Surgical Management of Intracranial Epidermoid Tumors—An Institutional Review of 234 Cases

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Abstract

Aim To study the demographics, clinical profile, management strategies, and treatment outcomes of intracranial epidermoid lesions.

Materials and Methods A single institutional retrospective analysis from a large institute in Western India was done of all patients operated for intracranial epidermoid tumors over a period of 20 years from January 1997 to January 2017. All patients were investigated with a contrast-enhanced computed tomography (CECT) as well as a magnetic resonance imaging (MRI), with a diffusion-weighted imaging to confirm diagnosis of epidermoid tumor. The presenting complaints and the demographics of all patients were recorded, and all patients underwent planned surgery, with or without a cerebrospinal fluid (CSF) diversion procedure as per requirement. Surgery ranged from a gross total resection to a near-total resection depending on the patient, location of the lesion, and involvement of adjacent structures. All patients underwent an immediate postoperative computed tomographic (CT) scan as well as a follow-up MRI. Patients were followed up to note clinical improvement as well as documentation of all complications.

Results Total 234 patients of intracranial epidermoid tumors were operated upon in a 20-year span. Males predominated with a total number of 146 (62.4%). The most common age group was the fourth decade. The most common presentations were headache (118 patients) and neuralgia in the trigeminal distribution (82 patients). Infratentorial compartment was the most common location of the tumors (152 patients), with cerebellopontine angle being the predominant site in the posterior fossa (107 patients). Both supra- and infratentorial compartments were involved in 28 cases. Gross total excision could be achieved in 202 patients. Reoperation was required in 12 cases. The mean follow-up period was 68 months. The most common complication encountered was that of postoperative CSF leak (24 patients). We had eight cases of recurrence and six deaths in our series.

Conclusion Epidermoid tumors can arise in virtually all intracranial locations. Gross total excision of the tumor is treatment of choice and can be curative for these benign lesions. Large lesions at presentation with involvement of multiple compartments can be difficult to manage and may require bicompartmental or second look surgery. Complete excision of the capsule with preservation of adjacent neuronal and vascular structures is of utmost importance to decrease postoperative morbidity and chances of recurrence. Perioperative steroids, avoidance of intraoperative spillage, and intraoperative endoscopic assistance are recommended in all cases.

Keywords
- ► epidermoid tumor
- ► infratentorial
- ► supratentorial
- ► cerebellopontine angle
- ► surgical outcomes

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Introduction

Intracranial epidermoid tumors account for roughly 1% of all intracranial tumors.[1] They are considered to be anomalies of gastrulation phase with entrapment of ectodermal cell rests in the developing neuroectoderm.[2] Characteristically these lesions spread from the basal surface of the brain to a variety of locations with most common being the cerebellopontine angle (CPA), infratentorial middle cranial fossa, and suprasellar regions.[3] These benign, slow-growing lesions can produce a myriad of symptoms depending on the location of the lesion; for example, trigeminal neuralgia is common with CPA epidermoid tumors and seizures are common with interhemispheric lesions. Presentation in these tumors is most often attributed to neural irritation rather than actual mass effect. Magnetic resonance imaging (MRI) is the imaging modality of choice for these lesions, with diffusion-weighted imaging (DWI), which helps differentiate these from their most common differential diagnosis, that is, arachnoid cysts.[4] The treatment of choice for these lesions is radical surgical resection. Although these tumors cross cisternal boundaries, majority of them can be resected with low morbidity, mortality, and recurrence with meticulous microsurgical techniques avoiding dreaded compilations of aseptic meningitis, neural deficits because of manipulation, and vascular injury.[3]

Materials and Methods

A retrospective analysis of 234 patients operated upon for intracranial epidermoid tumors in a 20-year period from January 1997 to January 2017 was conducted in a tertiary care center in Western India. Preoperatively all patients underwent a contrast-enhanced computed tomography (CECT) as well as an MRI with diffusion-weighted sequences to confirm the diagnosis. The complete demographic record was noted of all patients, and the clinical profile at presentation was analyzed. A subset of patients presenting with acute hydrocephalus underwent a cerebrospinal fluid (CSF) diversion procedure before the planned excision. All patients were operated upon with the aim of radical excision of the epidermoid tumor using microsurgical techniques with care to avoid adjacent vascular and neural damage. Surgical approaches were tailored to suit the site of the lesions with retrosigmoid approach used for CPA lesions most commonly whereas a combined retrosigmoid plus infratemporal approach was used for large bicompartmental (supratentorial + infratentorial) lesions. The other approaches used were the interhemispheric for midline frontal tumors, pterional for sellar and parasellar regions, subtemporal for middle fossa tumors, suboccipital craniotomy for midline posterior fossa and fourth ventricular tumors, and transcortical approach in other supratentorial tumors (►**Table 1**). The lateral semisitting position was used for all cases operated via a retromastoid or a bicompartmental approach (►**Fig. 1**). Meticulous separation of the adherent tumor capsule was given special importance. Intraoperatively, angled endoscopes were used to view the surgical field and localize microscopically hidden recesses to achieve complete resection. Endoscopes were used in our series since 2003, that is, past 14 years of the study. It was used in all cases of posterior fossa and bicompartmental, where the areas hidden by the tentorial incisura were seen much better. There were invariably some residual bits of tumor, which were out of the reach of the microscopic view. In supratentorial epidermoids, endoscopes were used in all cases of intraventricular epidermoids to check the areas that were not visible from the corticectomy. Patients in whom radical resection could not be achieved were closely followed up with a subset undergoing a second look procedure either by the same of a different approach. Intraoperative steroid (hydrocortisone), both parenteral and in the irrigation fluid, was administered to reduce the chances of aseptic meningitis. Immediate postoperative CT scans with MRI on follow-up of 1 month was done in all patients to document any residual lesions. The mean follow-up period was 68 months (minimum 18 months to maximum 92 months). Follow-up MRI was done at a mean interval of 36 months to document recurrence. All demographic, clinical, and perioperative data were retrospectively analyzed and tabulated.

