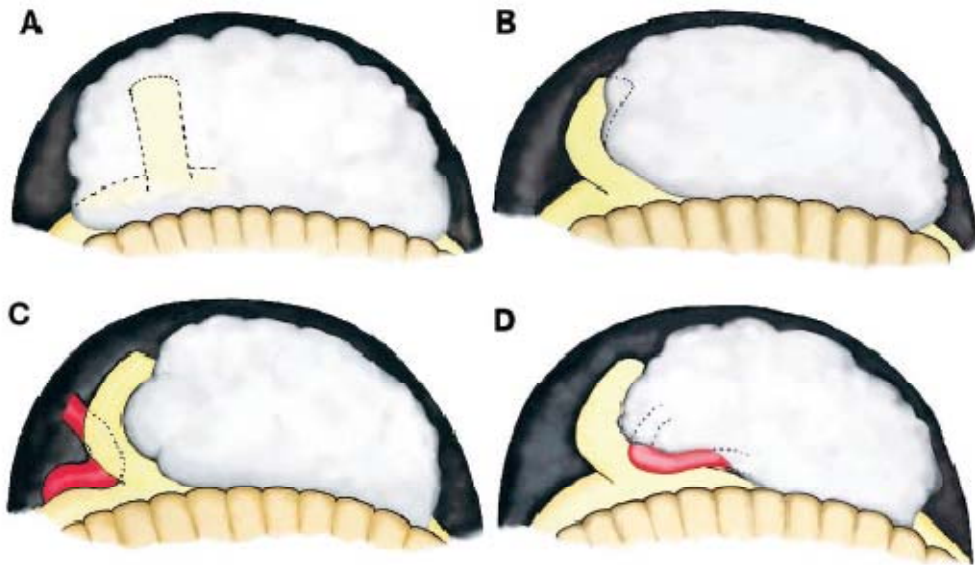
Various mechanisms by which epidermoid cyst can cause trigeminal neuralgia includes: direct compression of the nerve and irritation, (34,81) pushing the trigeminal nerve against a blood vessel,(58) or local irritation from cholesterol seeping (Figure 3).(109)

**Anatomic relationship between the tumor, the artery, and the cranial nerve (69).**

The relationships between the tumor and the neurovascular components were classified into four types according to the irritation pattern of the root entry/exit zone (REZ) of the respective cranial nerves:

1. *Type A*: The nerve is completely encased by the tumor without displacement.
2. *Type B*: The nerve is compressed and distorted by the tumor.
3. *Type C*: The nerve is displaced cranial or caudal direction by the tumor and is directly compressed by an artery on the opposite side of the nerve from the tumor.

4. *Type D*: The nerve is directly compressed by an artery displaced by the tumor located in the same side. Photographs and schematics of these relationships are presented in Fig. 3



**Figure 3:**Anatomic relationship between the tumor, the artery, and the cranial nerve .

Cyst rupture is considered a rare but serious complication. It initiates brief recurrent episodes of aseptic meningitis caused by the chemical action of keratin, cholesterol, and other proteinaceous derivatives on arachnoid membranes and nerve parenchyma (1,122)

In the fourth ventricle, the interval between onset of symptoms and diagnosis can be as short as 1 month to as long as 2 year. The most common presenting symptom is gait disturbance. Although some

patients present with pressure signs, papilloedem and sixth nerve palsies (92)

### **Natural history and behaviour:**

Epidermoid tumors tend to grow very slowly along natural cleavage planes, becoming clinically apparent only after years of undetected growth (9,13,110,132). These slow-growing histologically benign lesions possess a unique capacity to flow across the basal surface of the brain into any available space dissecting along natural tissue planes, and only in later stages of their growth do they behave as expanding lesions and surround and embed rather than displace the cranial nerves. However, suprasellar and intraventricular epidermoids (87) tend to become symptomatic much earlier compared to other locations

Epidermoid tumors are typically benign lesions, but there have been reports of malignant degeneration (2,4,18,39,75,91,95,113). These tumors have a tendency to recur if partially removed (102). It has been suggested that the growth rate of an epidermoid tumor is one generation

per month, essentially the same as the turnover time of normal human skin (9,107). Assuming only a single cell was retained after surgical resection, a patient would be at risk for recurrence of identical dimensions for a period equal to his age at the time of resection plus 9 months. This hypothesis is valid, assuming that the tumor is congenital in nature and that individual variations exist regarding cell size, rate of desquamation product breakdown, local structural effects , hormonal and immune responses etc. These tend to grow by spreading widely and extending into surrounding areas, adhering to neurovascular structures, such as cranial nerves, perforating arteries, veins and brainstem. Associated chemical meningitis also may play a role in the adhesion process. Lunardi et al (79) infer from their series that an epidermoid tumor cannot re-grow after surgery at the same preoperative constant rate. The amount of capsule left in situ, the volume of blood supply and surgical trauma all affect tumor re-growth to a variable degree.

**Radiology:**

On computerized tomogram (CT) scan, epidermoids typically appear as low density, non-enhancing lesions (19,23,32,34,40,45). On CT scan, the lesion usually appears as a hypodense mass with an attenuation value between -2 to +32 Hounsfield Units. Lesions with a

relatively high density between 80 and 120 HU can also occur (20,118) epidermoids may exhibit atypical features and appear hyperdense (53,90), have rim calcification (16,48,103) or marginally enhance with contrast (53).Li et al (75) observed that hyperdense epidermoid tumors as observed in 3% in their series was more common in females and probably is related to recurrent leakage of the irritating cyst contents and subsequent chemical inflammatory response. In contrast to typical epidermoid tumors and other lesions, hyperdense tumors are more prone to spread intra-operatively and result in severe aseptic meningitis (47). Similarly contrast enhancement in an otherwise typical epidermoid cyst suggested malignant degeneration (44).



**Figure 1.** Axial CT scan demonstrating a right-sided cerebellopontine angle (CPA) epidermoid cyst.

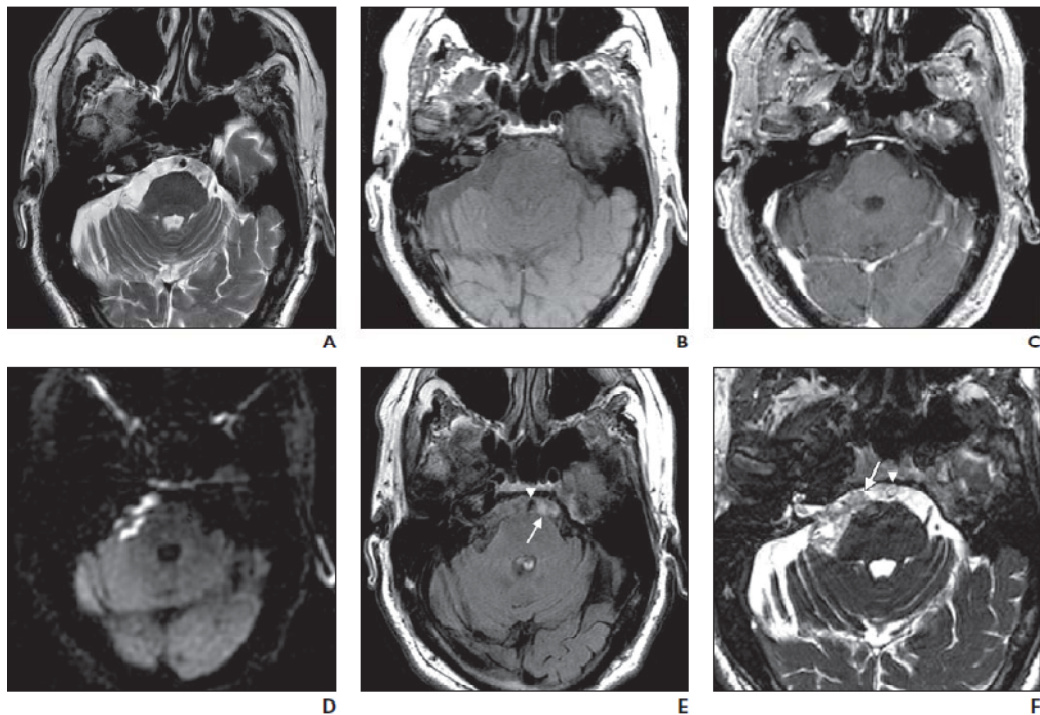
Magnetic resonance imaging (MRI) of brain has superseded CT for accurate radiological diagnosis of these lesions, mainly because diffusion-weighted imaging definitively differentiates tumor from CSF-filled cisterns and cysts (19,31,34,46,57,71,91,94,99,128,131).

Epidermoid tumors generally demonstrate minimal hypo-intensity, typically between CSF and brain parenchyma, on T1-weighted images, and hyper-intensity, similar to or greater than CSF, on T2-weighted images and do not enhance with gadolinium (57,94,99,128,131). Several studies (26,37,51,63) have reported that FLAIR sequence is superior to the conventional MR sequences in demonstrating the epidermoid cysts. The signal characteristics of an epidermoid on T2-weighted imaging have been attributed to the presence of increased water content within the tumor. Hemorrhage has also been reported in epidermoid cysts resulting in unusual signal intensities on MR imaging (25,55).

