ORIGINAL ARTICLE



Intracranial epidermoid tumor; microneurosurgical management: An experience of 23 cases

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ABSTRACT

Objectives: An intracranial epidermoid tumor is relatively a rare tumor, accounting for approximately 0.1% of all intracranial space occupying lesions. These are also known as pearly tumor due to their pearl like appearance. In this series, the localization of the tumor, presenting age and symptoms, imaging criteria for diagnosis, surgical management strategy with completeness of excision and overall outcome were studied prospectively. Here, we report our short experience of intracranial epidermoid as a whole.

Materials and Methods: Between January 2006 to December 2010, 23 cases of intracranial epidermoid were diagnosed preoperatively with almost certainty by computed tomography (CT) and magnetic resonance imaging (MRI) of brain in plain, contrast and other relevant studies. All of them underwent operation in Dhaka Medical College Hospital and in some Private Hospital in Dhaka, Bangladesh. All patients were followed-up routinely by clinical examination and neuroimaging. Average follow-up was 39 (range-71-11months) months. Patients of the series were prospectively studied.

Results: Supratentorial epidermoids were 04 cases and infratemporal epidermoids were 19 cases. Clinical features and surgical strategy varies according to the location and extension of the tumors. Age range was 19-71 years (37.46 years). Common clinical features were headache, cerebellar features, seizure, vertigo, hearing impairment and features of raised intracranial pressure (ICP). Investigation was CT scan or/+ MRI of brain in all cases. Pre-operative complete excision was 20 cases, but post-operative images showed complete excision in 17 cases. Content of tumor was pearly white/ white material in all cases except one, where content was putty material. Re-operation for residual/recurrent tumor was nil. Complications included pre-operative mortality one case, persisted sixth nerve palsy in one case, transient memory disturbance one case, and extra dural hematoma one case. One senior patient expired three months after the operation from spontaneous intracerebral hemorrhage. Rest of the patients were stable and symptom/s free till last follow-up.

Conclusion: In the management of such tumors, one should keep in mind that an aggressive radical surgery carrying a high morbidity and mortality and a conservative subtotal tumor excision is associated with a higher rate of recurrence, but earlier diagnosis and complete excision or near total excision of this benign tumor can cure the patient with the expectation of normal life.

Key words: Epidermoid tumor, intracranial, neurosurgical management

Introduction

Intracranial epidermoid appears like a cyst in neuroimaging studies. They are also known as cholesteatomas and pearly tumors. They are usually congenital benign lesion and

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Dr. Forhad Hossain Chowdhury, Neurosurgeon, Department of Neurosurgery, Dhaka Medical College Hospital, Dhaka, Bangladesh. E-mail: forhadchowdhury74@yahoo.com approximately 0.1% of all intracranial tumors. They may arise from displaced dorsal midline ectodermal cell rests between the third and fifth weeks of embryogenesis during neural tube closure. However, Cerebello-pontine angle (CPA) and parasellar location could be explained by the result of proliferation of multipotential embryogenic cell rests or lateral displacement of ectodermal cells by the developing otic vesicles.^[1,2] They grow by the accumulation of keratin and cholesterol, which are breakdown products created by the desquamation of epithelial cells.^[3] They have tendency to grow along available cisternal spaces. So there is no mass effect initially and remain asymptomatic for many years.^[4-7] Symptoms are mainly due to mass effect or involvement of cranial nerves. There are diverse opinions regarding the completeness of the excision especially when the tumor is extensive and where there is firm adhesion to pia matter. Modern neuroimaging helps in near confirm diagnosis pre-operatively, which helps in planning of proper

surgical approach for any individual cases. In case of elderly patients with shorter life expectancy, controversies are more regarding the tumor operation.

Materials and Methods

Between January 2006 to December 2010, 23 cases of intracranial epidermoids were diagnosed pre-operatively with almost certainty by computed tomography (CT) and magnetic resonance imaging (MRI) of brain in plain, contrast and other relevant studies. All of them underwent operation in Dhaka Medical College Hospital and in some private Hospital in Dhaka, Bangladesh. All patients were followedup routinely by clinical examination and neuroimaging. Average follow-up was 39 (range 71-11 months) months. Patients of the series were prospectively studied. Presenting symptoms were recorded at the first visit and re-assessed just before operation and findings were recorded accordingly. Approaches of operations, preoperative findings were also recorded. No attempt was made to analyze the outcome as the sample size was small in this mini clinical series of rare intracranial tumors.

