

# Analysis of Giant Intraventricular and Extraventricular Epidermoids, Defining Risk Factors for Recurrence, an Institutional Experience

## Abstract

**Background:** Multicompartmental intraventricular epidermoids behave differently from multicompartmental extraventricular lesions and localized lesions during its management. Few studies are available which have analyzed risk factors separately in these groups of cases for recurrence of these lesions and time to recur. **Materials and Methods:** In this retrospective observational study, 72 cases of intracranial epidermoid were treated over a span of 7 years. Cases were categorized into three groups. Group 1 comprised 15% (11/72) of cases with intraventricular multicompartmental, Group 2 with 22% (16/72) extraventricular giant tumors with multicompartmental involvement and size >4.5 cm, and Group 3 comprised 63% (45/72) of patients with lesions <4.5 cm and localized. Data pertaining to demography, clinical and radiological features, surgery performed, postoperative complication, histology, and follow-up were obtained from medical records available in the institute. **Results:** The average duration to treat was  $1.86 \pm 0.52$  (standard deviation [SD]) years, with headache as a major complaint in all the groups. Combined endoscope-assisted microsurgery was performed in 38.8% (28/72), microsurgery in 54.1% (39/72), and endoscopic excision in 6.9% (5/72) of cases. Tumor calcification was found in 23.6% (17/72) and preoperative capsular enhancement was seen in 19.4% (14/72) which persisted in 79% (11/14) of cases postoperatively on subsequent follow-up suggesting recurrence. On stepwise logistic regression analysis, preoperative capsular enhancement was a strong predictor of recurrence of tumor ( $P = 0.001$ ). The average follow-up was  $46 \pm 14.92$  (SD) months in Group 1,  $52.34 \pm 11.45$  (SD) months in Group 2, and  $63.36 \pm 18.42$  (SD) months in Group 3. **Conclusion:** Although the intracranial epidermoid is known to recur after long interval, tumor with specific characteristics can recur in short span of 5–6 years. Tumor characteristics such as preoperative capsular enhancement, multicompartmental distribution in vertebrobasilar territory, large size, and presence of calcification are strong predictors for recurrence. Performing endoscope-assisted microsurgery can decrease the postoperative morbidities but does not reduce the recurrence risk.

**Keywords:** Comparative study, different clinical features and management, extraventricular giant epidermoid, intraventricular giant epidermoid

## Introduction

Intracranial epidermoid comprises 0.2%–1.8% of all intracranial tumors with intraventricular lesion seen in 6%–10% of cases. Multicisternal involvement of intraventricular lesion is uncommon compared to the lesions located in extraventricular space such as cerebellopontine (CP) angle, sellar-suprasellar region, and quadrigeminal cistern. Involvement of multiple cisterns and compartment (supratentorial and infratentorial) makes the complete respectability of the tumor difficult, especially in one-stage surgery. The use of combined procedure involving

microsurgery and endoscopy both has been reported to increase the feasibility of complete resection of these tumors along with reduction in postoperative neurological complications, but the role of the combined procedure with neuronavigation assistance in decreasing the recurrence of tumor needs to be studied further. Tumor characteristics such as pre- and postoperative residual capsular enhancement, tumor calcification, location of the lesions, and multicisternal involvement are known to influence the recurrence rate. We aim to review the time to recur in the presence of these risk factors and redefine follow-up.

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## Materials and Methods

We retrospectively reviewed 72 cases of intracranial epidermoid operated in the Neurosurgery Department between July 2013 and July 2020. All the cases operated with confirmed radiological diagnosis, intraoperative pearly white appearance, and final confirmation on histology were included. Other resembling cystic lesions such as dermoid, arachnoid cyst, and lesion mimicking epidermoid as revealed on magnetic resonance imaging (MRI) brain with contrast, diffusion and flair images, apparent diffusion coefficient, and diffusion restriction images were excluded.

Cases were divided into three groups based on tumor location. Group 1 comprised intraventricular tumor of size >4.5 cm and extending into other compartments such as lamina terminalis, subchiasmatic, interhemispheric, and perimesencephalic cistern. Group 2 comprised cases with extraventricular location of size >4.5 cm and extending into nearby cisterns such as CP tumor extending into perimesencephalic cistern, quadrigeminal cistern lesion with extension into the third ventricle and transtentorial spread, and lesion in sellar-suprasellar location with extension into subchiasmatic, prepontine, and perimesencephalic cistern. Group 3 of intracranial epidermoid comprised lesions <4.5 cm and localized into single compartment only to CP region, quadrigeminal cistern region, or sellar/suprasellar region. The third group was considered as control to compare the behavioral pattern, clinical implication, and outcome in other two groups of giant multicompartamental tumor.