Table 1 Surgical approaches

Approach	Most common tumor location
Retrosigmoid suboccipital	Cerebellopontine angle tumors
Anterior interhemispheric	Interhemispheric
Far lateral approach	Infratentorial cisternal
Pterional subtemporal	Middle cranial fossa tumors
Subfrontal approach/ trans-sylvian	Suprasellar/parasellar
Supracerebellar infratentorial	Quadrigeminal cistern/pineal region
Transcortical	Intraventricular
Combined retrosigmoid + subtemporal	Bicompartmental tumors

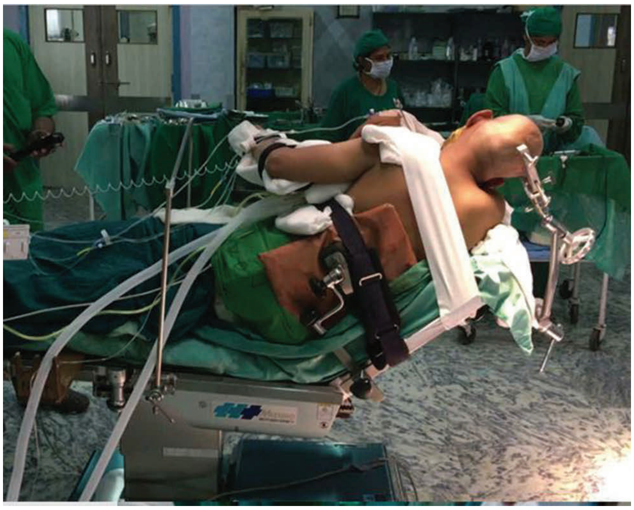

Fig. 1 Lateral semisitting position used for most patients. This position facilitates both retro- and presigmoid access with minimal complications encountered in.

Results

Total 234 patients were operated upon out of whom 162 were males and 72 were females. The most common age group at presentation was the fourth decade (►**Table 2**). Infratentorial location was more common (152) than supratentorial (54), whereas 28 patients had a bicompartmental involvement. Most common presenting symptom was that of headache (118) followed by neuralgia of the trigeminal region (82) (►**Table 3**). The most common anatomical location of the lesion was at the CPA (107) (►**Table 4**). Although hydrocephalus was seen in 44 patients, the most common locations of lesions in the patients were the quadrigeminal cistern and intraventricular, rather than CPA. Hormonal imbalance was seen in six tumors, five of which were suprasellar and one was intraventricular in location. A gross total excision as documented by a follow-up MRI at 1 month was documented in 202 patients. The other 32 patients were labeled as subtotal resection. CSF diversion (ventriculoperitoneal shunt) was required in 17 patients preoperatively, one out of which was lateral ventricular epidermoid, whereas the rest were either completely or partly infratentorial. The most common complication was that of a CSF leak. There were eight documented recurrences out of which five underwent second look surgery and three opted out of surgery (►**Table 5**). There was only one recurrence in the gross total resection category whereas the other seven underwent subtotal resection. Immediate postoperative morbidity defined as prolonged intensive care stay more than 10 days was seen in 36 patients, 31 (86%) of whom belonged to the gross total resection category. Prepontine and bicompartmental lesions had the highest immediate postoperative morbidity with 10 out of the 11 patients, and 13 out of 28 patients requiring prolonged intensive care unit (ICU) stay, respectively. There were six deaths in our series. Three patients died of pulmonary infections, one from pulmonary embolism and two died from central nervous system infection.