Role of diffusion weighted sequences (DWI) in the diagnosis has been extensively studied (10,26,37,51,56,57,59,71,91,112,126). The

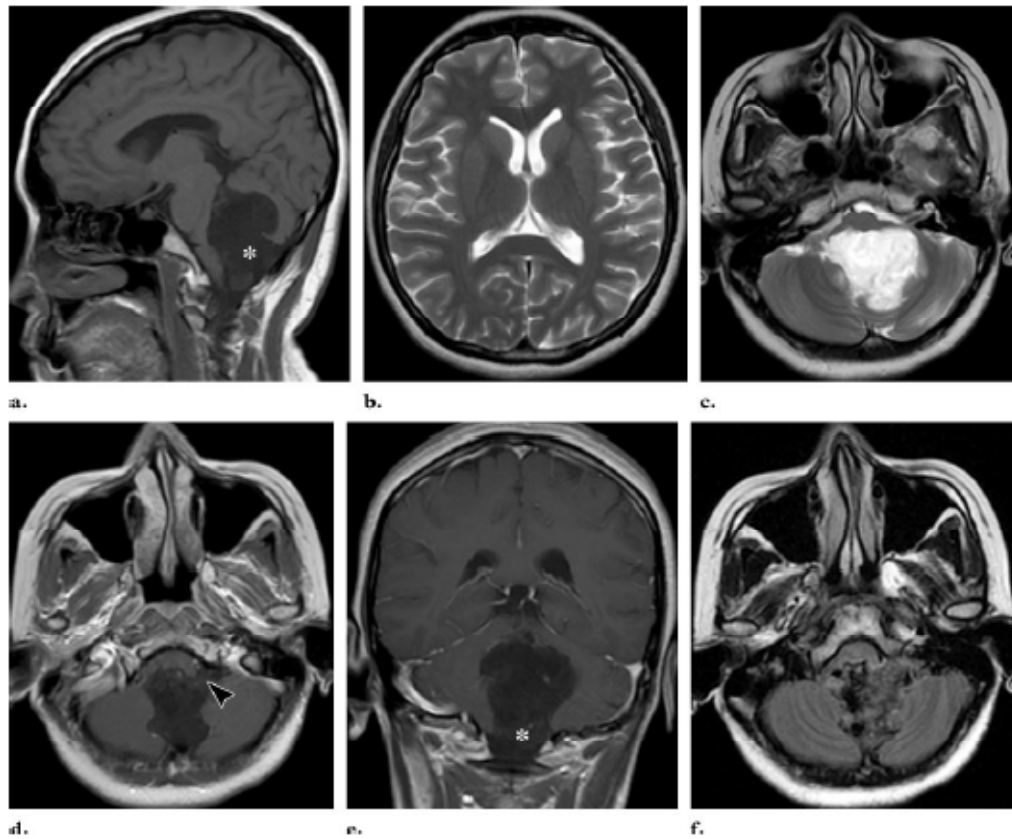
hyper-intensity of epidermoid tumors on echo-planar DW imaging is not caused by diffusion restriction in the lesions but by the intrinsic T2 shine-through effect (26). DW imaging is superior to other magnetic resonance imaging sequences in delineating the edges of the epidermoid cysts. Exponential DW images have shown that the hyper-intensity in the trace images are caused by increased T2 effect of the lesion rather than the decrease in ADC values (10,51). DWI is the most sensitive investigation to detect recurrence in an operated patient (3,102,108). Santhosh K et al (112) have shown that epidermoid cysts showed high fractional anisotropy(FA) with directionally-averaged mean diffusivity(Dav) values similar to that of normal white matter. Exponential apparent diffusion coefficient (eADC) maps did not show any restriction of diffusion. FA values were high, but not as high as that for the white matter. Planar anisotropy (CP) values were higher and linear anisotropy (CL) values were lower than those obtained for the white matter in various regions. M Jolapara et al.(59) in a study noted that diffusion tensor mode values were near -1, and CP values were high within the tumor. This suggested preferential diffusion of water molecules along a two-dimensional geometry (plane), which could be attributed to the parallel-layered arrangement of keratin filaments and flakes within these tumors. They concluded that DTI with DTM can

provide information regarding the micro-structural anatomy of the epidermoid cysts.

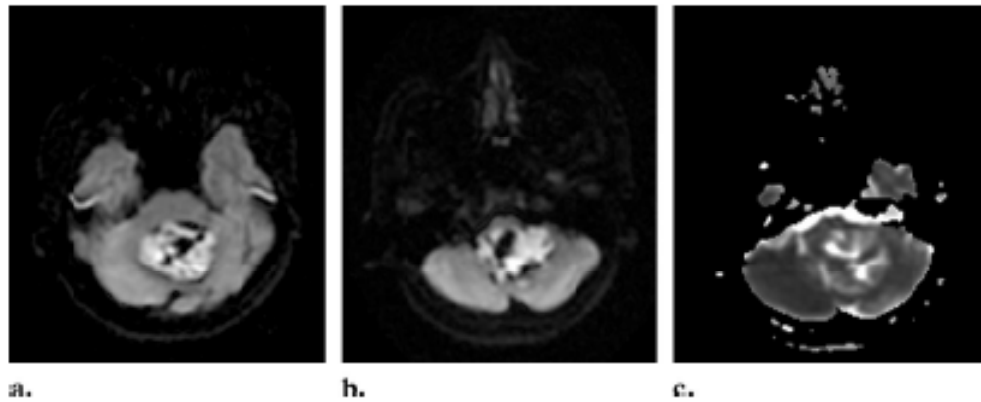


**Figure 4:** **A** and **B**, Epidermoid cyst of CPA appears iso- or hyperintense to CSF on T2-weighted images (**A**) and iso- or slightly hyperintense to CSF on T1-weighted images (**B**). **C**, Epidermoid cyst does not enhance after IV gadolinium administration. **D**, Unlike arachnoid cysts, epidermoid cysts show high signal intensity on diffusion-weighted images. **E**, Signal of epidermoid cyst does not completely suppress on FLAIR images; however, delineation of epidermoid cysts may be confounded on FLAIR images by CSF pulsation artifacts, as in this

example where hyperintense CSF flow artifact (*arrow*) lies to left of basilar artery flow void (*arrowhead*). **F**, MR cisternography more accurately delineates lobulated margins of epidermoid cyst (*arrow*) as well as cisternal structures such as basilar artery (*arrowhead*).



**Figure 5:** Epidermoid tumor depicted on sagittal T1-weighted (**a**), axial T2-weighted (**b, c**), axial (**d**) and coronal (**e**) gadolinium-enhanced T1-weighted, and axial fluid-attenuated inversion recovery (FLAIR) (**f**) images.



**Figure 6:** DW images (a, b) and the corresponding apparent diffusion coefficient map (c) demonstrate significant areas of restricted diffusion within the fourth ventricular mass (highsignal intensity on DW images, low signal intensity on apparent diffusion coefficient map).

### **Management:**

Because epidermoids are not sensitive to radiation or chemotherapy (27) treatment of these lesions relies exclusively on surgical excision (27,130). Operative morbidity and mortality from attempted removal of these cysts have declined remarkably in the last 20 years. Prior to the advent of the operative microscope, operative mortality ranged from 20% to 57%.(42) This rate dropped to approximately 6% for operations performed since 1951.(79) A review of case reports from the 1970s suggests an even lower operative mortality rate following the removal of this lesion (8,16,35, 80,109,132) Recently,

Yasargil and colleagues (133) reported their results in 22 patients with CPA epidermoids; in almost all of these cases cyst removal was total with a mean follow up of 5 years and no operative mortality.

There is no consensus in current literature regarding the extent of removal of epidermoid cysts. Although the goal of surgery is complete removal of the lesion without damage to adjacent neurovascular structures, portions of the capsule adherent to these structures often make this extremely difficult (5,8,38,48,50,65,69,77,79,111,115,120,132,133). Some authors promote radical resection of tumor to prevent recurrence, whereas others advocate a more conservative approach to minimize operative morbidity and mortality (30,35, 79,114,132).

Berger and Wilson (16) did not perform total removal in any of their cases, reporting minimum patient morbidity and no recurrences over 4.5 years of follow-up. On the other hand, Yasargil et al (133) achieved total removal in 95% of patients with minimum morbidity and a 9% recurrence rate over 5.2 years of follow-up. All other reports in literature report results which are somewhere in between these ends of the spectrum. Because there are so many differences and inconsistencies in the surgical methods and data cited in these series, it is difficult to

come to any firm conclusions. Another limitation is lack of a system for grading the complexity of these lesions. There is no available grading system to accurately quantify the extension of epidermoid tumors to adjacent spaces and degree of adherence to neighboring structures. These variables must be taken into account when considering extent of removal and resultant clinical outcome.

Total removal of tumor should be the standard goal when operating on epidermoid cysts. It has been suggested that with microscopic meticulous sharp dissection, every bit of the capsule should be removed to prevent a recurrence. However, in more complex cases where the epidermoid is densely adherent to vital neurovascular structures near/subtotal removal is a better option and carries little if any risk of increasing the rate of recurrence (16,67).

The lesion when confined to the CP angle is approached by a retromastoid craniectomy, whereas significant supratentorial extension needs a combined retromastoid and subtemporal approach or a staged procedure. However a retromastoid approach allows radical removal of supratentorial extension even in the case of large cysts, because these lesions expand the subarachnoid spaces and create surgical channel(111).

**Complications:**

Aseptic meningitis is the most dreaded known complication following surgery for epidermoid cysts (3,12,15,22,28,33,62,84,117,119). The capsule of an epidermoid cyst is the living portion of the tumor. Viable portions of capsule that remain after surgical excision is likely to re-grow and continue to generate the breakdown products of epithelial desquamation which is rich in cholesterol and fat tissue. This material is known to be irritant to the meninges and can cause meningeal irritation which is usually transient but partially uncapsulated tumor is fragile and these materials may exude in to subarachnoid space. It can also settle down to perivascular space (28,60). The responsible factor for adherence may be spontaneous rupture during tumor growth, which may occur during surgery or spontaneously and when it occurs, the content may spread along the subarachnoid space. The presence of tumor content in the subarachnoid space may induce complications, caused mainly by the inflammatory effects of the cholesterol breakdown products. This can lead to a meningeal inflammatory reaction, obstruction of CSF flow, or irritation of vascular and neural structures (15). Aseptic meningitis is the most frequent complication caused by the meningeal inflammatory reaction,

which also may lead to fibrosis around the cranial nerves and spinal nerve roots. Martin et al (84) suggested that occlusion of the dural sinuses lead to dural thickening. Granulomatous aseptic meningitis can also impair normal CSF flow and causes hydrocephalus.