## Statistical analysis

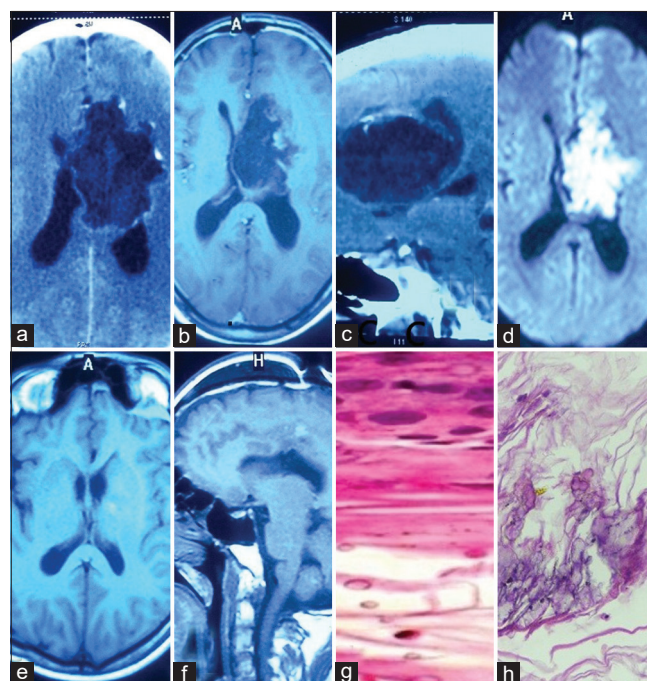
Continuous variables in this study were expressed as mean  $\pm$  standard deviation (SD) and were compared using independent *t*-tests. Categorical variables were expressed as number (percentage) and were compared with the Chi-square test or Fisher's exact test, as appropriate. Factors with  $P < 0.05$  in the univariate regression analysis were entered into the stepwise logistic regression analysis to identify the risk factors related to tumor recurrence. SPSS (Version 20.0; IBM Corp, Armonk, NY, USA) was used for statistical analysis. A two-tailed  $P < 0.05$  was considered statistically significant.

## Results

There were 15.7% (11/72) of patients in Group 1 with giant size intraventricular epidermoid with extraventricular extension, 22.22% (16/72) with extraventricular giant size epidermoid with multicompart spread in Group 2, and 62.5% (45/72) cases in Group 3 with localization to single compartment and size <4.5 cm [Table 1].

The mean age at presentation in Group 2 was  $41.63 \pm 8.95$  (SD) years which was slightly lower as compared to Group 1 and Group 3. There was a male predominance with the ratio of 1.2:1 (male:female) in all

the three groups. In Group 1, frontal horn epidermoids 27.27% (3/11) showed the tendency to involve interhemispheric fissure 67% (2/3) and medial frontal lobe [Figure 1a-h]. Temporal horn lesion 44.44% (4/11) had propensity to grow in subtemporal location and extended into cerebellomesencephalic and prepontine cistern [Figure 2a-d]. Atrial lesions were of largest size with an average diameter of 6.5 cm and extension into temporal horn [Figure 3a-e], whereas fourth ventricular tumor had inclination to invade into cervicomedullary cistern [Figure 4a-e]. There was almost equal distribution of lesions inside different compartments of the ventricles. Among Group 2 lesions, CP lesions were most common 50% (8/16) with extension to multiple cisterns as in perimesencephalic 62.5% (5/8) and in subchiasmatic cistern 12.5% (1/8) [Figure 5a-d]. Among giant quadrigeminal cistern, the extension was found into the third ventricle 60% (3/5) and superiorly into posterior interhemispheric fissure. Spread in all these locations was found along the course of major vascular territory. In Group 1, the extension was along choroidal vessel territory and partly basilar artery as in case of temporal horn lesion with subtemporal extension. In Group 2, extension of the lesions was along vertebrobasilar system (87.5%) and in internal carotid artery (ICA) territory (12.5%) [Table 1].

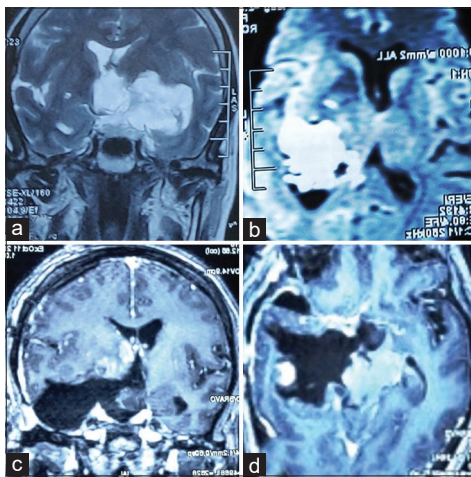


**Figure 1:** (a) Contrast-enhanced computed tomography head suggesting dystrophic calcifications and marginal capsular enhancement. (b) Lesion is hypointense on axial T1 magnetic resonance imaging. (c) Dilated lateral ventricles on sagittal magnetic resonance imaging. (d) Diffusion restriction on axial magnetic resonance imaging. (e) No residual lesion on axial T1 contrast. (f) No residual lesion on sagittal contrast magnetic resonance imaging. (g) Histopathology suggesting of squamous epithelial lining, keratin debris, and cells with empty nucleus. (h) Specs of calcification scattered among keratin deposits