Table 2 Age distribution

Age (y)	Number of patients	Percentage (%)
≤ 10	18	7.6
10–20	42	17.9
20–30	59	25.2
30–40	82	35
40–50	26	11.1
≥ 50	7	2.9

Table 3 Clinical features

Clinical features	Number of patients	Percentage (%)
Headache	118	50.4
Trigeminal neuralgia	82	35.04
Hemifacial spasm	9	3.8
Cranial nerve deficits	41	17.5
Visual disturbances	13	5.5
Parinaud's syndrome	8	3.4
Hydrocephalus	44	18.8
Seizures	18	7.6
Hormonal imbalance	6	2.5
Frontal lobe signs	12	5.1
Motor deficits	6	2.5
Altered sensorium	31	13.2
Cerebellar Signs	2	0.8

Representative Cases

Case 1: A case of right caudate nucleus region intraparenchymal epidermoid (►**Figs. 2**, **3**)
Case 2: A case of large left CPA epidermoid (►**Figs. 4**, **5**)
Case 3: A case of quadrigeminal cistern/pineal region epidermoid (►**Figs. 6**, **7**)
Case 4: Another case of quadrigeminal cistern/pineal region epidermoid (►**Figs. 8**, **9**)

Table 4 Location of tumor

Location	Number of patients	Percentage (%)
Cerebellopontine angle	107	45.7
Prepontine	11	4.7
Quadrigeminal cistern (pineal region)	8	3.4
Other infratentorial	26	11.1
Suprasellar/parasellar	14	5.9
Infratemporal middle fossa	6	2.5
Interhemispheric	18	7.6
Frontal lobar	6	5.9
Other supratentorial	10	4.2
Supratentorial + infratentorial	28	11.9

Table 5 Complications

Complications	Number of patients	Percentage (%)
CSF leaks/pseudomeningocele	24	10.2
Infections	8	3.4
Aseptic meningitis/ventriculitis	15	6.4
Pneumonia	13	5.5
Pulmonary embolism	1	0.4
New-onset cranial nerve palsies	14	5.9
New onset motor deficits	5	2.1
New-onset visual disturbance	1	0.4
Prolonged ICU stay (> 10 d)	36	15
Recurrence	8	3.4
Death	6	2.5

Abbreviations: CSF, cerebrospinal fluid; ICU, intensive care unit.

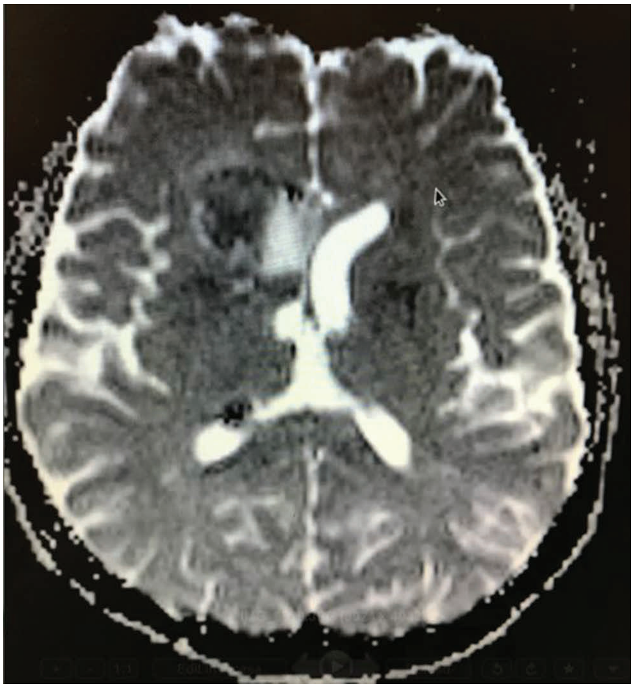

Fig. 2 Axial MRI showing a caudate region epidermoid cyst.

Fig. 3 Postoperative MRI.

Case 5: A case of intraventricular epidermoid operated by transcortical approach (►**Figs. 10**, **11**)
Case 6: A case of in interhemispheric epidermoid operated with a bifrontal craniotomy (►**Figs. 12**, **13**)
Case 7: A case of parafalcine epidermoid (►**Figs. 14**, **15**)
Case 8: A case of lateral intraventricular epidermoid (►**Figs. 16**, **17**)

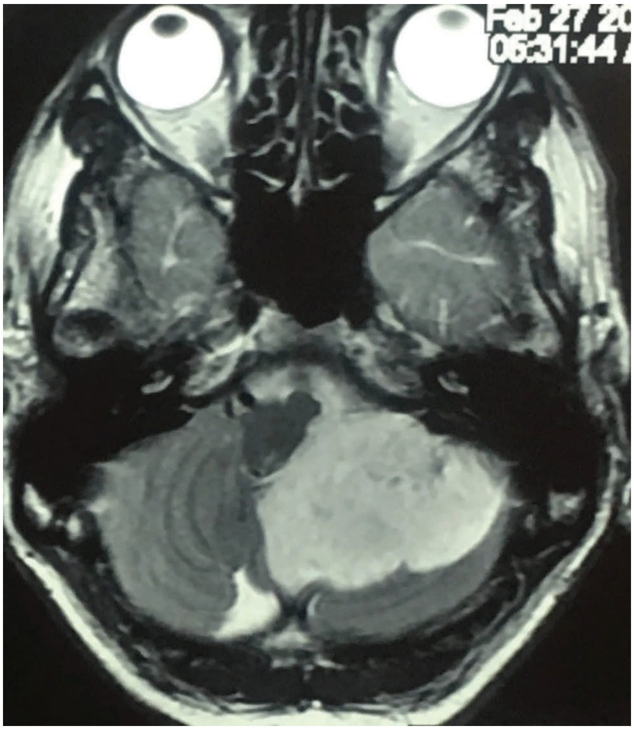

Fig. 4 MRI of left cerebellopontine angle epidermoid tumor.