Mollaret's recurrent aseptic meningitis (MM) is characterized by short attacks of meningeal irritation and fever (3,12,28,33,117,119). The course of MM is self-limiting. The attacks resolve spontaneously and without any sequel (119). Symptom-free intervals observed in such patients can be as brief as few days or as long as several years (12). CSF examination remains the sole diagnostic modality of MM (119). The CSF shows markedly increased cellularity with pleocytosis with initial polymorphonuclear, and subsequent mononuclear predominance (12,15). CSF examination of MM presents a spectrum of cytomorphologic features (117). Large endothelial cells with indistinct cytoplasm (Mollaret's cells) are typically present in the CSF. Mollaret originally described the large mononuclear cells - the so-called "Mollaret cells" seen in the CSF. Subsequent ultrastructural and immunocytochemical studies support a monocyte/macrophage lineage for these cells (33,117). It is suggested that cholesterol-containing debris

entering the subarachnoid space from the tumor cysts provoke an aseptic “chemical” meningitis of short course (15,119).

The incidence of chemical meningitis is less frequent in patients in whom the tumor is excised more completely, the keratinaceous debris is irrigated, and steroid therapy is used prophylactically (15,22,112). The main goal for treatment of MM is protection of neighbouring structures – covering them with cotton strips to avoid this complication is a standard method (12). However, despite profuse saline irrigation during surgery, the patient may develop aseptic meningitis during the postoperative course. Corticosteroid therapy is effective in all cases inducing a complete or partial remission of the neurological symptoms and signs. Some authors stress that radical tumor removal and profuse irrigation of the operative field with hydrocortisone before the dura is closed may wash out tumor debris and avoid postoperative aseptic meningitis. As steroid irrigation significantly decreased the peroperative morbidity of epidermoid tumor resection, indications for intravenous steroids may become more limited, thereby reducing cost (15,62).

Recurrences have been noted following incomplete excision of epidermoid cysts (1,111). Rutherford et al (108) have commented that early recurrence of an epidermoid, even in the absence of overt evidence

of infection, should produce a high index of suspicion that there may be a low-grade infective cause. The authors had noted coagulase-negative *Staphylococcus* growth after prolonged culture in an enrichment medium from a recurrent CP angle epidermoid cyst.

Primary intracranial squamous cell carcinoma is rare, with most cases arising from a preexisting benign epidermoid cyst (2,4,18,49,52,68,70,85,95,96,113,125,128). Kim et al (68) who reported a CP angle squamous cell carcinoma have speculated that the squamous cell carcinoma may have developed secondary to a chronic inflammatory response by the adjacent epidermoid cyst. Protracted chronic inflammation may predispose to the development of squamous cell carcinoma, as this form of cancer may be encountered in the margins of long standing ulcers and draining sinuses and in burn scars. Rare cases of leptomeningeal carcinomatosis has been reported (52,67). In their series of intra-ventricular epidermoids, Meng et al (87) reported that subtotal removal carries a high incidence of recurrence with the added rare possibility of carcinomatous degeneration of the tumor remnants.

### **Results of surgery:**

Yasargil et al (133) reported forty-three patients with intracranial intradural epidermoid tumors who underwent radical surgical resection. Supratentorial tumors were excised by the pterional (frontosphenotemporal) approach, mesencephalic tumors by either a supratentorial posterior interhemispheric transtentorial approach or an infratentorial/supracerebellar method, and posterior fossa tumors by either a medially or laterally positioned suboccipital osteoplastic craniotomy. One epidermoid tumor was subtotally resected because of dense adhesion to vital structures; the remaining were completely excised. The most frequent complications were aseptic/chemical meningitis and transient cranial nerve palsies. There were no peri-operative deaths. Mean follow-up was 5.2 years. Eighty-six percent of patients reported good to excellent results. No patient had experienced symptomatic or radiographic evidence of recurrence.

Yamakawa et al (132) reported thirty-three cases of intracranial epidermoid tumors treated over 25 years. In 28 patients, (84.9%), the tumor was removed totally or subtotally. Most of the patients could lead

an independent and useful life after operation (93.1%). Among the 29 patients in a long-term follow-up survey, seven tumors recurred after an average interval of 8 years and 10 months (from the first to second operation) and 12 years and 6 months (from the second to third operation). Patients with recurrent tumors were successfully treated, and excellent functional prognosis was observed even after the second or third operation. The 20-year survival rate was 92.8% (Kaplan-Meier method). Yamakawa's study (132) demonstrated that most of the patients, including those with recurring tumors, lead an independent useful life after surgery and that the cumulative 20-year survival rate reached 92%.

Madjid Samii et al (111) reported 40 patients with epidermoid cysts of the cerebellopontine angle (CPA) underwent surgery between 1980 and 1993. Total resection was achieved in 30 cases (75%); in 10 cases (25%) parts of the cyst capsule were left because they adhered to the brainstem and vascular structures of the CPA. One patient with very large bilateral epidermoid cysts, who underwent complete bilateral resection in one stage, died of pulmonary aspiration and infection. As of their latest clinical and radiological follow-up examinations (mean 5.7

years), 93% of the patients are able to lead useful lives. Three cases of cyst regrowth have been observed thus far.

Talacchi et al (120) reported 28 patients with epidermoid cysts of the posterior fossa underwent surgery between 1976 and 1996. Total resection was achieved in 57% of the cases. 12 patients (35%) from subtotal group developed regrowth out of which 30% were symptomatic and reoperated after 8.1 year. 5% of total removal group showed asymptomatic regrowth are on follow up. In 10 cases (25%) parts of the cyst capsule were left because they adhered to the brainstem and vascular structures of the CPA. One patient (3%) died in the perioperative period.

Schiefer et al (114) reported 24 patients with epidermoid cysts of the CPA underwent surgery between 1985 and 2005. Total removal was achieved in 13 patients, near total removal in 6 patients, and subtotal removal in 5 patients. Patients who underwent total removal had a median MRS score of 0, whereas those who underwent near/subtotal removal had a median MRS score of 1. The rate of recurrence was 23% in tumors considered totally removed and 27% in those near/ subtotally removed. Of the 6 patients with recurrences, 5 underwent a second operation. The mean duration of follow-up was 4.2 years.

Similar results have been published by other authors too, which have been summarized in Table 13.

Controversy exists regarding the extent of removal of epidermoid tumors(50). The tendency of the epidermoid tumors to creep and spread can make total surgical excision through a single corridor difficult. Similarly they tend to engulf vessels and attempts at total surgical excision can result in vascular injury. The capsule is often adherent and peeling of the capsule to ensure total excision can again result in injury to underlying neurovascular structures. If not excised totally, epidermoids carry the risk of recurrence, albeit slowly (102). Role of surgery is therefore to attain delicate balance – as totalexcision as possible without producing fresh neurological deficits. Role of radiotherapy for recurrences is controversial although Parikh et al (102) have found it to be beneficial.

Location of tumor affects the outcome - the functional outcome is good for those tumors located in areas amenable for total resection; in our series too the outcome is better in those patients where tumor is restricted to CPA and CPA with transtentorial extension.

## **AIM OF THE STUDY**

The aim of this study was to retrospectively study posterior fossa epidermoids treated surgically at our institute and to know their long term outcome of treatment.

## **MATERIALS AND METHODS**

Between January 1997 to December 2007, 50 consecutive patients with posterior fossa epidermoid cysts were operated on in the Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram. Clinical and surgical records, radiological studies, and slides of these cases have been retrospectively reviewed.

The clinical course was compared within each group. Presenting symptoms, onset of the same, and Neurological signs preoperatively and postoperatively and at the time of follow up recorded.

All of the patients were preoperatively evaluated with either nonenhanced and enhanced computerized tomography (CT) or Magnetic resonance (MR) imaging or both. MR diffusion weighted sequence with Absolute diffusion coefficient (ADC) was used for postoperative radiological follow up.

Maximum diameter measurements cannot be obtained for epidermoids because of their expansive growth along the least resistant

planes, often with a proteiform shape; therefore, a classification including either location or extension is required. The following assessment was determined using computed tomography (CT), magnetic resonance imaging (MRI), and operative records. Primary locations include the cerebellopontine angle (CPA), the fourth ventricle and other miscellaneous include cerebellar convexity. CPA epidermoids were observed to extend into one or more anatomic regions, hence further classified in five subgroups as per Madjid Samii classification(111) as mentioned in review of literatureTable 3.

Computed tomography characteristics reviewed included tumor density and homogeneity, presence and type of contrast enhancement, and calcifications. MRI examination was performed on different imaging units, using a field strength ranging between 0.5 and 1.5 tesla. The magnetic resonance (MR) image parameters used for this study included signal homogeneity and intensity (on proton-density [PD], T1-, and T2-weighted images), presence and type of contrast enhancement, calcifications, and lesion margins, characteristic of the tumor on Diffusion weighted (DWI) and Absoluted diffusion coefficient (ADC) sequences.

In all patients, radical excision was attempted. Using microsurgical techniques, the surgical strategy adopted was to follow the path created by the tumor via a single simple route. In all patients surgical field and the adjacent cisterns were irrigated with hydrocortisone solution (16) at the end of the procedure, and the patients were maintained peri- and postoperatively on steroids (Alternate Inj. Dexamethasone 4mg and Hydrocortisone 100mg intravenously every 6 hourly) (22,111)

Operative data, including surgical approach, extent of removal, morbidity, and mortality, were evaluated on both location and extension criteria. The completeness of cyst removal was assessed intraoperatively and with postoperative CT and MR imaging. Extent of removal divided in to total removal, subtotal removal group. Subtotal removal further classified in to residual lesion 'A' and residual adherent capsule 'B'. Each patient's functional status was determined at the time of discharge from the hospital according to Glasgow outcome score(GOS). Radiological follow-up studies were performed at intervals of 1 to 2 years and at last follow up by means of CT and/or MR imaging.