**Table 1: Primary site of origin of tumors in cisterns and its extension into multiple other cisterns**

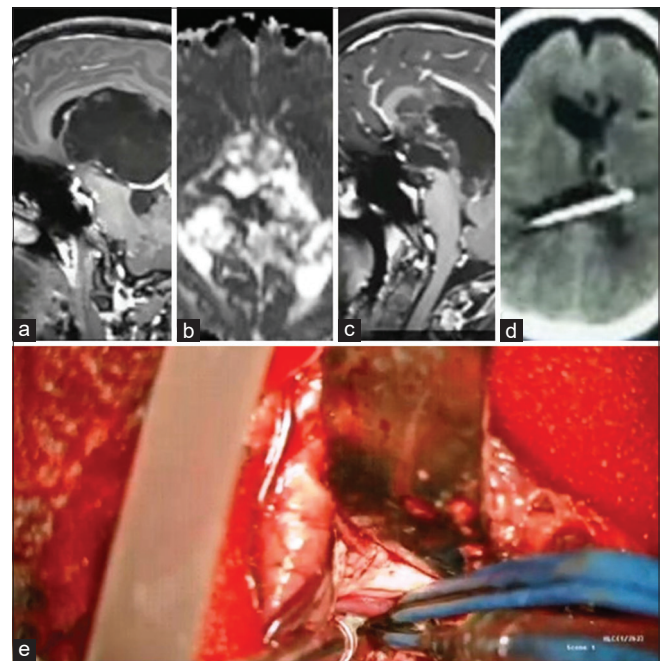
Cisterns Cases (n=36)	Intraventricular giant tumor Group 1 (n=11)		Extraventricular giant tumor Group 2 (n=16)		Control Group 3 (n=45) Lesion of size ≤4.5 cm, localized to single compartment (n=62.5%; 45/72)
	Intraventricular giant tumor (n=15.2%; 11/72)	Extension of intraventricular tumors into cisterns	Extraventricular giant tumor (primary site of origin) (n=22.2%; 16/72)	Extension into nearby cisterns (multicompartmental)	
Subchiasmatic	-	-	-	18.75 (3/16)	
Sellar and suprasellar	-	-	18.7 (3/16)		24.4 (11/45)
Quadrigeminal cisterns	-	-	31.25 (5/16)		22.2(10/45)
Cerebellomesencephalic cistern	-	-	-	31.25 (5/16)	
cerebellopontine	-	9.09 (1/11)	50 (8/16)*		53.3 (24/45)
Cervicomedullary cistern	-	9.09 (1/11)		31.25 (5/16)	
Interhemispheric fissure	-	18.18 (2/11)			
Perimesencephalic cistern	-	27.27 (3/11)		31.25 (5/16)	
Ambient cistern	-	27.27 (3/11)		31.25 (5/16)	
Middle fossa, parasellar, cavernous	-	27.27 (3/11)		31.25 (3/16)	
Prepontine cistern		27.27 (3/11)		31.25 (5/16)	
Frontal horn of lateral ventricle	27.2 (3/11)			-	
Temporal horn of lateral ventricle	27.2 (3/11)			-	
Atria of lateral ventricle	18.18 (2/11)			-	
4 <sup>th</sup> ventricle	27.2 (3/11)			-	

-: No data available, \*Significant association with  $P \leq 0.05$



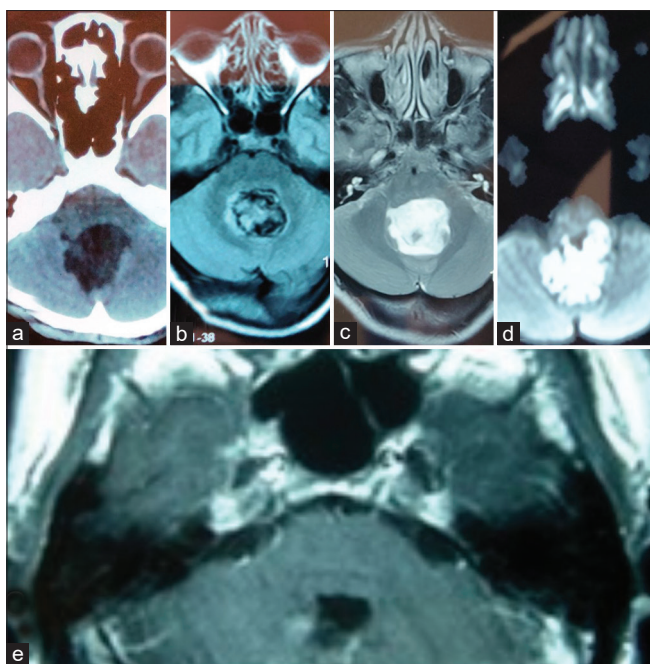
**Figure 2:** (a) Hyperintense lesion with multilobulated appearance in the left temporal horn with subtemporal extension on coronal T2 magnetic resonance imaging. (b) Axial magnetic resonance imaging image shows diffusion restriction on DW MRI sequence. (c) No residual lesion on coronal contrast magnetic resonance imaging sequence. (d) Axial contrast T1 magnetic resonance imaging sequence revealing marginal capsular enhancement in the vicinity of lesion in mesencephalic cistern where preoperative contrast enhancement was present

