

## Research Article

# Microsurgical excision of intracranial epidermoids: a short surgical series of 12 cases

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### ABSTRACT

**Background:** Intracranial epidermoids are rare congenital cystic lesions where total excision is often difficult resulting in recurrence. In the current series we tried to analyze and report, the clinical findings, imaging criteria for diagnosis, surgical management strategy to achieve radical resection and outcomes were studied prospectively. Here we report our short experience with intracranial epidermoids as a whole.

**Methods:** 12 cases of intracranial epidermoids confirmed radiologically between Feb 2009 and Dec 2013 were operated by different surgical approaches according to the location of the lesion by micro neurosurgical techniques to achieve total resection. All these patients were followed up for various periods and results analyzed.

**Results:** Of the 12 epidermoids 5 were in CP angle, 4 were in supratentorial location. Of these 2 patients died and Transient facial palsy was observed in two cases and lower cranial nerve palsy in another case which improved later. 2 patients required VP shunt and there were no recurrences in the series. Total resection of the lesion was achieved in majority of the patients (83.3%) by microsurgical techniques.

**Conclusions:** With the availability of good imaging techniques and modern micro neurosurgical techniques it is possible to achieve total or near total excision where ever possible to avoid recurrence of the lesions, resulting in total cure of the patient.

**Keywords:** Epidermoid, Microsurgical management, Recurrence

## INTRODUCTION

Epidermoids are rare congenital benign cystic lesions that can occur anywhere along the entire neuraxis. They arise from displaced dorsal midline ectodermal cell rests between the 3<sup>rd</sup>-5<sup>th</sup> week of embryogenesis during neural tube closure. In the intracranial compartment they are common in Cerebello-Pontine angle and Parasellar regions. There are diverse opinions regarding the extent of excision of these tumours especially when the tumour is large or is adherent to critical neurovascular structures. In the present short series we present here our experience in treating these lesions.

## METHODS

Between Feb 2009 and Dec 2013, 12 cases of intracranial epidermoids were diagnosed pre-operatively with CT and MRI of the brain at our institution, Osmania Medical College and Hospital. All of them were operated by different surgical approaches according to the location of the lesion by micro neurosurgical techniques to achieve total resection. All pts were followed up postoperatively by clinical examination and neuroimaging. Follow-up period was ranging from 6months to 40months. Average follow-up was 24 months.

In this prospective study total number of patients were 12. There were 4 supratentorial epidermoids, three in the Suprasellar Parasellar location and one in the frontal lobe extending to opposite side under the falx, 5 C-P angle epidermoids, one in the pineal region and two in the fourth ventricle. There were 7 females (58.3%) and 5 male patients (41.6%). The age range was 12 years-65years. There was one recurrent CP angle epidermoid, operated in the same institute seven years ago. Common clinical features were according to the site of location like headache and vomiting. Seizures was the presenting symptom in one case of frontal interhemispheric epidermoid. Suprasellar epidermoids presented with unilateral visual disturbances and majority of CP angle epidermoids presented with signs of cerebellar dysfunction. Sixth nerve palsy was found in two cases of C-P angle epidermoids. Only one patient out of five CP angle epidermoids presented with trigeminal neuralgia. Seizures and behavioral disturbances of long duration was observed in one case of giant C-P angle epidermoid with extension in to supratentorial compartment under the temporal lobe. All the patients were evaluated by CT and MRI Brain. Imaging findings were typically suggestive of epidermoids in all the cases. CT scan showed well defined hypodense lesions, slightly lower than CSF intensity, not showing any contrast enhancement. MRI findings were, well defined, irregular marginated lesions found in the basal regions with extensions along the cisternal spaces at the basal regions. They were hypointense in T1WI and hyperintense in T2WI, showing no enhancement following contrast injection. Diffusion weighted imaging and FLAIR imaging done for few cases to rule out Arachnoid cysts in the CP angle, which showed restriction of water movement. Surgical approach varied according to the location of the tumour. Epidermoids are large, totally avascular pearly white tumours allowing for easy identification of the tumours. Complete tumour excision was possible in majority of the tumours by strict adherence to good microneurosurgical techniques. At places where tumour capsule was tightly

adherent to important neural structures, small bits of capsule was left over deliberately to prevent postoperative morbidity.

## RESULTS

Complete excision was achieved in 9 (75%) patients and in two cases total resection was not possible because of poor access, one in the C-P angle and the other in the pineal region. Both were very large tumours with extension in to multiple compartments. In one patient of CP angle epidermoid there was some difficulty in total excision on table towards the brain stem side. Contents of the tumour were pearly white flakes in all the cases with thin capsule adherent to the underlying brain but could be separable by gently teasing away from the brain with microdissectors. Dissection in the C-P angle was more tedious in separating the tumour flakes underneath the neuro vascular structures which were done successfully in majority of the cases with careful microsurgical techniques under high magnification. In two patients small bits of tumour capsule attached to brainstem was left over deliberately to avoid post-op morbidity. Great care was taken by gentle saline lavage of the tumour bed to avoid CSF contamination by cholesterol crystals. Complications included were death in two cases, (16.6%) one giant epidermoid and another in 4<sup>th</sup> ventricular epidermoid because of meningitis in the post-op period 4 weeks later. Transient facial palsy was observed in two cases and lower cranial nerve palsy in another case (25%), both of them improved in 3 weeks' time. Two patients required ventriculo peritoneal shunt for obstructive hydrocephalus in the post-op period. Rest of all other patients improved well and are symptom free till last follow-up. No recurrence was observed in this series so far.