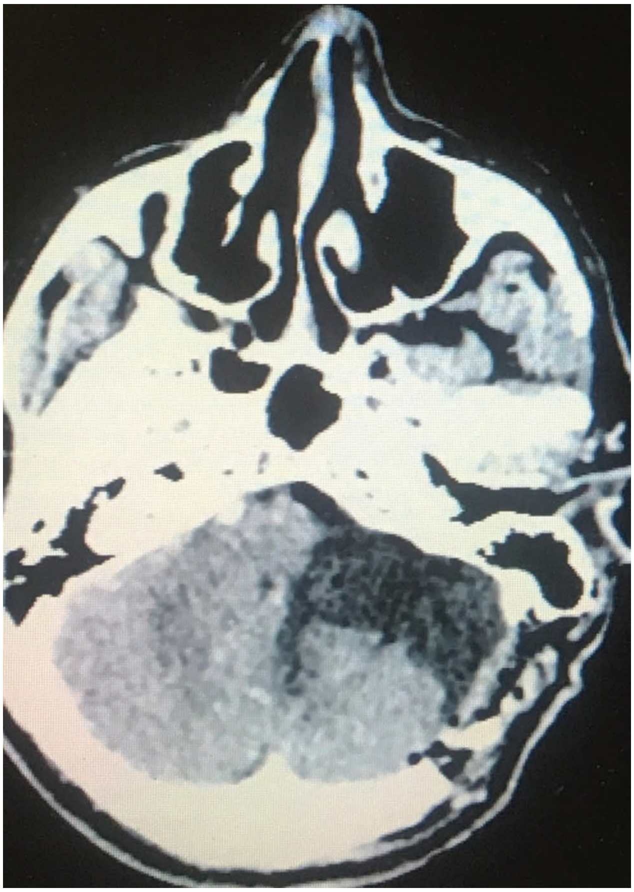

Fig. 5 Immediate postoperative CT scan.

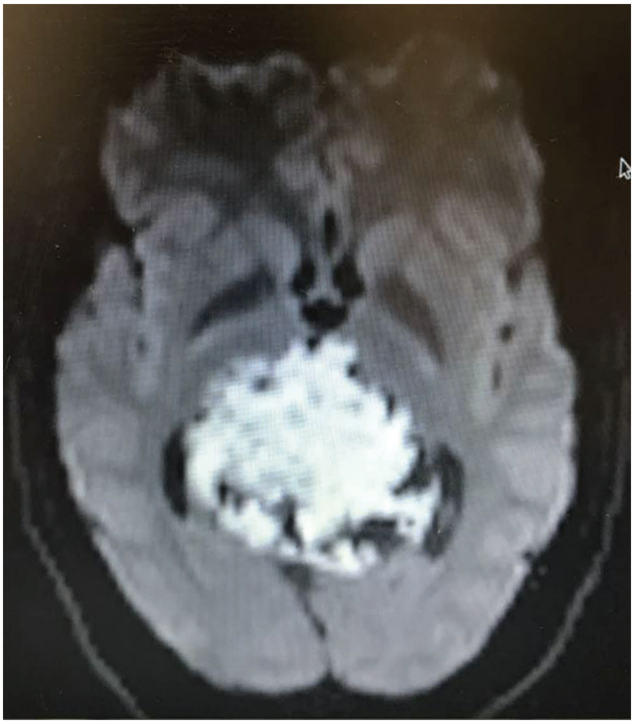

Fig. 6 Case of large quadrigeminal cistern/pineal region epidermoid cyst.

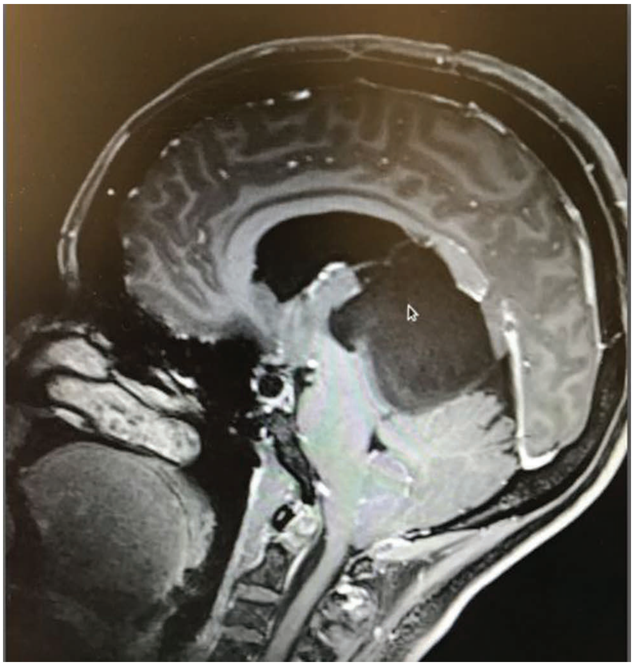

Fig. 8 Another case of quadrigeminal cistern/pineal region epidermoid operated by supracerebellar infratentorial approach.