Chronic headache was the most common presentation, and it was uniformly observed in all the cases. In Group 1 cases with intraventricular location, blurring of vision was the second common symptom present in 72.72% (8/11) of cases and was associated with papilledema. Group 1 cases have seizures such as presenting features in 36.36% (4/11), limb weakness 27.27% (3/11), and vertigo and nystagmus in 36.36% (4/11) of cases. In Group 2, multiple cranial nerve palsy was the most common complaint followed



**Figure 3:** (a) Noncontrast-enhancing lesion in atria and body of lateral ventricle hypointense on axial contrast preoperative magnetic resonance imaging. (b) Diffusion restriction on axial magnetic resonance imaging. (c) Postoperative sagittal contrast magnetic resonance imaging revealed complete excision. (d) Intraoperative image showing capsular dissection with calcification present in the wall with tracing of the lesion along choroidal vessels. (e) Postoperative computed tomography head revealing complete excision of lesion with shunt tube *in situ*

by headache in 56.25% (9/16), whereas in Group 3, seventh and eighth nerve palsies and seizures were seen in 6.66% (3/45) of cases [Table 2].

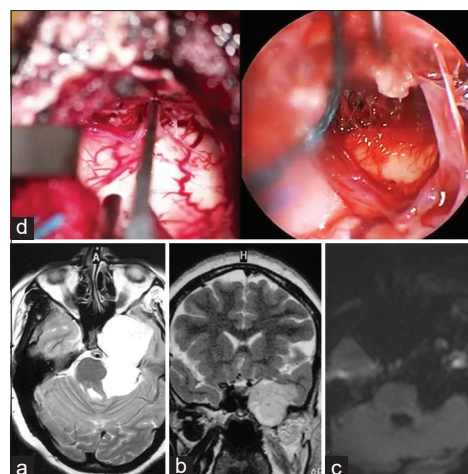


**Figure 4:** (a) Computed tomography head suggestive of hypodense lesion located in the fourth ventricle with expansion of the fourth ventricle. (b) On T1 axial magnetic resonance imaging, lesion in the fourth ventricle is hypo- to hyperintense. (c) On T2 axial magnetic resonance imaging, lesion in the fourth ventricle is hypo to hyperintense. (d) On magnetic resonance imaging, diffusion restriction was found. (e) Noncontrast computed tomography head showing complete excision of the lesion

In Group 1, different surgical approaches were employed depending on tumor location such as subfrontal transcortical approach for frontal horn lesion in two cases and interhemispheric approach in one case; similarly, lesion in the atrium was approached through the right superior parietal lobule and fourth ventricular tumor was approached through suboccipital telovelar approach. In Group 1, additional second-stage procedure with subtemporal approach was required in two cases and anterior petrosal approach in one case suggesting multistage surgery in 27% (3/11) of cases. In Group 2, CP lesions and multicisternal involvement were primarily dealt with retrosigmoid approach and additional procedure with subtemporal approach was required in 43% (7/16) of cases despite using combined procedure (microscopic with endoscopic assistance) [Figure 5a-d].

Thirty-nine cases were treated by microsurgery alone, 28 cases were treated by combined procedure (microsurgery and endoscopy both), and 5 cases were treated by endoscopy as a single procedure. In tumor with multicompartiment spread, combined procedure was performed in 8 cases of Group 1 and 10 cases of Groups 2 and 3 each [Table 3].

Staged procedure for residual lesions was required in seven cases who were treated by microsurgery and five cases who were treated by combined procedure (endoscope-assisted microsurgery) [Table 3].



**Figure 5:** (a) Axial T2-weighted magnetic resonance imaging showing hyperintense lesion in right cerebellopontine tumor with extension to the petroclival region. (b) Diffusion-weighted magnetic resonance imaging image of the same lesion shows restriction. (c) Postoperative diffusion weighted image shows complete excision with no restriction observed. (d) Intraoperative image of endoscope-assisted microscopy of capsular portion of epidermoid tumor in cerebellopontine region

Postoperative capsular enhancement was noticed in six cases who had undergone staged microscopic surgeries and five cases who underwent combined surgery. There was no procedural (staged microsurgery alone or combined procedure) benefit observed in terms of postoperative residual contrast enhancement on subsequent follow-up [Table 3].

Eleven of the 27 lesions with size >4.5 cm size had capsular enhancement, whereas only 6 of the 45 (tumor <4.5 cm) had capsular enhancement preoperatively, and there was a significant association between tumor size and postoperative contrast enhancement with  $P = 0.003$  [Table 4].

Preoperative capsular enhancement was present in three cases of Group 1, six cases of Group 2, and five cases in Group 3, but postoperative contrast enhancement was seen in three cases in Group 1, six cases in Group 2, and two cases in Group 3 [Table 5].