Details of observations and results were given in the Table 1 below:

**Table 1: Clinical details and post-operative outcome of patients in this series.**

Name	Sex/Age	Location	Approach	Extent of removal	Result at follow up
Ashok	M/27 years	4th ventricle	Midline sub - Occipital	Total excision	Improved
Parvathy	F/48 years	4th ventricle	Midline sub - Occipital	Total excision	Died 6 weeks later
Ratnakar	M/52 years	Pineal region	Left posterior parietal and left retrosigmoid	Subtotal excision	Died 8 weeks later
Ravinder	M/28 years	CP-angle	Combined retrosigmoid - Sub temporal	Near total excision	Improved
Siraj	M/43 years	CP-angle	Retrosigmoid	Total excision	Improved
Anusha	F/17 years	Parasellar	Pterional	Total excision	Vision not improved
Santosh	M/18 years	Suprasellar	Pterional	Total excision	Vision not improved
Lalita	F/27 years	Bi-frontal	Right frontal	Total excision	Improved
Ratnabai (Recurrent tumor)	F/55 years	CP-angle	Retrosigmoid	Total excision	Improved
Sandhya	F/10 years	Suprasellar	Pterional	Total excision	Improved
Yadamma	F/45 years	CP-angle	Retrosigmoid	Total excision	Improved
Shilpa	F/18 years	CP - Angle	Retrosigmoid	Total excision	Improved

**Representative case-1 (Table 1, No. 4)**

A 28 year male was referred from psychiatry hospital after treating for behavioral problems and psychosis for 3 yrs. and was not responding to medication, to rule out possible intracranial pathology. There was no h/o headache or vomiting. He had h/o recurrent seizure episodes for the past 3 months and started developing difficulty in walking with tendency to falls now and then. Clinically patient was conscious but incoherent at times. There was no papilledema. He had mild facial asymmetry on right side. No other cranial nerve palsies. He had cerebellar signs on right side in the form of dysmetria and incoordination. There was a tendency to fall on to right side. CT scan brain showed a large, irregular, well defined, hypodense mass lesion in the right Cerebello Pontine angle extending in to the supratentorial compartment through tentorial incisura in to the middle cranial fossa and under the right temporal lobe. There was severe brainstem compression from right side. There was no evidence of hydrocephalus. The lesion did not show any contrast enhancement. MRI brain showed hypointense lesion on T1WI and hyperintense lesion on T2WI (Figure 1).



**Figure 1: T2 weighted image showing right CP angle epidermoid.**

DWI showed restricted diffusion in to the lesion. Since the lesion was occupying both posterior fossa and middle cranial fossa, he was operated by a combined approach of retrosigmoid craniectomy and by sub-temporal approach. The cyst contained pearly white cholesterol flakes, totally avascular, extending through basal crevices ventral to the brainstem, through tentorial incisura in to the middle cranial fossa up to sylvian fissure. Near total excision of the cyst along with capsule was performed under high magnification with microsurgical techniques. Small

pieces of the tumour ventral to midbrain were adherent and not accessible were left over deliberately to avoid post-operative morbidity. Histopathological examination of specimen confirmed cyst wall lined by stratified squamous epithelium surrounded by fibrocollagenous tissue and containing abundant keratin material (Figure 2). Post-operatively he developed transient lower cranial nerve palsies and pseudomeningocele which resolved in three weeks' time. Repeat CT scan confirmed total excision showing only the residual dead space in the tumour bed. (Figure 3) subsequently one month later patient was again admitted for seizures and recurrent headache. CT scan at this stage showed gross hydrocephalus and a v-p shunt was placed and he improved very well and he is self-ambulant and no behavioral problems at last follow-up.