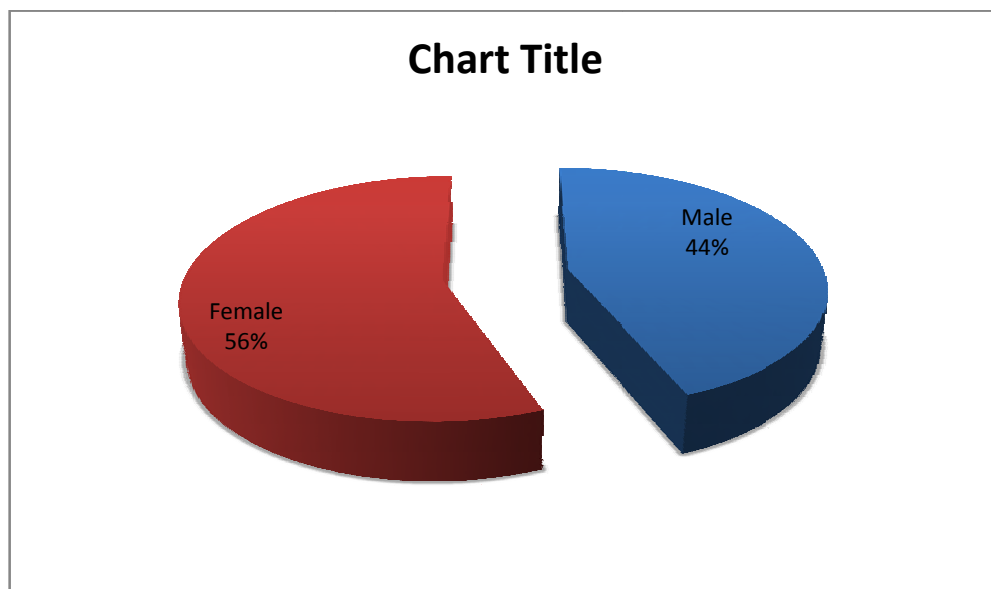
Long-term follow-up, both clinical and radiological, was analyzed, and Chi-square test performed to compare the rate of

recurrence and rate of second surgery for recurrence in total removal and subtotal removal group. Problems such as radiological detection and surgical attitude concerning recurrences were discussed.

## RESULTS

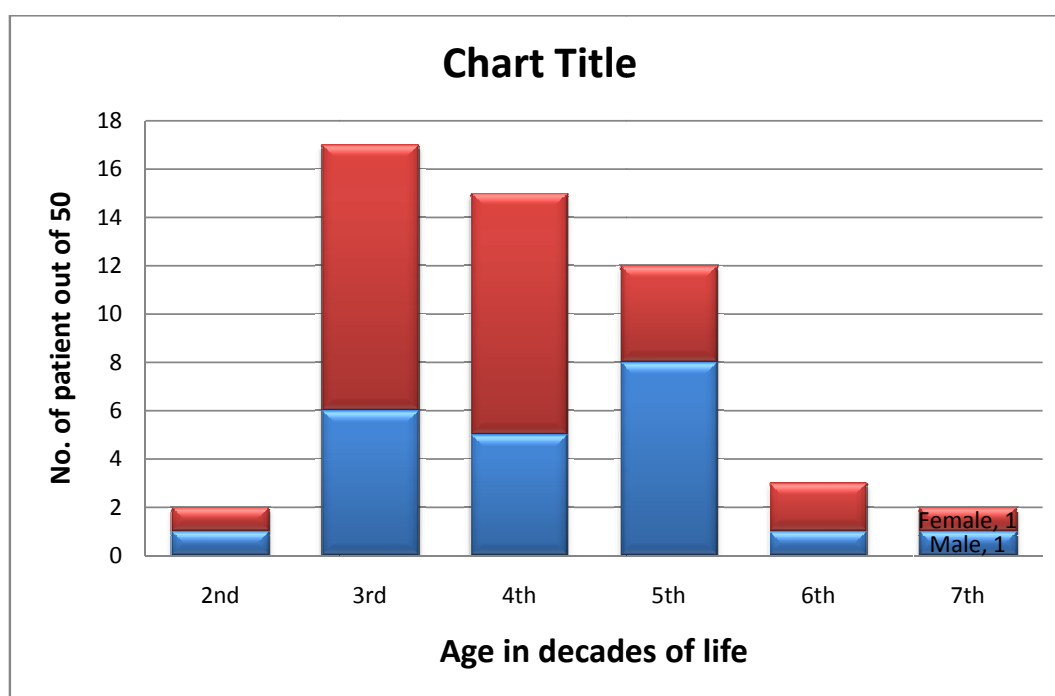
### Patient profile:

There were 28 females (56%) and 22 males (44%), with female to male ratio of 1:1.27.



**Figure 7:** Pie chart showing the skewed sex ratio

The average age of the patients was 37.8 years, with a range of 19 to 66 years. Majority of the patient presented in third and fourth decade of their life (Figure 8).



**Figure 8:** Age of presentation.

### **Clinical features:**

There were 36 patients with CPA epidermoid, 13 patients with fourth ventricle epidermoid and 1 with lateral cerebellar convexity epidermoid. Presenting clinical features are summarized in Table 5,6,7&8. The interval between onset of symptoms and diagnosis ranged between 1 month and 10 years (mean duration of presentation 2 year

6month). Presenting symptoms and signs on admission in patients with CPA epidermoid are listed in Table 5. In most of the patients with CPA tumors (n = 36), the first symptoms were subjective trigeminal neuralgia, hearing loss, and gait disturbance. Trigeminal neuralgia was seen in 13 cases(36%), Hearing loss was noted in 11(30.5%) and gait disturbance was seen in 9 cases(25%). Only in patients with epidermoids involving the middle fossa and supratentorial extension clinical onset was different(seizure and diplopia). Two of our CPA+Middle fossa extension patient had presented with seizure and headache. One patient had predominant complain about diminution of vision.

| <b>Symptoms</b>             | <b>No of Pt with CPA epidermoid<br/>n=36(%age)</b> |
|-----------------------------|----------------------------------------------------|
| <b>Gait disturbance</b>     | <b>9(25)</b>                                       |
| <b>Trigeminal neuralgia</b> | <b>13(36)</b>                                      |
| <b>Facial weakness</b>      | <b>5(13.8)</b>                                     |
| <b>Facial spasms</b>        | <b>4(11)</b>                                       |
| <b>Hearing loss</b>         | <b>11(30.5)</b>                                    |
| <b>Diplopia</b>             | <b>5(13.9)</b>                                     |
| <b>Vision deterioration</b> | <b>1(2.8)</b>                                      |
| <b>Facial numbness</b>      | <b>7(19.4)</b>                                     |

|                                |                |
|--------------------------------|----------------|
| <b>Seizure</b>                 | <b>2(5.6)</b>  |
| <b>Lower cranial symptoms</b>  | <b>5(13.9)</b> |
| <b>Hemiparesis</b>             | <b>4(11)</b>   |
| <b>Hemisensory anaesthesia</b> | <b>1(2.8)</b>  |

**Table 5:** Presenting Symptoms Related to CPA epidermoid (n=36)

Eidermoids extending to parasellar region tend to express supratentorial symptoms, whereas epidermoids extending to mesencephalic region were characterized by a symptoms related to cranial nerve III, IV, VI, V and brain stem compression(Table 5). The prevalent presenting symptom headache and gait ataxia were commonly observed in fourth-ventricle epidermoids (Table 6). Headache and gait ataxia were seen with equal frequency i.e. 69.2%(9patient).

| <b>Symptoms</b>               | <b>No of Pt with 4<sup>th</sup> ventricle epidermoid n=13 (%age)</b> | <b>Lateral cerebellar region(n=1)</b> |
|-------------------------------|----------------------------------------------------------------------|---------------------------------------|
| <b>Gait disturbance</b>       | <b>9(69.2)</b>                                                       | <b>1</b>                              |
| <b>Facial weakness</b>        | <b>3(23.1)</b>                                                       | <b>1</b>                              |
| <b>Hearing loss</b>           | <b>1(7.6)</b>                                                        | <b>--</b>                             |
| <b>Diplopia</b>               | <b>4(30.1)</b>                                                       | <b>--</b>                             |
| <b>Lower cranial symptoms</b> | <b>3(23.1)</b>                                                       | <b>--</b>                             |
| <b>Hemiparesis</b>            | <b>2(15.4)</b>                                                       | <b>--</b>                             |
| <b>Headache</b>               | <b>9(69.2)</b>                                                       | <b>--</b>                             |

|                                                                  |                |          |
|------------------------------------------------------------------|----------------|----------|
| <b>Cerebellar signs (e.g. dysmetria, nystagmus, gait ataxia)</b> | <b>8(61.5)</b> | <b>1</b> |
|------------------------------------------------------------------|----------------|----------|

**Table 6:** Presenting symptoms of Fourth ventricle(n=13) and lateral cerebellar region epidermoid(n=1).

Although the type and incidence of neurological signs varied according to location and extension, the predominant signs were cranial nerve palsy and cerebellar impairment. In CPA epidermoids, the most common single dysfunction was either fifth cranial nerve(n=22), seventh cranial (n=14) or eighth cranial nerve (n=12). Dysmetria seen in 7 patients, and abducent nerve palsy 3 patient (Table 7).

The highest rate of cerebellar signs, 61.5%(8 patients) was demonstrated in fourth-ventricle epidermoids. One patient with fourth ventricle epidermoid had bilateral lateral rectus palsy(Table 8).

| <b>Neurological signs in CPA epidermoid</b> | <b>Preop Abnormality(%)</b> |
|---------------------------------------------|-----------------------------|
| <b>II</b>                                   | <b>1(2.8)</b>               |
| <b>III</b>                                  | <b>2(5.6)</b>               |
| <b>V</b>                                    | <b>22(61.1)</b>             |
| <b>VI</b>                                   | <b>3(8.3)</b>               |
| <b>VII</b>                                  | <b>14(38.9)</b>             |
| <b>VIII</b>                                 | <b>12(33.3)</b>             |
| <b>IX, X</b>                                | <b>8(22.2)</b>              |

|                                                                  |                |
|------------------------------------------------------------------|----------------|
| <b>XII</b>                                                       | <b>1(2.8)</b>  |
| <b>Cerebellar signs (e.g. dysmetria, gait ataxia, nystagmus)</b> | <b>7(19.4)</b> |

Table 7: Surgical Results Related to Preoperative Status in Cerebellopontine Angle epidermoids

Lateral cerebellar convexity epidermoid which was seen in one of the patient had predominantly cerebellar signs and symptoms with VIIth nerve involvement.

| <b>Neurological signs in<br/>Fourth ventricle epidermoid</b>     | <b>Preop Abnormality(%)</b> |
|------------------------------------------------------------------|-----------------------------|
| <b>VI</b>                                                        | <b>4(30.8)</b>              |
| <b>VII</b>                                                       | <b>3(23.1)</b>              |
| <b>VIII</b>                                                      | <b>1(7.7)</b>               |
| <b>IX, X</b>                                                     | <b>3(23.1)</b>              |
| <b>Cerebellar signs (e.g. dysmetria, gait ataxia, nystagmus)</b> | <b>8(61.5)</b>              |
| <b>Papilloedema</b>                                              | <b>7(53.8)</b>              |

Table 8: Surgical Results Related to Preoperative Status in Fourth ventricle epidermoids(n=13)

### **Neuroimaging:**

6 patients were evaluated with CT scan alone, 14 patients were evaluated with MRI alone and remaining 30 had got both CT scan as well as MR Imaging done. CT revealed all 36 tumors to be hypodense

with respect to brain tissue. A small peripheral calcification was demonstrated in 10%, whereas subtle capsule-like enhancement was observed in 8%. In 72%, a homogeneous structure was observed in the tumors, even though precise evaluation of this parameter was often impaired because of the poor technical quality of the films available.

