Original Research Article

Microneurosurgical aspects in the management of posterior cranial fossa epidermoids: experience of 24 cases

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Received: 28 December 2019
Accepted: 06 January 2020

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ABSTRACT

Background: There is considerable debate in the surgical management of epidermoid cyst, whether gross total or subtotal resection yields better long term outcomes. We present our institutional experience in evaluating the clinical presentation, diagnosis, and surgical strategy and extent of resection in the management of posterior cranial fossa epidermoid cyst.

Methods: A retrospective review of 24 patients diagnosed with posterior cranial fossa epidermoid tumors surgically treated at the institution between January 2010 and July 2019.

Results: A total of 24 patients who underwent surgery for intracranial epidermoid lesions were identified. Of these 13 were in the Cerebellopontine angle region, eight were in the fourth ventricle, and three in lateral cerebellar convexity. The mean duration from onset of symptoms to surgery was 1.6 years. Cranial nerve dysfunction was noted in 73% of patients preoperatively, most of them being the CPA epidermoids. Total removal was achieved in 16 patients, near-total resection in 6 patients, and subtotal removal in 2 patients. Three patients developed recurrence radiologically of them only one patient became symptomatic. The mean duration of follow-up was 3.8 years. The content of the tumor was pearly white material in all cases. Complications noted in the present series were not related to the completeness of excision. Mortality was noted in one patient.

Conclusions: The present study highlights various precautions to be taken intraoperatively in the prevention of development of aseptic meningitis and concludes that total removal of epidermoids does not result in significantly increased morbidity and mortality and should be the goal of surgical treatment. However, near/subtotal resection of lesions that are densely adherent to neurovascular structures is justified, as there is no significant difference in the rate of recurrence. An endoscope can be used to assess the completeness of surgery.

Keywords: Cerebellopontine region epidermoids, Fourth ventricular epidermoids, Posterior fossa epidermoids

INTRODUCTION

Epidermoid or “pearly” tumors were described by Cruveilhier and designated the “most beautiful tumors of the body” by Dandy.1 Epidermoid cyst (EC) are extra-axial subarachnoid congenital benign slow-growing lesions also known as cholesterolomas or pearly tumors. They usually are rare lesions and represent approximately 0.2-2 % of all intracranial tumors.2 They are slow-growing lesions by the accumulation of keratin and cholesterol, which are the breakdown products of desquamated epithelial cells. Their slow growth and the soft consistency of cholesterolin allow progressive adaptation to the neurovascular structures with preferential development along natural pathways, that is, the subarachnoidal cisterns of the skull base. So, there is no mass effect initially and remain asymptomatic for many years. Symptoms are mainly due to mass effect...
when they increase in size or involvement of cranial nerves (CN). They commonly occur in the cerebellopontine angle (CPA), and parasellar location, which could be explained by the result of the proliferation of multipotential embryogenic cell rests or lateral displacement of ectodermal cells by the developing otic vesicles. Prevalently, EC extends to or involve the CPA and its vital neurovascular structures, thus commonly causing hearing loss, trigeminal neuralgia (TN), vertigo, or facial nerve palsy. As EC is neither sensitive to chemotherapy nor to radiation, surgical resection constitutes the exclusive treatment modality for these lesions. There is continued debate regarding the extent of resection, should we prefer gross total removal (GTR), which can result in unwarranted CN deficit and arterial injury, or should GTR be avoided. A safer, subtotal tumor removal (STR) be chosen. The objectives of the current paper are to report on our operative strategy and surgical technique and to identify the best operative management for posterior fossa (PF) epidermoid and extent of resection.