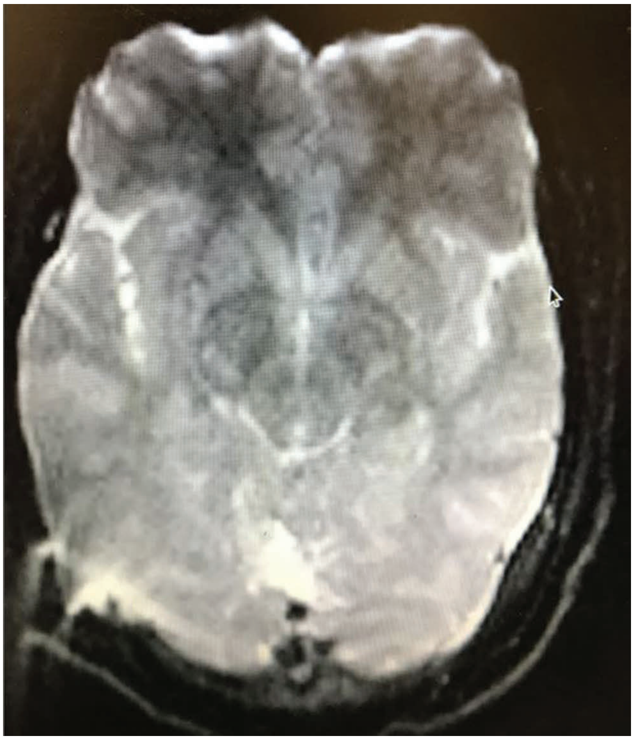

Fig. 7 Postoperative MRI showing complete excision.

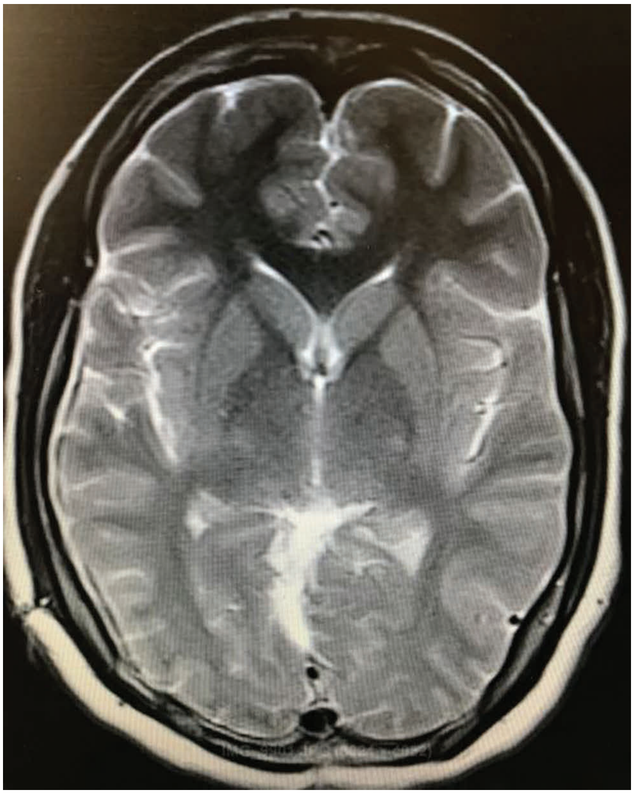

Fig. 9 Postoperative MRI showing complete excision of the lesion.

Discussion

Intracranial epidermoid tumors arise from aberrant closure of the dorsal neural tube.[5–7] As compared with the medially predominant dermoid tumors, the lateral preference of epidermoid tumors may be due to the proliferation of multipotential embryonic cell rests or the abnormal transplantation of epithelial cell rests carried laterally in the migrating otic vesicles or developing neuro-vasculature.[8] Rarely, they have been documented to be iatrogenically induced by repeated percutaneous subdural tapping.[9] Epidermoid tumors grow by accumulating keratin and