Observations and Results

Details of observations, findings, operations, and results are shown in Table 1.

Total number of patients was 23. Male patients outnumbered the female (M = 19, F = 04). Age range, 19-71 years (Average-37.46 years). Only four cases were supratentorial and rest were infratentorial. Among the supratentorial epidermoids, two cases were in ineterhemispheric fissure, one case was in cavernous sinus, and one case was in suprasellar prelamina terminalis area. Tumor distribution was eight cases in CPA, two cases in 4th ventricle, four cases in pineal region, two cases in pre-brainstem and basal cistern area, one case in cerebellar hemispheric fissure, and two cases in cerebellar hemisphere among the infratentorial cases. Clinical features were according to the location. Most common clinical features were headache, vomiting, and visual disturbance. Among the supratentorial cases epilepsy was common. Opthalmoplegia was found in cavenous sinus and premesencephalic epidermoid. In suprasellar case, there was psychosis, headache, vomiting, and altered level of consciousness with features of chemical meningitis. Cerebellar dysfunction was common in posterior fossa epidermoid. Cerebellar signs including gait ataxia, dysmetria, dysarthria and nystagmus were found in posterior fossa and 4th ventricular epidermoid (in eight cases). Among the CPA epidermoid, TN was common (four out of eight). Only CT scans was done for five patients; CT and MRI studies were performed in 10 patients and only MRI study were done in 8 cases. In the CT scan, hypodense lesion in the strategic localization with expansion of the mass along all the clefs of the basal cistern in the skull base which showed

minimal contrast uptake in the periphery. Only in suprasellar epidermoid, there was capsular calcification. In 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). In case of suprasellar epidermoid, pre-operative MRI of brain showed lesion intensity was more than CSF but less than the brain in T1W images, whereas lesion was hypointense than CSF in T2W images. At the same time, there was hyperintense material in both frontal horn and right temporal horn of lateral ventricle and different cisterns (most likely ruptured tumor content into ventricle and CSF spaces. Tumor material was lighter than CSF). Hydrocephalus with features of raised ICP was in six cases and pre-excision CSF diversion was not done in any case. Immediate post-operative patient recovered from features of raised ICP.

Different microneurosurgical approaches were used for epidermoid tumor at different sites [Table 1]. In one case ([Table 1] No 13), wrong sided (right sided) retromastoid retrosigmoid craniotomy was done and later, after closure of the wound, craniotomy was done on the pathological side. Among all the epidermoids, only in suprasellar epidermoid there was patchy calcification on the capsule with firm adhesion to surrounding structures. The other thing in this case was the content of this epidermoid was putty material, whereas content of all other tumors were same pearly white/ pearly white material. In two cases of CPA epidermoid causing TN, pre-operatively, there was deep indentation by the tumor at root entry zone where early post operatively patient developed facial dysesthesia that recovered within six weeks. Pre-operative total tumor removal (except adherent capsule) was in 20 cases. Whereas post-operative MRI showed 17 cases of total excision. Endoscope was used in three cases to check completeness of resection. Sitting posture was used only one case of pineal region epidermoid.

CPA epidermoid causing TN showed gratifying result after excision of tumor. Pre-operative hearing impairment (in two cases) persisted after operation. Early post-operative complications were extra dural hematoma (in suprasellar epidermoid, emergency reoperation needed), transient CSF rhinorrhea (in one CPA epidermoid), aggravation of Parinaud's syndrome (in one pineal epidermoid) and sixth nerve palsy. All recovered subsequently, except sixth nerve palsy. Preoperative cardiac arrest and death occurred in a patient with a large premesencephalic and basal cistern epidermoid. He had respiratory dysfunction from pulmonary tuberculosis and decision for operation was taken due to rapid deterioration of neurological status. The senior most member of the series recovered well, but died three months after the operation from spontaneous intracerebral hemorrhage. Rest of the patients were symptom free or in stable condition till last follow-up. Post-operative MRI showed no recurrence in complete excision cases and no increase in size of residual tumor. In one of pineal