**METHODS**

The study population constitutes a total of 24 patients who were diagnosed with PF epidermoid cyst. All 24 patients included in the study underwent surgery in Sri Venkateswara Institute of Medical Sciences (SVIMS), between the period of January 2010 to July 2019. Retrospectively collected data includes clinical presentation, radiological findings, surgical approach, the extent of resection (EOR), complications, and neurological outcome were studied. PF epidermoid cyst patients who were managed conservatively were excluded from the study. CT and MRI were used in all cases for diagnosis and follow-up. The location of the tumor determined the surgical approach. The aim of surgery was maximum safe resection with an emphasis on the preservation of CN to give our patients the highest quality of life possible following surgery. EOR was planned preoperatively based on MRI or computed tomography (CT) but ultimately decided intraoperatively when a tumor's adherence to critical structures could be best ascertained. The EOR was determined by examining surgical records and follow up imaging. Postoperatively, all patients received a CT scan in the immediate postoperative period to look for any surgical complications. During follow-up, all patients underwent an MR imaging examination to detect any residual or recurrence. The EOR was considered total if the whole keratinous debris from the core of the tumor, as well as the entire tumor capsule, had been removed. The EOR was considered near-total if the entire keratinous content had been resected, but adhesive parts of the tumor capsule remained in situ. The EOR was considered subtotal if some part of the tumor could not be excised for any reason. The mean follow up period was 3.8 years. No patient dropped out during follow-up. We reviewed the current literature in the context of previous clinical series on PF epidermoid tumors.

**RESULTS**

The total number of patients was 24 (M:F=13:11). Age range, 19-56 years. The mean duration from the onset of symptoms to surgery was 1.6 years. The commonest age group of the presentation was 31-40 yrs, 13 cases EC were located in the CPA region, in 8 cases, EC was located in the fourth ventricle, and in 3 cases, EC was located in lateral cerebellar convexity. Among 13 cases of EC located in CPA, in 3 cases, EC was extending to the opposite side through the preoptic cistern, in 3 other cases, CPA epidermoid was extending to middle cranial fossa. Clinical features were according to the location and extent of the tumor. Headache, CN deficit (73%), cerebellar symptoms were the commonest symptoms in PF epidermoid cyst. Cerebellar signs, including gait ataxia, dysmetria, dysarthria, and nystagmus. Among the CPA epidermoid, TN was noted in six patients. Facial weakness and facial spasms were noted in two patients, hearing loss was noted in two patients. Lower CN palsy was noted in 2 patients.

| Clinical features | Number of patients % |
|-------------------|----------------------|
| Headache (suboccipital region) | 17(70.83%) |
| Cerebellar symptoms | 8(33.33%) |
| Cranial nerve deficit | |
| Trigeminal neuralgia (lancinating pain on one half of the face) | 6 (25%) |
| Hearing impairment and Tinnitus and vertigo | 4 (16.6%) |
| features of seventh cranial nerve palsy | 2(8.3%) |
| Diplopia | 2(8.3%) |
| Nasal regurgitation | 2(8.3%) |
| Hydrocephalus | 6 (25%) |

**Table 1: Clinical features of posterior fossa epidermoid cyst.**

| C P angle epidermoids | Fourth ventricular |
|-----------------------|--------------------|
| Headache | Headache in suboccipital region |
| Involvement of cranial nerve | | |
| • V in the form of trigeminal neuralgia | Cerebellar symptoms |
| • VIII in the form of vertigo, hearing impairment and tinnitus | Swaying while walking |
| • VII in the form of LMN features | Slurring of speech |
| • VI on the form diplopia | Unsteady gait |
| • Lower cranial nerves in the form nasal regurgitation | |
| Cerebellar symptoms | Symptoms of hydrocephalus |

**Table 2: Common clinical features according to location.**
Table 1 depicts clinical features in PF epidermoids, (Table 2) shows clinical features according to location.

CT and MRI brain was done for all cases. In the CT scan, hypodense lesion, which showed minimal contrast uptake in the periphery. In the MRI study, the epidermoid was hypointense in T1 and hyperintense in T2 weighted images. Proton density (PD) differentiated the cyst from the cerebrospinal fluid (CSF). Hydrocephalus with features of raised Intracranial Pressure was in six cases, and pre-excision CSF diversion was done in 3 cases. The neurosurgical approach to EC depends on tumor location and growth pattern. Different micro neurosurgical approaches were used for EC at various sites. The suboccipital retro sigmoid retro mastoid approach was preferred route in all CPA epidermoid.

Midline suboccipital craniectomy (trans vermain) in fourth ventricular EC. In one case, the combined suboccipital and sub temporal approach was used as the tumor was extending from posterior fossa to middle fossa. Paramedian suboccipital craniectomy was done for lateral cerebellar convexity EC. The content of the EC in all cases was pearly white material. In the present study, GTR was possible in 16 cases.