Capsular enhancement was present in 14 cases preoperatively and 11 cases postoperatively, and this association was significant with  $P = 0.0001$ .

19.44% (14/72) of cases with preoperative intratumoral calcification had capsular contrast enhancement, and this association was found to be significant with  $P = 0.001$  [Table 6].

Capsular calcifications were present in 22.2% of choroidal artery territory which were intraventricular, 58.3% of vertebrobasilar (cerebellomedullary, CP, and perimesencephalic cistern), 8.3% of ICA (sellar-suprasellar cistern), and 11.1% of combined territory of vertebrobasilar with choroidal vessels in case of lesion extending from ventricular to subtemporal location. The calcified capsules in these territories were difficult to excise completely because

**Table 2: Demography and clinical profile of multicompartmental epidermoids**

Total number (n=36) clinical features	Intraventricular giant tumor Group 1 (n=11)		Extraventricular giant tumor Group 2 (n=16)		Control Group 3 (n=45)	P
	Intraventricular giant tumor preoperative (n=11)	Postoperative on follow-up	Extraventricular giant tumor preoperative (n=16)	Postoperative on follow-up	Control (n=9)	
	Age (years)	41.63±8.95 (SD)		44.5±28.38 (SD)		
Sex	Male=6, female=5 (1:2)		Male=9, female=7 (1.28)		Male=25, female=20 (1.25)	
Headache (variable degrees)	90.90 (10/11)*	54.54 (6/11)	22.22 (8/16)*	37.5 (6/16)	77.77 (35/45)*	
Weakness (variable distributions)	36.36 (4/11)	18.18 (2/11)	31.25 (5/16)	31.25 (5/16)		
Hearing loss (variable degree)	18.18 (2/11)	9.09 (1/11)	43.75 (7/16)	43.75 (7/16)	33.33 (15/45)	
Diplopia	18.18 (2/11)	-				
Gait disturbance and ataxia	27.27 (3/11)	18.18 (2/11)	31.25 (5/16)	31.25 (5/16)		
Trigeminal neuralgia	-	-	25 (4/16)	100 (0/16)	11.11 (5/45)	
Facial numbness	9.09 (1/11)	9.09 (1/11)	-	-	11.11 (5/45)	
Facial weakness (variable degree)	9.09 (1/11)	9.09 (1/11)	-	-	-	
Vertigo and tinnitus	36.36 (4/11)	27.27 (3/11)	56.25 (9/16)*	12.5 (2/16)	44.44 (20/45)	
Seizures	36.36 (4/11)	27.27 (3/11)	25 (4/16)	100 (0/16)	22.22 (10/45)	
Deterioration of vision	72.72 (8/11)	63.63 (7/11)	37.50 (6/16)	6.25 (1/16)	33.33 (15/45)	
Hydrocephalus	90.90 (10/11)*		31.25 (5/16)	6.25 (1/16)	-	
Papilledema on fundoscopy	72.72 (8/11)*	54.54 (6/11)	31.25 (5/16)	6.25 (1/16)	-	

:- No data available, \*Significant association with P≤0.05. SD - Standard deviation

**Table 3: Surgical procedure performed and recurrence of the lesion**

Procedure performed	Intraventricular-multicisternal (Group 1)	Extraventricular-multicisternal (Group 2)	Control (Group 3)	Repeat surgery with recurrence	P
Microscopic surgery	3	5	31	6	0.086
Combined microscopic and endoscopic	8	10	10	5	0.058
Endoscopic alone	0	0	5	0	-
Recurrence	3	5	3	11	0.067

**Table 4: Size of the tumor and capsular calcification present**

Capsular calcification	Size of the tumor (cm)				Total (%)
	2.5-3.5	3.5-4.5	4.5-5.5	>5.5	
Absent	8	18	29	6	61 (84.7)
Present	0	2	2	7	11 (15.3)
Total (%)	8 (11.1)	20 (27.8)	31 (43.1)	13 (18.1)	72
P			0.003	0.003	

**Table 5: Size of the tumor and preoperative capsular enhancement**

Contrast enhancement of the capsule	Size of the tumor (cm)				Total (%)
	2.5-3.5	3.5-4.5	4.5-5.5	>5.5	
Absent	8	17	28	5	58 (80.6)
Present	0	3	3	8	14 (19.4)
Total (%)	8 (11.1)	20 (27.8)	31 (43.1)	13 (18.1)	72
P			0.003	0.003	

of chances of injury to perforating vessels [Table 7]. Total removal was achieved in 81% of Group 1, 57% of Group 2, and 96% of Group 3 cases, which was confirmed on postoperative MRI and computed tomography head on

follow-up. Residual calcified capsule had been left after surgery in 19% (2/19) of Group 1, 43% (7/16) of Group 2, and 4% (2/72) of Group 3 cases [Table 3].