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No.	Age/sex	Location	C/F	Operation	Complication/s	Last F/U	Remark
1	19/M	4 th ventricle [Figure 1]	Quadriparesis, cerebellar features and features of raised ICP	Suboccipital craniotomy and removal through telovelar approach	None	Complete recovery	No residual tumor on post op MRI
2	28/M	Lt. Suprachiazmatic prelamina terminalis subfrontal [Figures 2 and 3]	Psychosis, sudden headache, vomiting, visual disturbance, unconciousness/s of chemical meningitis	Lt. pterional craniotomy	EDH (and reoperation) memory deficit	Psychosis and memory deficit recovered	Small residual on CT scan
3	30/F	Lt. CP angle with prepontine and posterior subtemporal extension	Trigeminal neuralgia, hearing loss, gait instability, cerebellar features	Lt subtemporal trans tentorial	None	TN and other features resolved except hearing loss	Small residual in prepontine area around basilar artery and subtemporal area
4	28/M	Rt CP angle	Occipital headache and TN	Rt retrosigmoid suboccipital approach	None	Symptoms resolved	No residual on imaging
5	35/M	Supracallosal anterior interhemispheric (Rt>Lt)	Headache, epilepsy	Anterior Interhemispheric approach	None	Headache resolved, epilepsy is controlled (Pt. is on AED)	Complete excision on post op MRI
6	20/M	Pineal region [Figure 4]	Headache, parinaud's syndrome	Occipital transtentorial approach	Aggravation of Parinaud's syndrome	Persistent Parinaud' syndrome	No residual tumor on MRI
7	30/M	Pineal region [Figure 5]	Headache	Sitting, infratentorial supracerebellar approach	None	Symptom gone	Small ?residual on post op MRI
8	27/M	Pineal region	Headache (occipital)	Infratentorial supracerebellar approach	None	Symptom gone	No residual on MRI
9	38/M	Lt. prepontine and premesencephalic	Quadriparetic (RT>Lt), headache, obtunded recently	Lt subtemporal transtentorial approach planned	Cardiac arrest before craniotomy	-	Expired
10	52/M	Rt CP angle [Figure 6]	Intractable TN, (alcohol injection in trigeminal ganglion in abroad)	Rt suboccipital retrosigmoid approach	None	TN resolved	No residual on MRI
11	22/F	Lt. petroclival region (CPA)	Features of Raised ICP, cerebellar features, quadriparesis	Lt retrosigmoid suboccipital approach	Lt sixth nerve palsy	HCP and other features resolved except sixth nerve palsy	Small residual around basilar artery
12	38/M	Lt. premesencephalic and basal cistern	Rt hemiparesis, opthalmoplegia, headache	Rt subtemporal transtentorial approach	None	Symptoms resolved	No residual tumor on post op MRI
13	48/M	Lt CP angle	TN	Lt retrosigmoid suboccipital approach (There was mishap; first wrong sided craniotomy done)	CSF rhinorrhea (resolved spontaneously)	TN cured	No residual tumor on MRI
14	39/M	Pineal region	Occipital headache	Occipital transtentorial	none	Headache gone	Small residual on post op CT scan
15	32/M	Lt CP angle	Headache	Lt retrosigmoid suboccipital approach	None	Symptom gone	No residual on post op MRI
16	53/F	Rt sided posterior fossa(Recurrent/ residual)	Headache and gait instability	Rt lateral suboccipital approach	None	Symptoms improved	No residual tumor on MRI
17	42/M	Rt cavernous sinus	Headache and diplopia, sixth nerve palsy	Rt sided temporal craniotomy and interdural approach to cavernous sinus	None	Symptoms improved	No residual tumor