In the remaining cases, it was not possible for complete removal as vital structures needed to be preserved. Near-total resection was done in six patients. A minute amount of tumor was left behind the basilar artery in two cases and two cases tumor capsule adherent to brain stem was left behind. One case was a recurrent case operated elsewhere, so some amount of tumor adherent to the brain stem and lower cranial nerves was left behind. Two cases underwent STR was done. In four cases, an Endoscope was used to check the completeness of resection. All patients underwent an early postoperative CT brain in the first 24 hrs to check for any complications. Table 3 shows the post complication. The neurological deficits are newly developed CN deficits post operatively.

Table 4 shows the fate of preoperatively present CN deficits in relation to recovery (early or late) or worsening or persistence. Out of six patients with TN who underwent surgery four patients had complete relief of symptoms postoperatively remaining two patients had relief of symptoms very slowly.

Table 3: Complications noted in this study in relation to extent of resection.

| Complications                        | GTR | STR and NEAR |
|--------------------------------------|-----|--------------|
| Aseptic/chemical meningitis          | -   | 1            |
| Transient cranial nerve palsy        |     |              |
| • Third CN palsy                     | 1   | -            |
| • Sixth CN palsy                     | 1   | 2            |
| • Seventh CN palsy                   | -   | 1            |
| • Lower CN palsy                     | 2   | 1            |
| Permanent cranial nerve palsy        |     |              |
| • Lower CN palsy                     | 1   | 1            |
| Post meningitic hydrocephalus        | 3   | 2            |
| Hemiparesis                          | 1   | -            |
| CSF rhinorrhea                       | 2   | -            |

(GTR- gross total resection, STR- subtotal resection, CN - cranial nerve)

Table 4: Cranial nerve outcome following surgery.

|                  | Preoperatively | Postoperative period | No recovery | Worsening |
|------------------|----------------|----------------------|------------|----------|
|                  | Recovered      |                      |            |          |
|                  | Early | Late |              |            |          |
| Trigeminal neuralgia | Six   | 4 | 2 |            |          |
| Hearing deficit   | Two  | No recovery | 2 | No worsening |
| Facial nerve palsy | Two  | No recovery | 2 |          |
| Lower cranial nerve palsy | One | Worsened in one patient |            |          |
| Diplopia          | One  | Late recovery | |          |

Table 5: Comparison of the present study with various other studies in literature.

| Name of the study | No. of cases | GTR (%) | Mortality (%) | Recurrence (%) | Follow up (Years) |
|-------------------|--------------|---------|---------------|----------------|------------------|
| Kobata et al⁷     | 30           | 56.7%   | 0             | 6.6%           | 11.4             |
| Akar et al¹¹      | 28           | 75%     | 3.5%          | 7%             | 6                |
| Samii et al¹²     | 40           | 75%     | 2.5%          | 7.5%           | 5.7              |
| Schiefer et al¹⁴  | 24           | 54%     | 0             | 25%            | 4.3              |
| Yasargil et al¹⁰  | 22           | 95%     | 0             | 9%             | 5.2              |
| Choudary et al¹⁵  | 23           | 73.9%   | 4.3%          | NA             | 3                |
| Kato et al¹⁶      | 27           | 10%     | 5%            | 20%            | NA               |
| Present study     | 24           | 66.6%   | 4.1%          | 4.1%           | 3.8              |
were
Salient
subsequently
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(54%).
Regarding
the
opening
dura and placing spatula to retract
cerebellum and visualizing the lesion. (E) pearly white
lesion seen along with fifth and seventh cranial nerves.

Figure 1: Imaging and intra-operative pictures cp
angle epidermoid cyst (A) MRI showing CPA
epidermoid extending anterior to the basilar artery on
to the opposite side (B) DWI showing restriction (C)
operative picture of the patient with lesion and
marking of incision for undergoing retrosigmoid
retromastoid suboccipital craniectomy. (D) After the
opening of the dura and placing spatula to retract
cerebellum and visualizing the lesion. (E) pearly white
lesion seen along with fifth and seventh cranial nerves.

But patients with hearing loss did not regain hearing even
after complete excision. Two patients required
postoperative external ventricular drainage for
postoperative hydrocephalus. One patient needed
ventriculoperitoneal shunting as a consequence post,
en meningitis hydrocephalus. (patient developed meningitis
post-EC removal) Regarding post-operative
complications, following fourth ventricular epidermoid
tumor were CSF leak, complications following CPA
Epidermoid were related to facial nerve palsy which
resolved in due course, transient CSF rhinorrhea (in one
CPA epidermoid), and sixth nerve palsy and chemical
meningitis in one patient. And one patient had lower CN
palsy, which was managed by RT feeding and
subsequently recovered. The complication was minimal
after the removal of lateral cerebellar convexity EC. One
CPA epidermoid patient died due to meningitis.