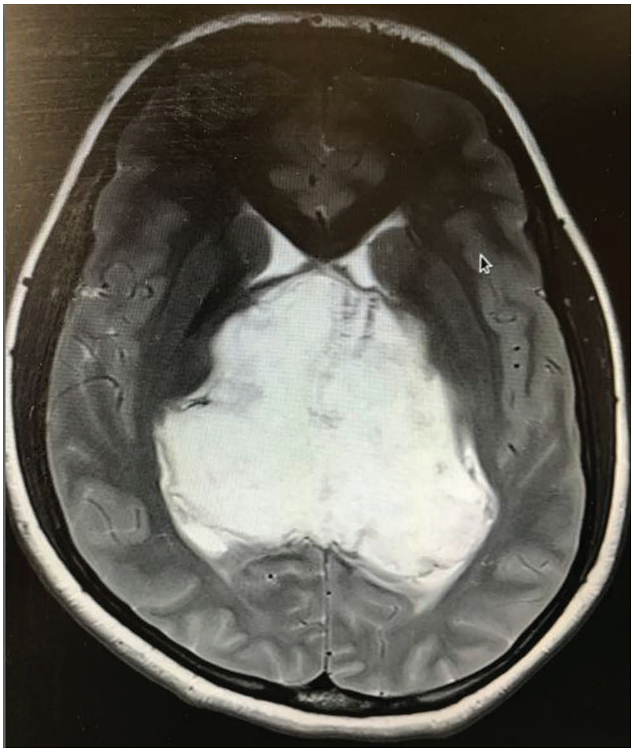

Fig. 10 MRI of a large intraventricular epidermoid cyst.

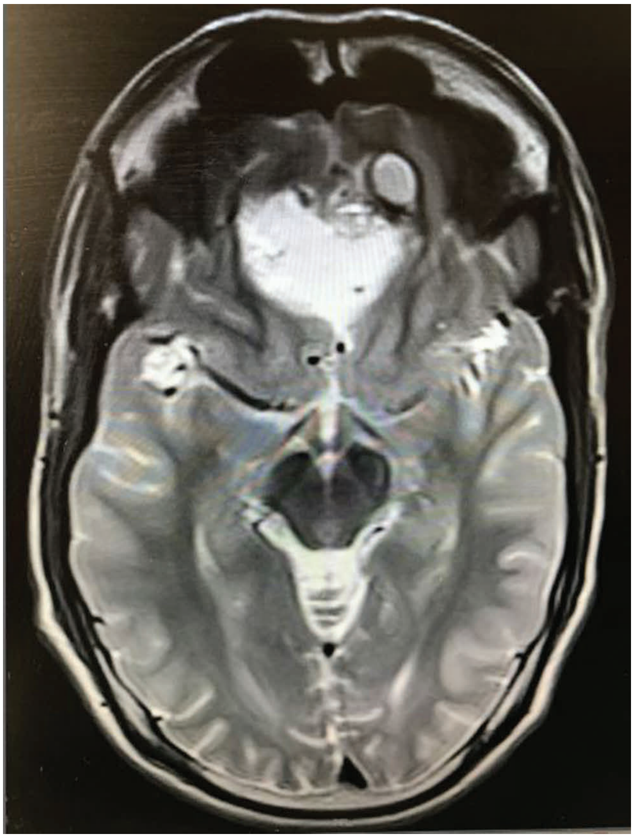

Fig. 12 Axial MRI showing an interhemispheric epidermoid cyst.

Fig. 11 Postoperative image showing conservative tumor decompression.

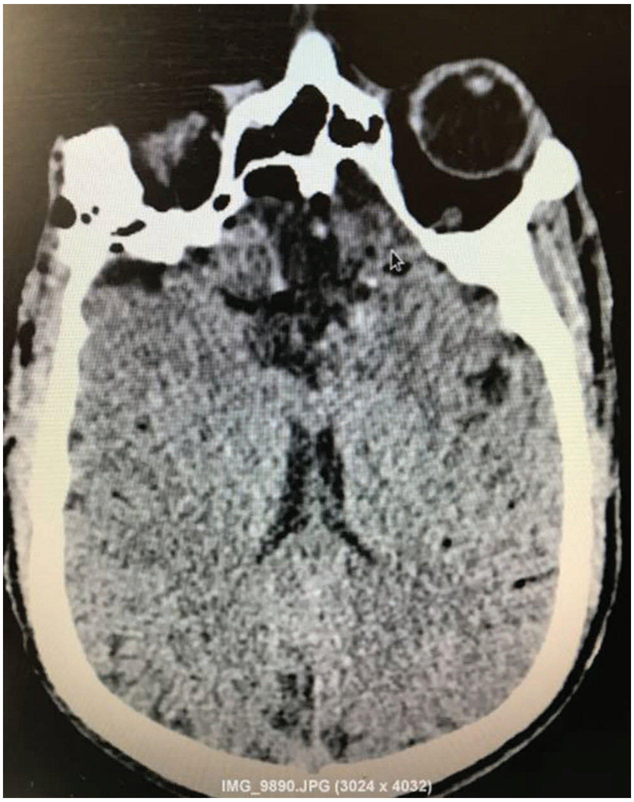

Fig. 13 Postoperative CT scan after resection of the lesion.

Fig. 14 MRI showing a right parafalcine region epidermoid cyst.