Table 1: Details of the cases of the series

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Table 1: (cont/-)								
No.	Age/sex	Location	C/F	Operation	Complication/s	Last F/U	Remark	
18	30/F	Posterior interhemispheric and suprasplenial and supracallosal region and pineal region	Headache, intarctable epilepsy	Post interhemispheric approach	None	Headache improved and epilepsy is controlled with AED	Residual tumor bilaterally away from midline	
19	71/M	4 th ventricle	Headache, vomiting, visual disturbance, gait instability	Suboccipital telovaler approach	Intracerebral hemorrhage o3 months after operation and expired	Patient improved from all symptoms before expired	_	
20	28/M	Rt CPA	Headache, right sided fall, and gait disturbance	Rtretromastoid retrosigmoid approach	None	Complete recovery	Complete removal	
21	41/M	Cerebellar interhemospheric	Headache, vomiting, vertigo, nystigmus, and visual disturbance	Suboccipital craniectomy	None	Full recovery	Complete removal	
22	34/M	Lt cerebellar hemisphere	Vertigo, headache	Suboccipital craniotomy	None	Symptoms gone	Complete removal	
23	44/M	Lt CPA	Headache, hearing impairment, vertigo, cerebellar features	Lt retromastoid retrosigmoid approach	None	Complete cure except hearing loss.	Complete removal	

M – Male; F – Female; Rt – Right; Lt – Left; CP – Cerebello-pontine; CPA – Cerebello-pontine angle; TN – Trigeminal neuralgia; ICP – Intra cranial pressure; MRI – Magnetic resonance imaging; HCP– Hydrocephalus; EDH – Extradural hematoma; AED – Anti epileptic drug; MRC – Medical research council; CSF – Cerebrospinal fluid; C/F – Clinical features; F/U – Follow-up; SI – Serial; Op-Operative and CT-computed tomography



Figure 1: (a) MRI of brain TW1 image, saggital view showing 4th ventricular epidermoid (Ep), (b) Peroperative view showing 1-incised dural margins, 2-cerebellar tonsil, 3-Posterior inferior cerebellar artery, 4-floor of 4th ventricle, and 5-epidermoid tumor

region epidermoid, post operative MRI showed residual tumor with hyperintensity [Figure 5c and d] in all image sequences (though pre-operatively tumor excision seemed to be complete and checked by endoscopy).

Representative case-1

A 28-year-old right handed male presented with sudden, severe, headache, vomiting, visual disturbance followed by unconsciousness [Table 1, No. 2, Figures 3 and 4]. He had history of three episodes headache, vomiting, visual

Asian Journal of Neurosurgery ______ Vol. 8, Issue 1, January-March 2013 disturbance followed by psychosis in last two years. He was on antipsychotic drugs. On clinical examination, he was obtunded and there were signs of meningeal irritation. CT scan showed a right sided preponderance cystic lesion with wall calcification suggestive of craniopharyngioma. MRI of brain showed a hypointense (hyperintense than CSF and hypointense than brain parenchyma, there was hyperintensity at the periphery of the lesion) lesion in predominantly right suprasellar suparchiasmatic pre-lamina terminalis area. There was also multiple small hyperintense lesion in both frontal horn, right temporal horn, and basal and posterior fossa CSF spaces (seemed the ruptured content of the tumor, lighter than CSF, causing chemical meningitis). The MRI diagnosis was not certain of epidermoid, dermoid, or craniopharyngioma. Pre-operatively, patient was treated with injectable steroid. Pre-operative exposure of tumor through right pterional craniotomy revealed epidermoid tumor with putty substance in the tumor. There was patchy calcification on the capsule with firm adhesion to surrounding tissue. It was seemed that tumor removal was complete. Patient recovered well from anesthesia. But subsequent recovery was not satisfactory and patient developed right sided hemiparesis with further deterioration of vision. CT scan of 4th POD showed left frontotemporal extradural collection with small residual tumor

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Figure 2: MRI of brainT1W images (a) saggital view showing suprachiasmatic prelaminaterminalis subfrontal epidermoid with hyper intense peripheral zone, (b) and (c) axial showing hyperintense epidermoid content in frontal and temporal horn (after rupturing into ventricle)