Whereas MR T1-weighted and T2-weighted images showed epidermoids as hypointense and hyperintense respectively, compared with cerebrospinal fluid (CSF) and brain tissue, Fluid attenuated inversion recovery (FLAIR) images proved more useful for diagnostic evaluation. Other features included a slightly nonhomogeneous structure 54%(n=23) and regular margins 64% (n=28). 20 patient also had Diffusion weighted(DWI) sequence with absolute diffusion coefficient. All of our patient showed DWI restriction. Calcifications and contrast-enhancement patterns were as revealed by CT scan.

| <b>Tumor location</b>    | <b>Subgroup</b>                      | <b>No of patients(n=50)</b> |
|--------------------------|--------------------------------------|-----------------------------|
| <b>CPA lesion (n=36)</b> | <b>CPA alone</b>                     | <b>11</b>                   |
|                          | <b>CPA+ transtentorial extension</b> | <b>10</b>                   |
|                          | <b>CPA+Middle fossa extension</b>    | <b>7</b>                    |
|                          | <b>CPA+Foramen magnum extension</b>  | <b>3</b>                    |

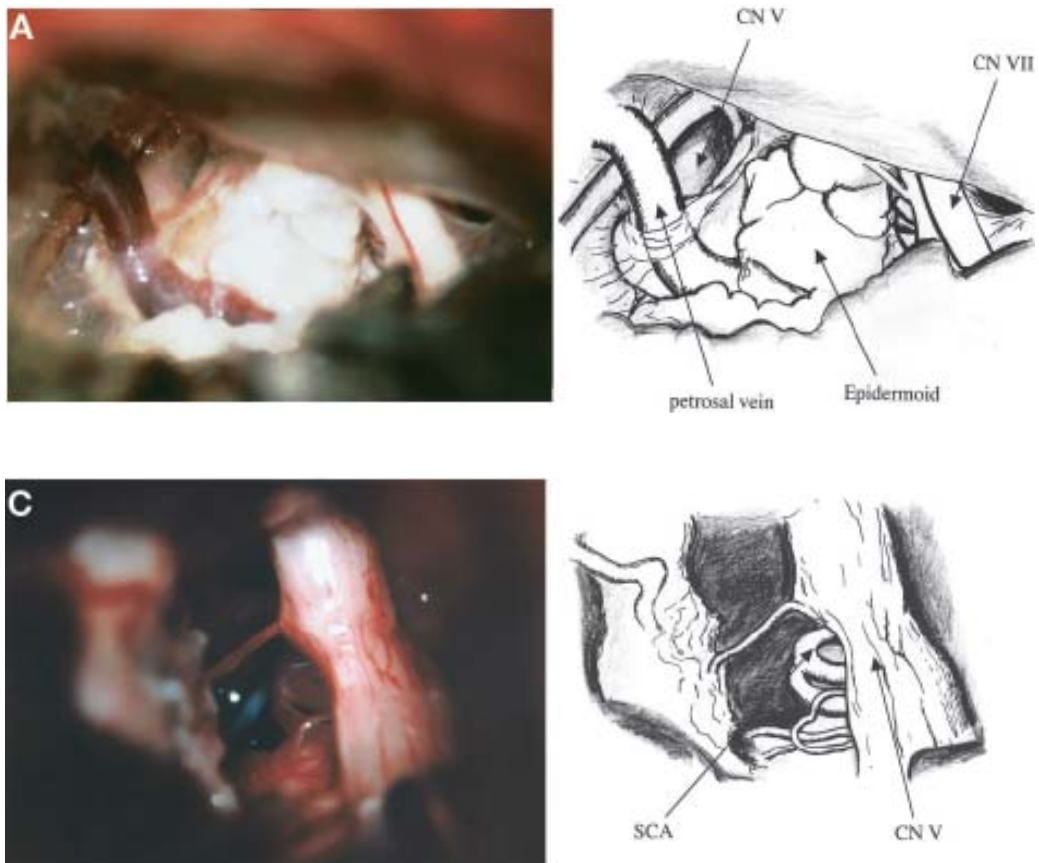
|                                              |           |
|----------------------------------------------|-----------|
| <b>CPA+Transtentorial &amp; FM extension</b> | <b>5</b>  |
| <b>Fourth ventricle</b>                      | <b>13</b> |
| <b>Right lateral cerebellar convexity</b>    | <b>1</b>  |

**Table 9:**Location and extension of epidermoid cysts in 50 patients\*

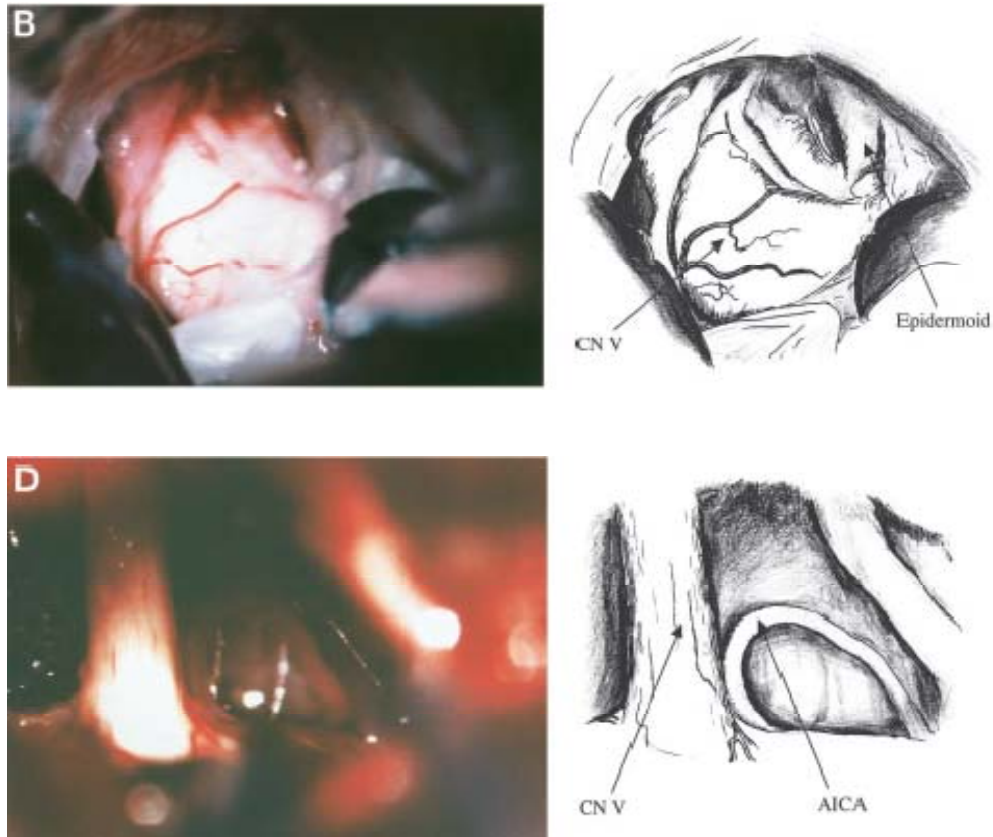
Posterior fossa epidermoid base on our finding were located at CPA, fourth ventricle and right cerebellar convexity. Concerning tumor location, CPA lesion where subdivided in 5 groups depending on there extension. Location and extension of epidermoid cystsare as shown in Table 9.

### **Surgery:**

In addition to the 50 primary surgeries in our patient population, 3 patients required a second surgery because of recurrence, making a total of 53 operative events. The selected surgical approachesinclude standard retrosigmoid suboccipital craniectomy(RMSOC), Frontotemporal craniotomy and subtemporal approach i.e., middle fossa approach (MFA), combined subtemporal middle fossa and retromastoid suboccipital approach i.e., combined approach (CA) for CPA epidermoid and Midline suboccipital craniectomy (MSOC) for fourth ventricle epidermoid. Right cerebellar convexity epidermoid was operated by retromastoid suboccipital approach Table 10.



**Figure 14- I.** Photographs (*left*) and drawings (*right*) illustrating the anatomy involved in epidermoid resection. ‘A’ before and ‘B’ after excision.



**Figure 14-II.** Photographs (*left*) and drawings (*right*) illustrating the anatomy involved in epidermoid resection. ‘C’ before and ‘D’ after excision of epidermoid in 2 of our CPA epidermoid tumor.