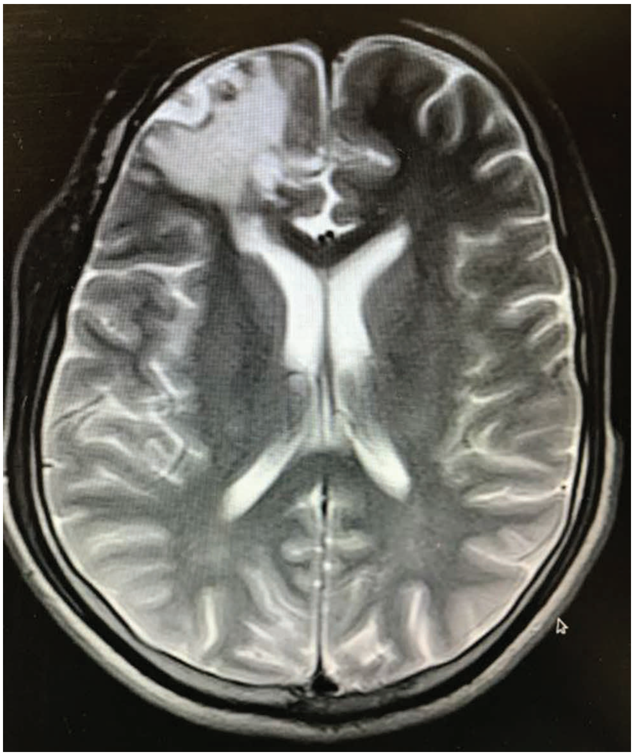

Fig. 15 Postoperative MRI.

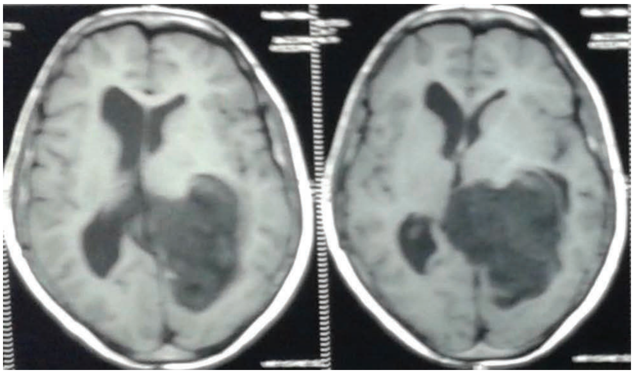

Fig. 16 Axial CT scan images showing large left lateral ventricular epidermoid cyst.

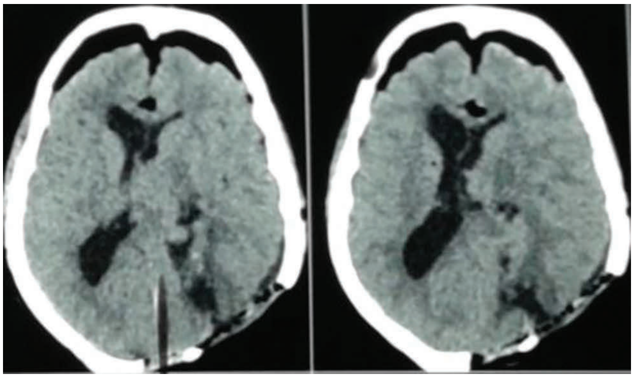

Fig. 17 Postoperative CT scan images.

cholesterol. These arise from the breakdown products of desquamated epithelial cells. These products also give these tumors their soft and pliable consistency and their classic growth patterns.[5] As these tumors grow, their increased volume is compensated by their "flow" pattern through the paths of least resistance, that is, the subarachnoid spaces of the sulci, fissures, cisterns, and ventricles. These tumors conform to the shapes of the cavities which they enter and do not displace the adjacent vascular and neural structures until all available subarachnoid space is occupied. This unique characteristic leads to delayed onset of symptoms.[3] Due to this slow growth, the most common age group of presentation is between 20 and 40 years.[10] This is consistent with our study where the most common age group was the fourth decade (35%). The most common location of intracranial epidermoid tumors is the CPA (40–50%), and they represent 5 to 10% off all CPA lesions. This has also been our finding with 107 (45.7%) of the 234 patients having CPA tumors.

The delayed symptoms are generally attributed to the irritative effects of these tumors more so, than due to the mass effect.[11] The infratentorial lesions most often present with cranial nerve involvements and cerebellar dysfunction.[12,13] The local irritation most often gives rise to trigeminal neuralgia and hemifacial spasm before any other neural deficits.[14] The supratentorial tumors often present with cranial nerve deficits, seizures, and hypothalamic impairment due to their proximity to the parasellar region and the temporal lobes.[11,15] Our series has similar findings with a common symptom in the infratentorial epidermoid tumors being trigeminal distribution neuralgia (35.4%). An infrequent but well-documented mode of presentation is that of recurrent aseptic meningitis and its sequelae.[16,17] Although no patient in our series had this presentation, 15 patients developed culture-negative meningitis after surgery.