Table 5 shows results in the present study along with
results from various studies from literature, the results of
present study are more or less same.

Salient operative steps common to all procedures in study
were;

- positioning and incision depend on location
- dural opening
- placement of cotton pads surrounding the tumor to
prevent spillage of cystic contents into subarachnoid
space thereby preventing chemical meningitis

- careful inspection of anatomy and gradual,
piecemeal reduction of the tumor thereby creating
working space between tumor capsule and
surrounding neurovascular structures
- sharp dissection of capsule done to and free it from
adjacent neurovascular structures
- if the capsule is tightly adherent and removal of the
capsule is likely to injure vital structures, capsule
may be left behind.
- The leftover capsule should never be coagulated.
- Irrigation of surgical site continuously with
hydrocortisone (100mg/l in lactated ringer solution)
also reduces the chances of post-operative
communicating hydrocephalus and aseptic
meningitis.
- Intraoperative steroid administration
(dexamethasone)
- Careful attention was paid to identifying and
preserving the arachnoid plane at the tumor–
brainstem interface, which facilitated complete
tumor resection and minimized small vessel and
brainstem injury.
- All open mastoid cells were sealed with bone wax to
prevent CSF leaks.
- Usage of the endoscope allows inspection of
obscured but important corners of the operating
field without the need to retract neurovascular
structures or to resect dura and bone edges.
Compared with a microscopic surgical field alone,
the endoscope provides a view of the operative field
from a different angle.

Figure 1 shows the preoperative imaging of a patient
from the study along with operative position and
intraoperative picture after opening dura and appearance
of epidermoid cyst.

DISCUSSION

Many studies suggest EC has no male or female
predilection, as in our study, females (11) and males (13).
EC patients typically are between 20 and 40 years of
age. Present study most of the patients were in the fourth
decade correlating with previous studies. The commonest
age group in our study was 31-40 years. The range of
patient age was 19-56 yrs. The most common location of
PF epidermoids in the present study being CPA, 13 of 24
(54%). Literature suggests 60% of all intracranial
epidermoids occur in CPA. The commonest tumors in the
CPA region are acoustic neuromas and meningiomas, EC
being the third commonest in that region. Literature
suggests that the second most prevalent location in the PF
is the fourth ventricle accounting for 5-18% of all
intracranial epidermoids, 8 of 24 in the present study
were noted in the fourth ventricle. Three were noted in
lateral cerebellar convexity. Spinal epidermoid is very
rare. The mean duration of symptoms in the present
study was 1.6 years. EC has a thin capsule composed of
keratinized stratified squamous epithelium, which
desquamates and gives rise to the cyst contents, keratin, and cholesterol. They tend to grow very slowly along natural cleavage planes, sulci, fissures, cisternal spaces, ventricles until all available subarachnoid space is used up. Symptoms become clinically apparent, because of this, the symptoms are noticed in the delayed stage. Symptoms and signs tend to appear at the later stage due to the physical presence of tumor-causing irritative effects. Infratentorial epidermoids cause CN deficits and cerebellar symptoms. The most common presenting symptoms in patients with CPA epidermoids are TN and hearing impairment, followed by dizziness, headaches, diplopia, and facial nerve paresis.5

Table 6: Differential diagnosis of cystic lesions of brain.