Complete removal was achieved in 31 patients (62%). Of the remaining 19 patients, there were 13 patients with capsule fragments attached to neurovascular structures for fear of increasing neurological

deficits. In the other 6 patients, the tumor remnants, which were distant from the primary tumor location hence could not be reached, or escaped/hiding. The highest rate of total tumor removal (eight of eleven tumors) was for epidermoids confined to the CPA, and for CPA with transtentorial tumor (eight of ten tumors). Rate of tumor removal in different subgroup is summarized in Table 10.

| <b>Tumor Location</b>                        | <b>No of Patient</b> | <b>Surgical Approach</b> | <b>Total Removal</b> | <b>Subtotal removal (A+B)</b> | <b>Residual lesion (A)</b> | <b>Residual adherent capsule (B)</b> |
|----------------------------------------------|----------------------|--------------------------|----------------------|-------------------------------|----------------------------|--------------------------------------|
| <b>CPA alone</b>                             | <b>11</b>            | <b>RMSOC</b>             | <b>8</b>             | <b>3</b>                      | <b>0</b>                   | <b>3</b>                             |
| <b>CPA+ transtentorial extension</b>         | <b>10</b>            | <b>RMSOC</b>             | <b>8</b>             | <b>2</b>                      | <b>1</b>                   | <b>1</b>                             |
| <b>CPA+Middle fossa extension</b>            | <b>7</b>             | <b>2 MFA+<br/>2 CA</b>   | <b>5</b>             | <b>2</b>                      | <b>1</b>                   | <b>1</b>                             |
| <b>CPA+Foramen magnum extension(FM)</b>      | <b>3</b>             | <b>RMSOC</b>             | <b>1</b>             | <b>2</b>                      | <b>0</b>                   | <b>2</b>                             |
| <b>CPA+Transtentorial &amp; FM extension</b> | <b>5</b>             | <b>RMSOC</b>             | <b>1</b>             | <b>4</b>                      | <b>3</b>                   | <b>1</b>                             |
| <b>Fourth ventricle</b>                      | <b>13</b>            | <b>MSOC</b>              | <b>7</b>             | <b>6</b>                      | <b>1</b>                   | <b>5</b>                             |
| <b>Right cerebellar convexity</b>            | <b>1</b>             | <b>RMSOC</b>             | <b>1</b>             | <b>0</b>                      | <b>0</b>                   | <b>0</b>                             |
| <b>Total</b>                                 | <b>50</b>            | <b>--</b>                | <b>31</b>            | <b>19</b>                     | <b>6</b>                   | <b>13</b>                            |
| <b>Long term</b>                             | <b>16/38</b>         | <b>--</b>                | <b>2/22</b>          | <b>14/16</b>                  | <b>5</b>                   | <b>9</b>                             |

|                            |          |           |          |          |          |          |
|----------------------------|----------|-----------|----------|----------|----------|----------|
| <b>Recurrence</b>          |          |           |          |          |          |          |
| <b>Out of 38 patients</b>  |          |           |          |          |          |          |
| <b>Recurrence operated</b> | <b>3</b> | <b>--</b> | <b>0</b> | <b>3</b> | <b>2</b> | <b>1</b> |

**Table 10:** Extent of Removal Related to Tumor Location and Extension

(n=50) and Long term recurrence and re-explorations(n=38)

### **Complications:**

Early postoperative neurological deterioration, although minimal and transient, occurred in most of the patient particularly seventh nerve paresis. Eleven patients apart from 14 patient with CPA epidermoid who had seventh nerve paresis, developed new onset deterioration of its function, however on follow up on 13 patient out of these 14 patient had residual weakness. Four patient developed lower cranial dysfunction and required RT feedings, 1 patient required tracheostomy. 2 patients from fourth ventricle epidermoid developed postoperative VIIth nerve paresis. One patient with lateral cerebellar convexity epidermoid did not develop any new deficit and on follow up his symptoms completely improved. At an intermediate follow-up, many dysfunctions had cleared, resulting in a notable functional improvement Table 11.

One patient died intraoperatively due to massive myocardial infarction and surgery could not be completed. Another patient with

CPA tumor was brought with poor GCS underwent retromastoid suboccipital approach and subtotal excision. Post operatively patient required tracheostomy and mechanical ventilation. Patient was gradually weaned of ventilator however remained in vegetative state and was discharged from hospital. Patient succumbed after 14 months following surgery due to respiratory complication.

One patient required a ventriculoperitoneal CSF shunt for hydrocephalus. One patient CPA epidermoid developed CSF

| <b>Neurological signs in CPA epidermoid</b>                      | <b>Preop Abnormality(%)</b> | <b>Postop Abnormality(%)</b> |
|------------------------------------------------------------------|-----------------------------|------------------------------|
| <b>II</b>                                                        | <b>1(2.8)</b>               | <b>0</b>                     |
| <b>III</b>                                                       | <b>2(5.6)</b>               | <b>0</b>                     |
| <b>V</b>                                                         | <b>22(61.1)</b>             | <b>3(8.3)</b>                |
| <b>VI</b>                                                        | <b>3(8.3)</b>               | <b>3(8.3)</b>                |
| <b>VII</b>                                                       | <b>14(38.9)</b>             | <b>13(36.7)</b>              |
| <b>VIII</b>                                                      | <b>12(33.3)</b>             | <b>6(16.7)</b>               |
| <b>IX, X</b>                                                     | <b>8(22.2)</b>              | <b>0</b>                     |
| <b>XII</b>                                                       | <b>1(2.8)</b>               | <b>0</b>                     |
| <b>Cerebellar signs (e.g. dysmetria, gait ataxia, nystagmus)</b> | <b>7(19.4)</b>              | <b>1(2.8)</b>                |

**Table 11:** Surgical Results Related to Preoperative Status in Cerebellopontine Angle epidermoids .

rhinorrhoea after retromastoid suboccipital approach and required thecoperitonal shunt. One patient developed Pseudomeningocele and followed by CSF leak and managed with lumbar drain for 5days. Exact data regarding incidence of aseptic meningitis could not be found out due to lack of documentation.

| Neurological signsin<br>Fourth ventricle epidermoid          | Preop Abnormality(%) | Postop<br>Abnormality(%) |
|--------------------------------------------------------------|----------------------|--------------------------|
| VI                                                           | 4(30.8)              | 3(23.1)                  |
| VII                                                          | 3(23.1)              | 5(38.5)                  |
| VIII                                                         | 1(7.7)               | 0                        |
| IX, X                                                        | 3(23.1)              | 2(15.4)                  |
| Cerebellar signs (e.g. dysmetria,<br>gait ataxia, nystagmus) | 8(61.5)              | 5(38.5)                  |
| Papilloedema                                                 | 7(53.8)              | NA                       |

**Table 12:** Surgical Results Related to Preoperative Status in Fourth ventricle epidermoids(n=13)

### **Long term follow up:**

Out of 50 patientsone patient expired intraoperatively due to massive myocardial infarction, another patient who had been operated with poor GCS continued to remain in vegetative state and died after 14 month. 4 patients never turned up after being discharged from hospital

with GOS of 5 and 6 patient lost to follow up after one year of surgery. At last follow up there GOS were 5.

Remaining 38 patients were included for long term outcome assessment, 22 were from total removal group and 16 were from subtotal removal group. The average follow-up period was 9.4 years(Range 4.8yr to 15.3yr). Patients who underwent subtotal removal had a longer follow-up period than those obtaining total removal (9.3 and 8 yr, respectively).

At the time of last follow up 16 out of 38 patient (42%) showed evidence of tumor regrowth(5 residual lesion 'A' growth, 9 tumor capsule residual 'B' growth and 2 from total removal group) Table 7. Three patients (7.9 %) were operated the second time for progressive neurological deterioration, after mean follow up of 10.9 yr and are doing well after second surgery(GOS= 5)(Table 10).

After supposedly total removal, two patients experienced recurrence; however both the patients are asymptomatic. One patient belonged CPA group and asymptomatic after 10.7 year and other from fourth ventricle group asymptomatic after 8 year. Rest of the other patients with tumor regrowth are asymptomatic after 7 years (Table 10).

Analysis of our data using Chi-square test, comparing long term recurrence rate in total removal group and subtotal removal group, long term recurrence rate of total removal group is significantly less with P value=0.0057 ( $P < 0.05$ ). However same for the second surgery of recurrence P value = 0.18 i.e., not significant ( $P > 0.05$ ).

## **DISCUSSION:**

Epidermoid cysts are uncommon congenital lesions belong to the spectrum of “tumors of disordered embryogenesis” and are known to occur at diverse locations in the neuraxis. Epidermoids represent 0.2 to 1% of all primary intracranial tumors (107). These slow-growing benign tumors commonly occur in the parapontine or parapituitary regions. Cerebellopontine angle(CPA) epidermoids constitute 40% of all intracranial epidermoids (100) and are of considerable surgical interest. Epidermoid tumors have a thin capsule composed of keratinized stratified squamous epithelium, which desquamates and gives rise to the cyst contents, keratin, and cholesterol (107). Epidermoids presents with a distinct growth pattern, i.e., they "flow" across the basal surface of the brain into the subarachnoid space and only in later stages of growth do they behave as expanding lesions, remaining clinically silent for many years (9,13,106,129).

## **DEMOGRAPHICS:**

Current study comprised 28 females (56%) and 22 males (44%), with female to male ratio of 1:1.27. On comparison with the previously done studies we found that Schiefer and Link et al (114) in also found slight female preponderance, Kobata et al 69 , Talacchi et al (120) , Madjid Samii et al (111) in their study also noted female predominance. Our finding was in concordance with these study finding. The average age of the patients was 37.8 years, with a range of 19 to 66 years. Majority of the patient presented in third and fourth decade of their life.

Schiefer and Link et al (114) study showed mean age at the time of diagnosis of 38.8 years; range, 7 to 73 years. In Madjid Samii et al s study (111) patient's age ranged from 19 to 66 years with mean of 42 years. In Talacchi et al (120) study patients were ranging in age from 18 to 65 years (mean age, 45.8 yr). All of these study were confirmed our finding of epidermoid tumor presenting most commonly in third and fourth decade of life.

## **CLINICAL FEATURES:**

The mean duration from onset of symptoms to diagnosis was 2.5 year (range 1 month to 10 year) however since our study had 36 patient with CPA and 13 patient with fourth ventricular lesion and 1 patient

with lateral cerebellar convexity tumor. On further analyzing we found Mean duration of presentation of CPA group to be 2.8 year and that of fourth ventricle group, 1.6 year.