On stepwise multiple regression analysis, preoperative capsular enhancement was the only risk factor which retained the power of significance with P = 0.0086 and area under curve on response operative curve of 0.88 [Table 8 and Figure 6].

There was a mild improvement in Karnofsky score following surgery in all the three groups, and there was no significant difference noted among all the three groups on postoperative Karnofsky and Glasgow Outcome Score [Table 9]. Mortality was observed in 9.09% (1/11) in Group 1 compared to 6.25% (1/16) in Group 2 due to recurrent meningitis and septicemia whereas no mortality occurred in group 3. Earliest recurrence observed in the present study in the form of increase in size of capsular enhancement was 4 years in a Group 2 case [Figure 2d]. Recurrence was more common in Group 2 cases as compared to Groups 1 and 3 as noticed on analysis of time to recur on Kaplan–Meier survival curve [Figure 7]. There was residual capsular remnant in 27.27% (3/11) in Group 1, 31.2% (5/16) in Group 2, and 6.66% (3/45) in Group 3 cases with mild increase

**Table 6: Tumor type and preoperative capsular enhancement**

Contrast enhancement of the capsule	Type of the tumor group			Total (%)
	Multicompartmental intraventricular	Multicompartmental extraventricular	Control	
Absent	5	10	43	58 (80.6)
Present	6	6	2	14 (19.4)
Total (%)	11 (15.3)	16 (22.2)	45 (62.5)	72
<i>P</i>	0.034			

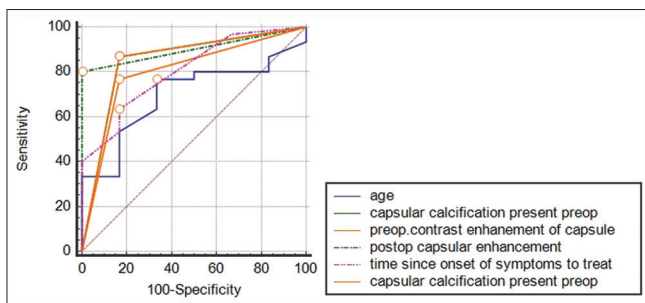
**Table 7: Vascular territory and capsular calcifications in multicompartment epidermoid**

Capsular calcifications	Arterial territory involved				Total (%)
	Choroidal	Vertebral	Internal carotid artery	Vertebral plus choroidal	
Present	2	32	0	4	38 (52.8)
Absent	14	10	6	4	34 (47.2)
Total (%)	16 (22.2)	42 (58.3)	6 (8.3)	8 (11.1)	72
$\chi^2$	13.192				
df	3				
Significance level	<i>P</i> =0.0042				
Contingency coefficient	0.518				

**Table 8: Stepwise multiple logistic regression analysis of risk factors for recurrence**

Variable	Coefficient	SE	OR	95% CI	Wald	<i>P</i>
Capsular calcification present	-1.35661	1.03757	0.2575	0.0337-1.9680	1.7095	0.1910
Cistern involved	-0.63104	0.59852	0.5320	0.1646-1.7196	1.1116	0.2917
Preoperative capsular enhancement	-3.43314	1.30740	0.0323	0.0025-0.4187	6.8956	0.0086
Constant	10.81632	3.33395	-	-	10.5254	

SE - Standard error; OR - Odds ratio; CI - Confidence interval

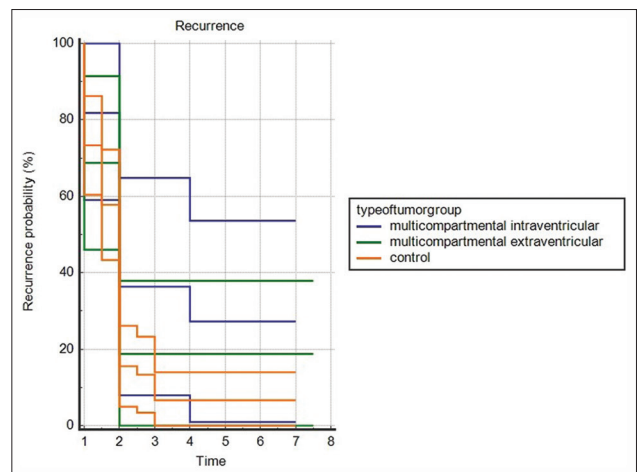


**Figure 6: Comparative response operative curve showing high predictive value of preoperative capsular enhancement for recurrence, with area under curve of 0.88**

in tumor bulk in the vicinity of excised lesion suggestive of recurrence observed till the last follow-up in Group 1 of  $46 \pm 14.92$  (SD), in Group 2 of  $52.34 \pm 11.45$  (SD) months, and in Group 3 of  $63.36 \pm 18.42$  (SD) months [Table 9].