| Lesion                                           | Differentiating radiological characteristics                                                                 |
|--------------------------------------------------|-------------------------------------------------------------------------------------------------------------|
| Classical epidermoid                            | • Epidermoids classically follows a CSF intensity pattern on CT and MRI.                                    |
|                                                  | • Hypodense on noncontrast CT, as they are avascular, no enhancement on contrast CT                           |
|                                                  | • Hypointense on T1WI-MRI, hyperintense on T2WI-MRI, suppress on FLAIR (used to differentiate them from the arachnoid cysts which do not suppress on FLAIR) and show diffusion restriction. |
|                                                  | • These tumors are usually solid and have no triglycerides. These classical epidermoids are referred to as black epidermoids. |
| Atypical epidermoid (white epidermoid)17         | • Hypodense on CT, Restriction absent on DWI 18                                                            |
|                                                  | • Hyperintense on MRI T1WI1, Hypointense on MRI T2WI                                                       |
| Malignant transformation of benign epidermoid cyst19 | • Histological diagnosis is confirmatory                                                                  |
|                                                  | • Presence of edema and tissue invasion, rapid growth, and new enhancement on contrast are some features of malignant transformation of epidermoid cyst |
|                                                  | • Immunohistochemistry of the malignant epidermoid cyst reported in the literature showed the positivity of tumor cells with P53 protein. |
| Dermoid cyst                                     | • Midline location, grow fast and rapid, leading to the occurrence of clinical features                     |
|                                                  | • It contains dermal appendages like hair follicles, teeth, lipids.                                        |
|                                                  | • The dermoid cyst appears in 2nd to 3rd decade of life.                                                  |
|                                                  | • Hypodense on CT, Hyperintense on T1WI and iso-hyperintense on T2WI                                      |
|                                                  | • The lining of a dermoid cyst is a simple stratified squamous epithelium supported by collagen.            |
| Arachnoid cyst                                   | • Located mostly at middle cranial fossa                                                                   |
|                                                  | • Hypointense on both FLAIR and DWI                                                                        |
| Neurenteric cyst                                 | • Ovoid, extra-axial, well-delineated lesion located at the midline in the posterior fossa.                 |
|                                                  | • It may be spontaneously hyperdense on CT.                                                                  |
|                                                  | • MRI, the cyst usually appears hyperintense to CSF signal on T1, T2, and FLAIR sequences, owing to protein-rich content. No enhancement or restricted diffusion is typically seen. |
| Colloid cyst                                      | • Mass arising at the foramen of Monro frequently manifests as obstruction of the lateral ventricles.     |
|                                                  | • The well-defined round cyst may be from several millimeters to 3 cm in size and attaches to the superior anterior aspect of the third ventricle roof |
|                                                  | • Hyperattenuating at nonenhanced CT, it has variable signal intensity at MR imaging and is often hyperintense on T1-weighted and FLAIR images. |

In the present study, six patients had TN four of these patients relieved in the immediate period after surgery. TN and hemifacial spasm may result from nerve compression by an artery displaced by the EC or from nerve displacement towards the artery by the tumor. Kobata et al, highlighted that in patients with EC, hemifacial spasm occurs only in cases of neurovascular conflict.7

At the same time, TN may often also result from displacement or kinking of the trigeminal nerve related to the presence of a tumor, and in cases unrelated to a tumor may also be caused by arachnoid adhesions or venous pathology. They believe that this difference is related to the length of the Redlich-Obersteiner zone, which is sensitive to direct compression or displacement and is nearly three times longer for the trigeminal nerve compared to the facial nerve (2.2 mm vs. 0.8 mm). As a result, TN due to trigeminal nerve displacement is more likely than hemifacial spasm due to facial nerve displacement. In my present study, two patients had hemifacial spasm compared to six with TN.
Rarely EC may present with a recurring history of chronic aseptic meningitis with prolonged episodes, which can occur due to spontaneous rupture of cyst contents into subarachnoid space, which is highly irritative. No patient in the present study presented with recurrent aseptic meningitis the clinical features in the present study matched to those in literature. The appearance of EC on MRI shows low signal intensity on T1WI and high signal intensity on T2WI. Conventional MR images sometimes cannot reliably be used to distinguish EC from arachnoid cysts since both lesions are very hypointense relative to brain parenchyma on T1-weighted MR images and very hyperintense on T2-weighted images. Fluid-attenuated inversion recovery (FLAIR) and diffusion-weighted (DW) sequences can successfully be used for the diagnosis of EC by revealing its solid nature. DW imaging is superior to other MR sequences in delineating the borders of the epidermoid cyst. FLAIR MR imaging is based on the nulling of the signal from CSF. EC characteristically present on this sequence as heterogeneous lesions with central parts of the tumor being hyperintense relative to the hypointense CSF,T2WI is hyperintense and has a high value on ADC. Capsular enhancement of the cyst is not often seen; the high contrast enhancement of epidermoid capsule is rare. All the patients in the present study needed MRI DWI images for confirmation of diagnosis. The primary differential diagnosis for an EC is arachnoidal cysts, dermoid cysts, cystic neoplasms, neurocysticercosis, neurenteric cyst, etc. (Table 6) differentiates EC from all these lesions.