Comparing our data with existing literature of CPA epidermoid Schiefer and Link et al found mean duration from onset of symptoms to diagnosis was 3.1 years (range, 1 month-18 years). Madjid Samii et al (111) reported

Mean duration was 42 years (range 19 to 66 years). On average, the onset of the symptoms occurred 11.4 years (range, 2–28 yr) before the initial operative procedure as reported by Kobata et al (69). This exceptionally delayed presentation probably may be due selection bias. This study included only those patients who presented with trigeminal neuralgia or hemifacial spasms. Our results were matching the above two study group. Although our patient has slightly earlier presentation but it can be explained by technological advancement and easy availability of health care services and relatively older previously done studies. Tancredi A et al 121 found in their study of 9 fourth ventricle epidermoid, duration of clinical history ranged from 2 months to 6 years (mean: 2.2 years). Nassar et al 92 reported 4 cases with duration of symptoms from 1month to 2 year (mean 0.6 year). Our results of fourth ventricle were in between these ranges.

In most of the patients with CPA tumors ( $n = 36$ ), the first symptoms were trigeminal neuralgia, hearing loss, and gait disturbance. Trigeminal neuralgia was seen in 36%( $n=13$ ), Hearing loss was noted in 30.5%( $n=11$ ) and gait disturbance was seen in 25%( $n=9$ ). Only in patients with epidermoids involving the middle fossa and supratentorial extension was the clinical onset different (seizure and diplopia). Parasellar extended epidermoids tended to express supratentorial symptoms, whereas mesencephalic extended epidermoids were characterized by a higher disturbance rate, especially regarding the brain stem. Scheifer and Link et al (114) showed headache, eighth cranial nerve dysfunction and followed by fifth cranial nerve dysfunction as a common presentation. Kobata et al (69) performed a meta-analysis of 263 cases of CPA epidermoid tumors, which showed that hearing loss (37.6%), trigeminal neuralgia (29.7%), dizziness or vertigo (19.4%), facial palsy (19.4%). Madjid Samii et al (111) reported hearing loss followed by dizziness, gait unsteadiness as commonest symptoms.

The prevalent presenting symptom headache and gait ataxia were commonly observed in fourth-ventricle epidermoids. Headache and gait ataxia were seen with equal frequency i.e. 69.2%( $n=9$ ). Talacchi et al (120) in their study having 5 out of 28 patients, of fourth ventricle epidermoid found 100% patient with cerebral ataxia and 60% with

headache. Nassar et al 92 in found 75% of fourth ventricle epidermoid patient presented with headache and 75% patient with cerebellar ataxia. Therefore our results correlated with these studies.

Although the type and incidence of neurological signs varied according to location and extension, the predominant signs were cranial nerve palsy and cerebellar impairment. In CPA epidermoids, the most common single dysfunction was either fifth cranial nerve 61.1%, seventh cranial 38.9% or eighth cranial nerve 33.3%. Dysmetria 19.4% , and abducent nerve palsy 8.3%. The highest rate of cerebellar signs, 61.5%(n=8) was demonstrated in fourth-ventricle epidermoids. Lateral cerebellar convexity epidermoid which was seen in one of the patient had predominantly cerebellar signs and symptoms with VIIth nerve involvement.

Scheifer and Link et al (114) found, eight cranial nerve dysfunction in 54% and followed by fifth cranial nerve dysfunction in 43% as a common presentation. Kobata et al (69) had 26.7% patient having trigeminal dysfunction. Madjid Samii et al 111 reported eight nerve paresis in 55% followed fifth nerve in 43%. Talacchi et al (120) had 50% patient with eight cranial nerve signs, 45% with facial nerve signs, 50% with cerebella signs, and 25% with fifth nerve involvement

signs. Our result matched with available literature but for sign of fifth nerve dysfunction found in more number of patients. We counted impaired corneal reflex as a sign of fifth nerve dysfunction even though there were no hypesthesia or any motor weakness. Probably we had more patients in CPA group extending in to transtentorial cistern and middle fossa.

### **NEUROIMAGING:**

Confident preoperative radiologic diagnosis of epidermoid tumors has greatly increased since the advent of MRI. On CT scanning, epidermoids typically appear as lowdensity, nonenhancing lesions (32,41,47,72). However, epidermoids may exhibit atypical features and appear hyperdense (53,90), have rim calcification (16,47), or marginally enhance with contrast (53). Our 5 (10%) patient showed calcification at periphery and 4 (8%) patient showed peripheral patchy contrast enhancement.

In Talacchi et al study a small peripheral calcification was demonstrated in only five tumors (18%), whereas subtle capsule-like enhancement was observed in three tumors (10%).Madjid Samii et al found 7.5% of their patient showing cyst wall enhancing on CT scan after administration of contrast.

Schiefer et al (114) in their study found this atypical feature i.e., calcification and enhancement on CT scan in 8% patient. In this regard our data is in concordance with their data.

Magnetic resonance imaging has superseded CT for accurate radiologic diagnosis of these lesions, mainly because diffusion-weighted imaging definitively differentiates tumor from CSF-filled cisterns and cysts (19,31,34,46,99,101,111,126,130,131). Epidermoid tumors generally demonstrate mild hypointensity, typically between CSF and brain parenchyma, on T1-weighted images, and hyperintensity, similar to or greater than CSF, on T2-weighted images and do not enhance with gadolinium (57,93,99,116,122). Diffusion-weighted imaging, while not available for many patients at the beginning of our series, proved extremely useful in diagnosis for those in which it was used and is the optimum way to follow up patients postoperatively for recurrent or residual epidermoid. Our Imaging findings were also no way different from existing literature.

| Authors & Years                      | Period Evaluated | Location Of Epidermoid | No. of cases | Total Removal (%) | Patient Mortality (%) | Cyst Recurrence (%) | Follow Up (yrs) |
|--------------------------------------|------------------|------------------------|--------------|-------------------|-----------------------|---------------------|-----------------|
| Berger & Wilson, 1985                | 1972–1983        | CPA                    | 13           | 0                 | 0                     | 0                   | 4.5             |
| Sabin, <i>et al.</i> , 1987          | 1967–1987        | CPA                    | 20           | 5                 | 5                     | 10                  | 6               |
| Salazar, <i>et al.</i> , 1987        | 1971–1981        | CPA                    | 17           | 0                 | 6                     | 24                  | 6.8             |
| Rubin, <i>et al.</i> , 1989          | 1976–1987        | CPA                    | 7            | 57                | 0                     | 0                   | 4.6             |
| Yamakawa, <i>et al.</i> , 1989       | 1963–1988        | CPA                    | 15           | 47                | 6.6                   | 20                  | 8               |
| Yasargil, <i>et al.</i> , 1989       | 1967–1988        | CPA                    | 22           | 97                | 0                     | 0                   | 5.7             |
| De Souza, <i>et al.</i> , 1990       | 1966–1986        | CPA                    | 30           | 18                | 3.3                   | 13.3                | 5               |
| Lunardi, <i>et al.</i> , 1990        | 1951–1988        | CPA                    | 17           | 35                | 12                    | 30                  | 9               |
| Samii, <i>et al.</i> , 1996          | 1980–1993        | CPA                    | 40           | 75                | 2.5                   | 7.5                 | 5.7             |
| Doyle and de la Cruz, 1996           | 1978-1993        | CPA                    | 13           | 54                | --                    | 31                  | 8.6             |
| Mohanty <i>et al.</i> , 1997         | 1986-1994        | CPA                    | 25           | 48                | --                    | 0                   | 3.5             |
| Brunori <i>et al.</i> , 1997         | 1980-1994        | CPA                    | 12           | 33                | --                    | 17                  | 6.5             |
| Talacchi <i>et al.</i> , 1998        | 1976-1996        | PF                     | 28           | 57                | 3                     | 14                  | 8.6             |
| Kobata <i>et al.</i> , 2002          | 1982-1995        | CPA                    | 30           | 57                | --                    | 10                  | 11.5            |
| Schiefer and Link <i>et al.</i> 2007 | 1985-2005        | CPA                    | 24           | 54                | --                    | 25                  | 4.3             |
| Current study 2012                   | 1997-2007        | PF                     | 50           | 62                | 2                     | 42%                 | 9.4             |

**Table 13:** Review of major reports of CPA epidermoid cysts; TR= Total removal, ( -- ) = clear data not available, CPA= cerebellopontine angle, PF= posterior fossa.

## **SURGERY:**

Tumors confined to the CPA were totally removed with greater success than those lesions that had infiltrated adjacent cisterns. In our series 31 patient (62%) underwent total excision and 19 patients (38%) subtotal removal due either the cyst capsule was strongly adherent to the basilar artery and its branches and to cranial nerves, or tumor was extending to middle fossa and was beyond reach. We compared our surgery results with existing literature, findings are summarized in the Tabel 13.

Madjid Samii et al (111) achieved total excision of CPA epidermoid in 75% of their patients. They further classified CPA in to five group, they achieved 73% in CPA only group, 100% in CPA with transtentorial extension group, 60% with extension to middle fossa group, 78% with extension to foramen magnum and 75% total removal rate in transtentorial and foramen magnum extension.

Talacchi et al achieved total excision of epidermoid in 57% of their patient with posterior fossa epidermoid. In his study 5 patients were having fourth ventricle epidermoid out of which total excision could be achieved in 60%.

Kobata et al (69) achieved total excision of epidermoid in 57% of their reported series. Tancredi et al (121) in their case series of 9 patients

with fourth ventricle epidermoid achieved total removal in 22.2%. Scheifer et al (114) reported 54% total removal rate. On comparing the same with our data i.e., 62%, it falls in between above mentioned rates. Our value for total removal of fourth ventricle epidermoid was 54% which is within the range mentioned in the literature.

### **COMPLICATIONS:**

However, initial neurological impairment remained unchanged, and almost half of the patients suffered mild transient deterioration, mainly related to cranial nerve dysfunctions (Table 11&12). This outcome is consistent with previously reported data (111,120) and can be explained by the widespread diffusion of these tumors and the consequent manipulation of the nervous tissue (132).

Right cerebellar convexity epidermoid sustained positive outcome.

Results of fourth ventricle epidermoid have been compared with various available literature in the Table 14.