Figure 3: Axial CT scan of the same patient of Figure 3. a, b, and c (pre-operative)- showing suprasellar epidermoid with calcified wall, d, e, and f (post-operative)-showing left sided fronto-temporal extradural collection with tumor dead space due to failure to collapse tumor wall (calcified)



Figure 4: MRI of brain T1W images saggital views, (a) preoperative, showing pineal region epidermoid, (b) postoperative, showing no residual tumor



Figure 5: MRI of brain showing pineal region epidermoid tumor, A-axial view and (b) saggital view. (c) and (d) Post operative MRI axial and saggital view respectively showing hyperintense residual tumor

or dead space within tumor cavity that failed to collapse due to calcification. Re-operation was done immediately to remove the extradural hematoma. Then he recovered hemiparesis and started to recover vision, but developed recent memory deficit. From 3rd week after second operation, he began to recover from memory deficit and recovered completely after one year. He was free of symptoms till last follow-up (last two years). Neuroimaging (CT scan) after one year showed same residual tumor/dead space within tumor capsule.

Representative case-2

A 52-year-old male presented with right sided trigeminal neuralgia (TN) for the last 15 years for which he visited several physicians [Table 1, No. 10, Figure 6]. Last three years, it was intractable. Two years back, when he was in a foreign service, trigeminal ganglion was infiltrated with absolute alcohol in a foreign hospital. No neuroimaging was done at that time. MRI of brain showed a right sided CPA epidermoid. Tumor was microsurgically excised completely through right retromastoid retrosigmoid lateral suboccipital craniectomy. During operation a portion of tumor was found deeply impinging the root entry zone. After dura closure bony chips were repositioned at the craniectomy site, post-operative period was uneventful. His TN was cured but there was dysesthesia at the right sided face for six weeks which resolved spontaneously. Post-operative MRI showed no residual or recurrent tumor at the end of two years after operation.

Discussion

Epidermoids are slow-growing benign tumors and have a peak incidence in the fourth decade.^[8] Typically patients are



Figure 6: MRI of brain T1W images axial view. (a and b) preopeartive showing right cerebello-pontine angle and prepontine epidermoid. (c and d) Post operative, showing no residual tumor but there is residual dead space

between 20 and 40 years of age.^[9] There may be increased prevalence in males, although this is not found in all series.^[9,10] In our series, male predominance was in obvious. This socalled "beautiful tumor" has an irregular cauliflower-like outer surface that grows and encases vessels and nerves.^[11] The cyst content is characteristically composed of a pearly material that results from desquamation of the cyst wall. The cyst content is characteristically composed of a material is composed of keratin and cholesterol crystals.^[12] But in one of our cases, we found the content was putty material like other extracranial epidermoid which is probably rare in intracranial cases. About 90% intracranial epidermoids are intra dural and 7% of CPA tumors.^[13] Approximately 60% of all intracranial epidermoids occur in CPA. They are the third most common tumors of the CPA after acoustic neuromas and menengiomas.^[13] The second most prevalent location in the posterior fossa is the fourth ventricle accounting for 5-18% of all intracranial epidermoids.^[13] Fifteen percent of epidermoids occur in middle cranial fossa and spinal occurrence of epidermoid is very rare.^[13,14] Ten percent of epidermoids are extradural, mostly within skull.^[13]

Signs and symptoms of epidermoid cysts are due to gradual mass effect, with presentation including: Headaches (most common), cranial nerve deficits, cerebellar symptoms, seizures, raised intracranial pressure.^[13,15] Recurrent aseptic meningitis is uncommon but recognized similar to the less common dermoid cyst.^[13]

The major differential diagnosis for a epidermoid cyst are arachnoidal cysts, hamartomatous lipomas, dermoid cysts,

cystic neoplasms,^[11,16] neurocystocercosis, neuroenteric cyst etc. They can usually be differentiated by CT scan or MRI images of brain. Conventional MR images sometime cannot reliably be used to distinguish epidermoid tumors 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. In contrary, fluid-attenuated inversion recovery (FLAIR) and diffusion-weighted (DW) sequences can successfully be used for diagnosis of epidermoid cysts by revealing its solid nature.^[8,17-20] 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. Epidermoids characteristically present on this sequence as heterogenous lesions with central parts of the tumor being hyperintense relative to the hypointense CSF.^[8,19,21]

Hydrocephalus is said to be uncommon because of the long-standing nature of the lesion and also because CSF can permeate through the crevices of the lesion. Rarely, pre-excision CSF diversion is required.^[13] However, in the present study, six (24.5%) patients had ventriculomegaly with features of raised ICP, all improved after definitive surgery.