Epidermoid tumors have characteristic radiologic features that nearly always confirm the diagnosis before surgery. These lesions typically appear as low-density lesions on CT imaging.[18] MRI is highly sensitive for diagnosis and helps fully determine the extent of these lesions particularly in the posterior fossa and in the saggital plane. The MRI findings, which have been our observations as well, are that of uniform T1 and T2 relaxation times in these tumors, with infiltration of cisternal spaces.[19] These tumors restrict on MRI with DWI that helps differentiate them from arachnoid cysts that do not restrict due to free water molecules.[20] Epidermoid tumors may show minimal postcontrast rim enhancement that is generally attributed to the peritumoral granulation secondary to leakage of irritant tumor contents.[21]

The goal in treating epidermoid tumors must be complete surgical resection. Most intracranial epidermoids occur predominantly in seven regions: anterosellar/frontobasal, parasellar/temporobasal, retrosellar/CPA, suprasellar/chiasmatic posterior fossa basal, intraventricular, and mesencephalic/quadrigeminal/pineal. The sellar region tumors are approaches via standard pterional craniotomy. Infratentorial lesions located centrally and in or around the fourth ventricle can be tackled via a midline suboccipital craniotomy, whereas CPA tumors are generally managed via a lateral retrosigmoid suboccipital approach. The tumors located in the pineal region (mesencephalic/quadrigeminal cisternal) can be managed via an infratentorial-supracerebellar approach. Supratentorial lesions, whether interhemispheric, cortical, or intraventricular, are managed in a tailor-made approach, depending on the exact anatomy of the lesion.[3] Although bicompartmental tumors can be managed using only a single-stage surgery, by following the conduit created by the tumor,[3] we have observed that it can be difficult to access remote areas and achieve total resection with preservation of the adherent vascular and neural structures via a single approach. Hence a staged procedure or a dual-access craniotomy is advocated by us in large bicompartmental lesions. It has been postulated that the growth rate of epidermoid tumors is one generation per month, essentially the same as normal human skin.[3,5] Therefore, any residual tumor can have a potential to recur in ones remaining life span. Another argument in favor of complete resection is that the risk of postoperative chemical/aseptic meningitis is directly related to the amount of retained tumor lining.[3] Although these lesions are slow growing and pliable, they can adhere to or encase adjacent vessels and nerves.[4] Therefore, although a complete resection may offer long-term cure and a normal life expectancy,[22] it can carry a high risk of morbidity. Piece meal reduction in the mass, with gentle teasing of the capsule, is the ideal technique for resection of these lesions. In most cases, a plane can be developed between the capsule and overlying arachnoid.[3] It is also advocated to check the postoperative field by using angled endoscopes to view areas of the cavity hidden from the microscopic view to achieve a total resection of the residual capsule.[23] Occasionally, a granulomatous reaction can cause a densely adherent capsule over vital structures that cannot be sacrificed, and small tufts of capsule can be left behind in these cases to avoid morbidity and with the hope that if recurrence develops, the rate of growth would be slow enough to obviate second surgery.[3] It has also been our observation that gross total excision of the tumors carries a high postoperative morbidity and prolonged intensive care stays. Out of 36 patients, 31 (86%) in whom prolonged ICU stay was required underwent gross total excision. On the flipside, seven (87.5%) out of our eight documented recurrences underwent only a subtotal resection.

With modern microsurgical techniques, gross total resection can be achieved in most cases. However, surgery in these tumors carries a small but significant risk of complications. The chances of aseptic meningitis can be reduced by avoiding spillage of contents in the subarachnoid space, complete removal of the tumor wall, and perioperative steroid administration.[11,24,25] We have been able to limit the rates of aseptic meningitis to 6.5% in our institute by following these practices and recommend perioperative steroid administration in all patients unless contraindicated. Though there are no available data to confirm the efficacy, intraoperative topical steroid administration via irrigation fluids was also practiced during our study. Though the CSF may turn out to be culture negative eventually, it is safer to start antibiotic therapy on suspicion of meningitis, because the initial spinal fluid profiles of these cases are similar to those of bacterial meningitis.[26]

Conclusion

Intracranial epidermoid tumors are benign, slow-growing, and potentially curable lesions. With advanced microsurgical techniques, complete resection should be the goal of treatment. With slow and meticulous piece meal removal, capsular bits can be teased from the surrounding structures in most cases. Angled endoscopes come in handy to review the nooks and corners of the operative field for any residual tumor, because these tumors conform to the cisternal spaces within which they lie without deforming the adjacent anatomical structures. Perioperative steroid administration and avoidance of spilling of contents in the subarachnoid spaces are essential to prevent chemical meningitis. Large and bicompartmental tumors can be best accessed by two different routes either in the same sitting or as a planned second surgery. Recovery of patients after excision of these lesions is excellent with low postoperative complication rates.

Conflict of Interest
None.

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