White EC is atypical in that they, due to their high protein content and consequently high viscosity, they are hyperintense on T1WI sequence, hypo- to hyper-intense (commonly hypointense) on T2 and show no diffusion restriction. They are usually cystic, and in addition to their high protein content, they typically have high lipid content with mixed triglycerides containing polyunsaturated fatty acids and no cholesterol, further contributing to their high viscosity. There is no uniform opinion regarding the extent of surgery in EC in literature. Some advocate GTR others advocate STR and leaving behind tumor adherent to vital nervous vascular structures. Berger and Wilson did not perform in any of their cases, reporting minimal patient morbidity and no recurrences over 4.5 years of follow-up. On the other hand, Yasargil et al, achieved total removal in 95% of patients with minimum morbidity and a 9% recurrence rate of over 5.2 years of follow-up. All other reports fall somewhere in between these ends of the spectrum. In the present study, 16(66.6%) patients underwent GTR to 2(8.3%) patients of STR.

Although the universal goal of surgery is the complete removal of the lesion without damage to adjacent neurovascular structures, portions of the capsule adherent to these structures often make this extremely difficult. Akar et al, found that the presence of CN dysfunction before surgery increased the incidence of dysfunction postoperatively, which was also consistent with experience. Chemical meningitis can occur by spillage of the cyst contents during operation, which usually is transient and self-limiting and can be managed successfully with steroids. Excision of the capsule by sharp dissection, irrigation of the CPA cisterns with hydrocortisone solution during the surgery, and delayed withdrawal of steroids in the post-operative period have been advocated as possible measures for preventing chemical meningitis. There was one death in the present study that occurred in CPA epidermoid, and it was due to meningitis. Due to the risk of recurrence, GTR should be the goal of the surgery, but in other reports was found no difference in the risk of recurrence between completely and incompletely removed tumors.

**Role of endoscope**

Compared with a microscopic surgical field alone, the endoscope provides a view of the operative field from a different angle. This feature improves the assessment of the extent of the surgical excision. In 1998 Fries and Perneckzy evaluated their experience with endoscope-assisted microsurgery in the treatment of 205 brain tumors. The authors did not report whether their patient population included EC or other CPA tumors. However, they concluded that endoscope-assistance might reduce the trauma related to resection and improve operative outcomes. In present data, although endoscope was used only in four cases (CPA epidermoids), author used to see the completeness of resection and extent to the opposite side. No complications were attributed to the direct application of the endoscope in the present study.

**CONCLUSION**

EC are rare slow-growing tumors that primarily increase in size by desquamation of epithelial cells rather than by multiplication. Multiple CN deficits are more noted in CPA epidermoids. EC is soft friable, avascular, and easy to resect. Most of the time, the brain is lax and easy to retract. GTR is the treatment of choice. Meticulous care to be taken to excise an EC always the goal should be to completely resect at the same time to preserve the neurological function of the patient by microsurgical operative steps. For achieving a GTR, the most important is the complete resection of capsule. While teasing off the capsule of epidermoids away from the nerve or vessel, the neurovascular structure may be injured and resulting in postoperative morbidity. Sometimes it is better to leave off some tiny portions of tumors attached to the neurovascular structures. Therefore, while removing the capsule one should be careful. In many CPA epidermoids, hearing loss and diplopia persisted after surgery, whereas marked improvement was observed in the TN. Finally, the time taken by recurrent EC to produce symptoms is a very long period, so it is better to follow up on a small recurrent case of EC than to produce...
a neurological deficit in the quest for GTR. This is true in CPA epidermoids. Prevention of aseptic meningitis can be done by covering all the exposed areas of the brain with cotton pledgets, intraoperative steroid cover, and continuous irrigation of field with steroid with normal saline. The endoscope can be used to assess the completeness of resection, allows inspection of obscured but important corners of the operating field without the need to retract neurovascular structures or to resect dura and bone edges. No attempt should be made to remove either the lesion or its capsule with the endoscope alone.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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Cite this article as: Chandra VVR, Prasad BCM, Niranjan V, Rajesh P. Microneurosurgical aspects in the management of posterior cranial fossa epidermoids: experience of 24 cases. Int J Res Med Sci 2020;8:606-12.