The surgical approach is generally determined by the location and the extent of the lesion. The lesion, when confined to the CPA, is approached by a retromastoid craniectomy, whereas significant supratentorial extension needs a combined retromastoid and subtemporal approach or a staged procedure. However, the tumor commonly extends into the hiatus and this incisural part can be completely removed with the posterior fossa approach.^[14,22] In one of our CPA epidermoid that extended in prepontine and posterior subtemporal area could not be removed completely due to arterial adhesion and encasement as well as limited exposure.

There is a controversy regarding the extent of removal. Although the aim of surgery is for complete removal, few authors advocate total removal of the tumor.^[22-24] It has been suggested that with microscopic meticulous sharp dissection, every bit of the capsule should be removed to prevent a recurrence.^[24] However, adherence of the capsule to the important neurovascular structures in and around the brain stem often leads to its incomplete removal.^[1] A conservative approach with decompression and the removal of the nonadherent portion of the capsule has been suggested by others.^[13,25-28] Coagulation of the residual capsule is also advocated to minimize recurrence,^[13] but is not widely practiced. It also may be dangerous to coagulate the remnant adherent capsule near the exquisitely sensitive cranial nerves, brain stem, or important vessels in the CPA. A relatively high incidence of cranial nerve dysfunctions has been mentioned in the previously reported series, the majority of which, however, improve by the time of discharge or during the follow-up period.^[22,25,28] In one of our patients, sixth nerve

palsy developed after surgery that did not recover even after two years. In cases where tumor capsule in firmly adhesive to surrounding structures, we did not go for dissection or coagulation of this part of capsule.

Chemical meningitis can occur by spillage of the cyst contents during operation, which usually is transient and self-limiting^[22] 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,^[25] and delayed withdrawal of steroids in the post-operative period^[24, 27, 29] have been advocated as possible measures for preventing chemical meningitis. We did not face any postoperative chemical meningitis. Communicating hydrocephalus can develop as a result of meningitis and might require CSF diversion procedures.^[25,30,31]

Nevertheless, epidermoids are often the most troublesome to cure because of their insinuating growth into different spaces and cisterns in addition to engulfing cranial nerves and vessels which make it difficult for radical excision.^[22] It is not surprising that prior to the microsurgical era, operative mortality ranged from 20% to 57%.^[27] But now-a-days, surgery induced mortality is very low as a result of trmendous improvent in neuroimaging and microsurgical skill as wel as conservative radical aproach in intracranial epidermoid excision.

Partial removal of a lesion leads to recurrence, which often occurs after a prolonged period as a result of the slow growth rate of the tumor.^[23,26] As the hypodense areas revealed by CT persist for a prolonged period, even after complete removal of the tumor, possibly because of a long-standing deformation of the neural structures, a diagnosis of recurrence at an early stage is often not possible. MRI is useful to diagnose an early recurrence.^[24,30,32] However, subsequent surgery is often indicated only when the recurrent lesion is symptomatic.^[30] In our series, follow-up period is small, probably for that reason we did not find any recurrence or symptomatic recurrence in residual tumors. Malignant transformation of an epidermoid cyst into a squamous cell carcinoma is a known but extremely rare occurrence. Malignant transformation is suggested by enhancement after contrast administration or rapid growth.^[19]

Conclusion

In the management of such tumors, one should keep in mind that an aggressive radical surgery carrying a high morbidity and mortality and a conservative subtotal tumor excision is associated with a higher rate of recurrence, but early diagnosis and complete excision or near total excision of this benign tumor can cure the patient with the expectation of normal life.

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