Tancredi et al(121) reported 9 cases of fourth ventricle epidermoid. Their total removal rate was 22%. 3 patients (33%) developed recurrence, 1 patient required CSF shunt, and had 2 mortalities. One patient developed vestibulocerebellar symptoms. 2

patients developed IVth cranial nerve palsy post operatively and noticed improvement of gait ataxia.

| Authors & Years      | Period Evaluated | No. of cases | HCP | Total Removal (%) | Patient Mortality (%) | Cyst Recurrence (%) | Follow Up        | Postoperative results and notes                                                                        |
|----------------------|------------------|--------------|-----|-------------------|-----------------------|---------------------|------------------|--------------------------------------------------------------------------------------------------------|
| Tancredi (2002)      | 1975–2000        | 9            | 3   | 22                | 22                    | 33                  | 5yr to 23yr      | 6, asymptomatic; 1, slight vestibulocerebellar syndr; 1, CSF shunt 3, recurrence                       |
| Talacchi (1998)      | 1976–1996        | 5            | 4   | 60                | 20                    | 14                  | 8.6years         | 2, IVth c.n. palsy, improvement of gait ataxia; 2, unchanged;                                          |
| Nassar (1995)        | 39year           | 4            | 2   | 75                | 0                     | --                  | 5month to 13year | 3, asymptomatic; 1, speech and movement disorder; 2, CSF shunt                                         |
| Current study (2012) | 1997-2007        | 13           | 7   | 54                | 0                     | 46                  | 9.3yr            | 2, VIIth c.n. palsy, improvement of gait ataxia, VI, VIII & LCN symptoms; 1, CSF shunt, 1, re-operated |

**Table 14:**Cases of 4th ventricle epidermoid cysts reported since 1974,

HCP Preoperative hydrocephalus;

Talacchi et al (120) in their study of posterior fossa epidermoid, had 5 cases with fourth ventricle epidermoid. They achieved total removal in 60% of patient and found recurrence in 14% patient at mean follow up of 8.6yr.

On comparing the same with our study, having 13 cases belonging to fourth ventricle group, we achieved total removal rate in 7

(54%) patient, and out of remaining 6 patient 5 were from tumor remnant group and 1 from capsule residual group.

Complications other than cranial nerve deficit, cerebellar signs or brainstem signs, such as aseptic meningitis, Mollaret's meningitis, we could not gather exact data due to lack of proper documentation.

Primary intracranial squamous cell carcinoma arising from a preexisting benign epidermoid cyst, or leptomeningeal carcinomatosis is not seen in any of our patients.

#### **LONG TERM OUTCOME:**

Postoperative perspectives because epidermoids are benign in nature, assessment is required not only to determine the extent of surgical removal, but also to monitor tumor recurrences. During the initial follow-up period, the surgeon's assessment of the extent of removal is more reliable than radiological information (79), because the immediate shrinkage of the hypodense and hypointense area on computed tomographic and MR T1-weighted images, respectively, leaves open questions regarding residuals. However, the surgeon's intraoperative judgment may not prove accurate in later follow-up.

As expected, a review of the literature showed that some reports suggested that near/subtotal removal was the best approach, whereas

others showed that total removal was a better strategy (Table 13). For example, Berger and Wilson (16) did not perform total removal in any of their cases, reporting minimum patient morbidity and no recurrences over 4.5 years of follow-up. On the other hand, Yasargil et al (133) achieved total removal in 95% of patients with minimum morbidity and a 9% recurrence rate over 5.2 years of follow-up. Tancredi et al reported 33% recurrence and 11% requiring second surgery for recurrence. All other reports (Table 13) including ours fall somewhere in between these ends of the spectrum except for rate of recurrence. Because there are so many differences and inconsistencies in the methods and data gathered in these series, it is difficult to come to any firm conclusions. One major limiting factor in the current available literature is a limited duration of follow-up. In this study we have managed to achieve long term follow up (Average of 9.4yr). As epidermoid cysts are very slow growing tumors that usually require more than 4 to 5 years of follow up to develop recurrence. However other limitation is lack of a system for grading the complexity of these lesions. There is no available grading system to accurately relate the tumor's extension to adjacent spaces and degree of adherence to neighboring structures. These variables must be taken into account when considering extent of removal and subsequent clinical outcome.

In our study recurrence rate is found to be 42% which is higher as compared to the existing literature (Table 13). On analysing the same, this finding can be explained by the fact that follow up period in our study is longer, and also that most of our subtotally removed tumor belonged to residual lesion 'B' (Table 10).

In spite of supposed total removal, two patients experienced recurrences. Long term recurrence rate found to be significantly low in case of total removal group (9.5%). Long term recurrence for subtotally removed tumor is significantly higher than total removal group. Although not all recurrence are symptomatic/require second surgery.

Patients with asymptomatic recurrences may be followed up by comparing serial neuroimaging studies for differential diagnosis between CSF cyst and tumor regrowth (8). However, both CT and MRI may pose doubts because the brain reexpansion is extremely slow and incomplete.

Three patients (7.8%) required second surgery for symptomatic recurrence after average follow up of 10.9 years. In Talacchi et al (120) reported study 30% were operated on a second time for progressive neurological deterioration, after a mean interval of 8.1 years from subtotal excision. 35% of their subtotally removed epidermoid showed

recurrence. In Tancredi et al (121) reported study of fourth ventricle epidermoid 11% patient were operated on a second time for cystic recurrence diagnosed between 10 and 17 year after primary surgery. In our study rate of second surgery for recurrence is low as compared to the quoted study, reason being majority of their patient presented with recurrence were symptomatic and therefore operated, however in our study most of the recurrence are asymptomatic and were kept under surveillance.

The timing of subsequent surgery is controversial. Surgeons still debate whether second surgery should be performed at the first radiological evidence of recurrence, at the time the tumor presumably extends beyond the original operative field, or after the onset of symptoms. Because of radiological uncertainties, the general attitude is to wait either for clear radiological evidence of recurrence or for the renewal of symptoms (8,16,79,133).

## CONCLUSIONS

Based on our experience there is no compelling evidence to suggest that total removal of epidermoid results in increased morbidity and mortality. On long term follow up there was evidence to suggest that symptomatic recurrence requiring re-exploration occurs after long duration (~9 year). The rate of recurrence is significantly higher after subtotal removal as compared to total removal of epidermoid. Hence, total removal should be the standard goal when operating on posterior fossa epidermoid cysts. However it is essential not to produce new neurological deficits for such a benign lesion where progression free survival is so long. It is preferable to operate on recurrent epidermoid rather than having a patient with poor quality of life indices following total excision.

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## **Proforma for Posterior fossa Epidermoid**

Name

Age

Sex        M / F

Hospital No

### **Presenting complaints**

Raised ICP headache (Yes / No)

Visual deficit (Yes / No)

Diplopia (Yes / No)

Trigeminal neuralgia

Facial numbness

Facial weakness

Hearing loss

Gait disturbance

Lower cranial symptoms

Hemiparesis

Hemisensory anaesthesia

Seizure

Incidentally detected

History of meningitis

Other

### **Examination**

Higher mental function

Fundus (normal / papilloedema/ optic atrophy)

Visual acuity(specify) Visual field (specify)

EOM (full / restriction)

IIIrd nerve Paresis (Yes / No)

IVth nerve Paresis (Yes / No)

VIth nerve Paresis (Yes / No)

Nystagmus (Yes / no specify)

Vth nerve Motor (Yes / No); Sensory (Yes / No)

VII th cranial nerve (Yes / no specify)

VIII th cranial nerve (yes /no specify)

Lower cranial nerve deficit (yes/no specify)

Cerebellar sign

Other

### **CT scan**

Plain (hypo / iso / hyper)

Calcification (Yes / no)

Contrast enhancement (yes/no)

Hydrocephalus(Yes / no)

### **MR Imaging**

Size

Location

Extent

T1 hypo/ hyper/ Iso

T2 hypo/ hyper/ Iso

FLAIR

Diffusion, ADC...

perfusion,

Contrast (Yes / no)

Hydrocephalus

**Surgery (Yes / no specify)**

Surgery approach

Surgery corridor

Plane of cleavage

Extent of removal

intraoperative use of steroid (yes/no specify)

**Post op deficit**

Visual acuity(specify) Visual field (specify)

EOM (full / restriction)

IIIrd nerve Paresis (Yes / No)

IVth nerve Paresis (Yes / No)

VIth nerve Paresis (Yes / No)

Nystagmus (Yes / no specify)

Vth nerve Motor (Yes / No); Sensory (Yes / No)

VII th cranial nerve (Yes / no specify)

VIII th cranial nerve (yes /no specify)

Lower cranial nerve deficit (yes/no specify)

Cerebellar sign (Yes / No)

Aseptic meningitis (Yes / No)

CSF rhinorrhoea/ Otorrhoea (Yes / No)

VP shunt required (yes/no), pre op or post op.

**Post op CT scan (Yes / no)**

Hydrocephalus (Yes / No)

Infarcts (Yes / No)

Residual (Yes / No)

**Post op MRI**

Hematoma (Yes / No)

Infarct (Yes / No)

Hydrocephalus (Yes / No)

Residual (Yes / No)

**HPR (specify)**

**Condition at discharge**

**Outcome**

Outcome at 6 weeks

Outcome at 6 months

Outcome at 1 year

Outcome at 2 year

Outcome at 5 year

Recurrence (Yes / no)

Second surgery for recurrence (Yes / No)

## **ABBREVIATIONS**

ADC-absolute diffusion coefficient  
CA-combined approach  
CISS-constructive interference steady state  
CPA- cerebellopontine angle  
CT – computerized tomography  
CSF – cerebrospinal fluid  
DSA – digital subtraction angiogram  
DWI-diffusion weighted image  
FLAIR-fluid attenuated inversion recovery  
FM-foramen magnum  
GCS – Glasgow coma scale  
GOS - Glasgow outcome scale  
HA – headache  
HCP – hydrocephalus  
LCN - lower cranial nerve  
MCF - middle cranial fossa  
MFA-middle fossa approach  
MRI-magnetic resonance imaging  
MSOC - midline suboccipital craniectomy approach  
PF- posterior fossa  
PD-proton density image

RMSOC-retromastoid suboccipital craniectomy approach

RT- ryle's tube

T1WI- T1 weighted image

T2WI- T2 weighted image