### Discussion

Different theories have been proposed regarding the genesis of this lesion such as embryonic inclusions, trauma, and differentiation of multipotent cell rest and epithelial cell remnants while few have suggested that the cyst arises from pial tissues in plexal tufts. True epidermoid arises from ectodermal cell rest present since birth and ectopically



**Figure 7: Kaplan-Meier survival curve showing short-term recurrence of the lesion of epidermoids in the present study**

present anywhere in temporal bone depending on where the cell rests are situated. The origin of these tumors near temporal bone region can be explained by the above theory, but uncommon site like intraventricular region supports the hypothesis of origin from pial tissue of plexal tufts.<sup>[1-3]</sup> Unlike epidermoid of the ear, these lesions remain noninfected.

**Table 9: Postoperative results of giant multicompartmental epidermoid**

Parameters	Intraventricular giant tumor (n=11)	Extraventricular giant tumor (n=16)	Control (n=45)
Total removal (percentage cases)	81.81 (9/11)	75 (13/16)	93.34 (42/45)
Subtotal resection	18.18 (2/11)	25 (4/16)	6.66 (3/45)
Residual calcified capsule	18.18 (2/11)	25 (6/16)	6.66 (3/45)
Noninfective complications (percentage cases)	-	-	-
Cranial neuropathy (percentage cases)	18.18 (2/11)	62.5 (10/16)*	22.22 (10/45)
Weakness in limbs	18.18 (2/11)	43.75 (7/16)	22.22 (10/45)
Hydrocephalus	54.54 (6/11)*	6.25 (1/16)	-
Brain infarct after perforator injury	9.09 (1/11)	37.50 (6/16)	-
CSF leak from wound and wound bulge	18.18 (2/11)	25 (4/16)	11.11 (5/45)
Infective complications (percentage cases)	-	-	-
Septic meningitis	63.63 (7/11)*	18.75 (3/16)	-
RTI	36.36 (4/11)	13 (4/16)	11.11 (5/45)
UTI	18.18 (2/11)	12.50 (2/16)	-
KPS score (preoperatively/last follow-up)	82.3±17.9/84±19 (SD)	76.38±18.9/79±26 (SD)	84.8±19.4/86±18 (SD)
Glasgow outcome score (on last follow-up)	4.2±2.31/5±2.4 (SD)	4.89±2.84/5±1.2.8 (SD)	4.87±3.12/6±1.9 (SD)
Mortality	9.09 (1/11)	6.25 (1/16)	0
Duration of follow-up (months)	46±14.92 (SD)	52.34±11.45 (SD)	63.36±18.42 (SD)

-: No data available, SD - Standard deviation of mean; CSF - Cerebrospinal fluid; UTI - Urinary tract infections; RTI - Respiratory tract infections; KPS - Karnofsky Performance Scale, P = 0.069

The mean age at presentation was 41 years with the youngest case at 6 years in the intraventricular group and oldest at 70 years. The average age of detection of intracranial epidermoid is 40 years in different studies which is similar to the present study.<sup>[4-6]</sup> There was no specific gender prevalence reported in intracranial location, but in our study, it was 1.2:1 with slight male predominance.

Group 1 cases with giant intraventricular lesion presented earlier with clinical features of raised intracranial pressure (ICP) as compared to other groups. This may be due to obstruction of cerebrospinal fluid pathway leading to recurrent waxing and waning headache with papilledema and blurring of vision. In extraventricular lesions of Groups 2 and 3, patients presented late in the disease course with multiple cranial nerve palsy. This delay might be due to multiple cistern involvement which let the tumor grow in anatomical contour and thus preventing its early detection.<sup>[7-10]</sup> In this study, a percentage of cases with intraventricular tumor (Group 1) who presented with features of raised ICP and blurring of vision were higher as compared to other studies.<sup>[6-11]</sup> This may be due to difference in tumor size of the study group. Although the tumor in temporal horn had been reported in the literature, presentation with seizures was seen in only a few cases even in the presence of parenchymal infiltration. The mechanism of development of this clinical feature may be attributed to infiltration of brain tissue by cyst, chemical meningitis, or architectural changes in epileptogenic area. Only 1.55% of cases of intracranial epidermoid have parenchymal invasion, especially in temporal location, and presentation with seizures is uncommon.<sup>[5,6]</sup> In this

study, cases with clinical features of seizure were higher as compared to other studies.<sup>[8-10]</sup> Intraventricular tumors extending to posterior fossa (CP angle) may present with features of localized cranial nerve palsy as observed in our study. Cases in the control group have no features of raised ICP, but seventh and eighth cranial nerve involvements in 33.33% (3/9) of cases and one case, respectively, have frontal lobe features with antisocial behavior.<sup>[9,10]</sup> We did not find any correlation between duration of onset of clinical features and symptoms correlating with recurrence.

In this study on MRI, 20% of epidermoid tumor had multiloculated hypoattenuating appearance with occasional calcification seen as hyperattenuating area with occasional peripheral capsular contrast enhancement. Peripheral capsular enhancement may be due to inflammatory process at the interface of tumor and normal brain parenchyma or active proliferation of squamous epithelial lining. In the current study, intratumoral calcification was observed in 22% of intraventricular and 58.3% of extraventricular tumor, which is comparatively higher to other studies who reported the 5%–18% incidence of calcification.<sup>[12-15]</sup> This variation may be due to prolonged duration to treat in the present study.

In the present study, strength of association between calcification in the tumor and cyst recurrence is similar to that reported by Yasargil *et al.*, Fornari *et al.*, Chowdhary *et al.*, and Desai *et al.* but was higher compared to the study by Sabin *et al.*<sup>[12,16-19]</sup> It suggests that postsurgical remnant of active proliferating component of tumor capsule in vicinity of the calcifications leads to subsequent recurrence.

In the present study, capsular enhancement in primary tumor (19.4%) and subsequently on postoperative follow-up (15.2%) was higher as compared to the study by About *et al.* who reported capsular enhancement in 7.7% of the primary cases.<sup>[20]</sup> Kallmes *et al.* reported capsular enhancement on initial imaging in 33% of cases which is higher than the incidence reported in the present study.<sup>[21]</sup> In this study, radiological features of capsular enhancement were suggestive of active component of the tumor and had been significantly associated with increased recurrence of the tumor. Sometimes, malignant transformations in the postoperative enhancing capsule had also been reported in literature.<sup>[22,23]</sup> These findings suggest that preoperative capsular enhancement has good predictive value for recurrence of the tumor and such cases need close follow-up.

In the study by Singh *et al.*, combined procedure facilitated the complete resection of tumor in 79% of cases. Tuchman *et al.* reported 70% and Ebner *et al.* reported 79% benefit in complete resection of the lesion which is in accordance with the findings of the present study.<sup>[24-26]</sup> In the current study, the number of cases with multicompartamental spread was more compared to other studies where staged operation was required. Multistage operation with good exposure of the lesion results in lesser recurrence as compared to incomplete exposure in single-stage surgery as demonstrated in many studies.<sup>[12-17]</sup> In the present study, we did not notice significant influence of operative procedure on recurrence of the tumor though we did observe decreased postoperative complication like septic meningitis and cranial nerve palsy in the combined surgery group as compared to staged procedures.

It is difficult to distinguish epidermoid capsule intraoperatively from arachnoid membrane. Frozen section sent intraoperatively in these cases was mostly nonconclusive in the present study and other studies too.<sup>[14-16]</sup> In such circumstances, imaging characteristics such as capsular enhancement and calcifications are better parameters for prediction of recurrence. Recurrence in these tumors had been reported as early as 1 year by Rutherford *et al.*<sup>[27]</sup> Talacchi *et al.* reported median time of recurrence of 8 years,<sup>[28]</sup> whereas 10% recurrence at 5 year and 19% recurrence at 10 years were reported by Ren *et al.* in their study on atypical intracranial epidermoids.<sup>[29]</sup> In this study, 15.2% recurrence at 5 years had been observed on follow-up which is similar to these studies. In our study, we observed tumor calcification and capsular enhancement to be more common in the region of vertebrobasilar distribution as compared to choroidal and ICA vascular territory. These are the transitional zone around tentorium cerebelli where multiple perforators supplying brain stem are present. The presence of adherent capsule and calcification in these regions together with limited visualization even with endoscope-assisted microsurgery makes the complete

excision of the tumor difficult and risky. Intraoperative confirmation to assess completeness of resection in these areas may increase the risk of neurovascular complication, and we could not find frozen section from suspected site of much benefit.

There are some conflicting reports in the literature regarding capsular enhancement, but the predominant opinion is that they do not enhance, presumably due to minimal vascularity. However, there are two reports describing rim enhancement in cases as reported in the study by Talacchi *et al.* and Kallmes *et al.*<sup>[21,28]</sup> These studies suggested that the enhancement was subtle and not completely circumferential as found in the present study also. Enhancement of periphery is well documented in few cases reported with malignant degeneration in residual lesion.<sup>[22,23]</sup>

Adjuvant radiotherapy can be considered in cases with multiple recurrence as suggested in the study by Morshed *et al.*<sup>[30]</sup> Gamma Knife surgery had been reported to be effective CP angle epidermoid with trigeminal neuralgia, where reduction in tumor size results in relief of symptoms, but long-term follow-up is missing regarding effect of radiotherapy on the recurrence of tumor.<sup>[31]</sup> In the present study, no patient was advised radiotherapy, but it can be considered in future follow-up on multiple recurrence.

## Conclusion

Although the intracranial epidermoids usually recur after long interval, it may recur after short span too as observed in the present study. Tumor characteristics as preoperative capsular enhancement together with multicompartamental distribution in vertebrobasilar territory, large size, and presence of calcification had demonstrated high predictive value for recurrence. Performing endoscopy-assisted microsurgery can decrease the postoperative morbidities, but it may not reduce the recurrence of the lesions.

## Patient consent

Detailed written informed consent had been taken at the time of enrollment for this study from the patient/next of kin/guardian to use the information and their data for teaching and clinical research purposes.

This manuscript is a unique submission and is not being considered for publication with any other source in any medium.

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## Conflicts of interest

There are no conflicts of interest.

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