**Figure 2: HPE with eosin and hematoxylin showing features of epidermoid.**



**Figure 3: CT brain showing post-operative changes following excision of epidermoid.**

**Representative case-2 (Table 1, No. 3)**

A 62 year old male was brought with complaints of behavioral abnormalities and difficulty in walking following trivial head injury 1m ago. There was no h/o vomiting but one episode of generalized tonic clonic seizure the day before his admission. Clinically he was conscious, incoherent at times, speaking inappropriate words .cranial examination was normal and there were no sensory or motor abnormalities. He had left sided cerebellar signs and gait ataxia. CT scan brain showed large irregular hypodense lesion found in the left trigone region extending in to pineal region and left C-P angle. The lesion did not show any contrast enhancement and there was obstructive hydrocephalus. MRI brain showed the same lesion as hypointense on T1 and hyperintense on T2WI, similar to CSF intensity (Figure 4). pre-operative diagnosis of epidermoid was done and he was operated by parieto-occipital craniotomy and left trigonal approach as most of the lesion was in the trigone. The tumour contained pearly white soft flakes which were slowly evacuated easily. We tried to excise the thin capsule outside the ventricle and thorough saline irrigation given at the end of the procedure and at a second sitting he was again operated after 3days by retromastoid suboccipital craniectomy and tumour excision. This time entire tumour in the C-P angle excised and part of the tumour going through the tentorial incisura in the pineal region also excised. Small portions of the tumour which was leftover because of non-accessibility and non-visualization. Post-operatively he made good recovery and improved in his gait. 3 weeks later he was re admitted with seizures and symptoms of meningitis. He has developed gross hydrocephalus. He was managed conservatively by external ventricular drainage and antibiotics. He responded well hence EVD was converted to regular v-p shunt. Subsequently, He slowly deteriorated and died of aseptic meningitis after 20 days.



**Figure 4: T 2 weighted images showing left parietooccipital epidermoid.**

**DISCUSSION**

Epidermoids are benign developmental cysts, rarely found in the intracranial compartment with a frequency of

less than 1% of all intracranial tumours.<sup>1</sup> They develop from retained ectodermal cell rests during the 3<sup>rd</sup>-5<sup>th</sup> week of fetal life before neural tube closure.<sup>2</sup> They are intradural and can be found anywhere in the cranial cavity including vault .Majority of the times they are located in C-P angle about 60%, followed by 4<sup>th</sup> ventricle and suprasellar regions.<sup>3-5</sup> They have an irregular cauliflower like outer surface<sup>5</sup> and contain pearly white keratin material laden with cholesterol crystals hence the name pearl tumour. They are commonly found at the bases and tend to grow into the available cisternal spaces encasing all the neurovascular structures in doing so, and are tightly adherent to underlying structures at some places posing great difficulty in excising them.<sup>4-6</sup> They are common in the age group of 2<sup>nd</sup> to 4<sup>th</sup> decade and males outnumber females in incidence, but in this series we had more number of female patients. They are very slow growing cysts and present with symptoms very late according to their location and by gradual mass effect. Common presenting symptoms include headache, seizures, trigeminal neuralgias and behavioral problems when found in the supratentorial compartment. Clinical findings include cranial nerve palsies, cerebellar dysfunction and hemi paresis when brainstem is compressed.<sup>6,7</sup> The common differential diagnosis for an epidermoid cyst are arachnoid cysts, dermoids and cystic neoplasm.<sup>6-8</sup> Generally imaging findings are very specific for epidermoids and diagnosis is certain with MRI most of the times. CT scan shows hypodense mass lesion, irregular shaped that does not show contrast enhancement. On MRI they are hypointense in T1WI and hyperintense like CSF in T2WI because of their solid nature. DWI and FLAIR images are helpful in differentiating other cystic lesions.<sup>9</sup> Depending on the signal characters of MRI they are classified as black and white epidermoids. Black epidermoids are hypointense to T1 and hyperintense in T2 weighted sequences and white epidermoids are hyperintense in both T1 and T2 weighted images. Epidermoids does not enhance with contrast and lack surrounding edema. Occasionally hydrocephalus may be found but may not be significant to the extent of requiring a CSF diversion procedure pre-operatively. Surgical treatment is by total excision of the cyst where ever possible and the approach depends on the location and extent of the tumour which can be accurately assessed by MRI pre-operatively.<sup>10,11</sup> The contents of the cyst are totally avascular pearly white solid keratin flakes which are easily removable by gentle dissection from the surrounding neurovascular structures. The capsule is thin and is adherent to brain parenchyma but can be teased out slowly by sharp micro dissection techniques under high magnification.<sup>12</sup> These are tightly packed solid tumours and does not easily give in for internal decompression. Endoscopic assistance is often helpful in assessing the total removal at corner places where microscopic is not accessible. At places where capsule is tightly adherent to brainstem can be left over in order to avoid post-operative morbidity. Coagulation of the residual capsule has been advocated by some authors but is no longer practiced now. Frequent thorough saline irrigation is very

important to prevent CSF contamination, post-operative development of aseptic meningitis and hydrocephalus.<sup>13</sup> In the present series two of our patients developed meningitis in the postoperative period and died. Subtotal resection will lead to recurrence which we have observed in one case. Post-operative result will be good to excellent after a brief period of morbidity if near total excision is achieved especially with small tumours, as was observed in this series. Large size tumours extending to other compartments tend to fare poorly as total excision may not be possible always because of poor access or development of post-operative complications. Nevertheless surgical excision of epidermoid tumours is most often challenging and troublesome to cure because of its insinuating growth into available spaces engulfing the vessels and nerves which make it difficult to radical excision. Prior to the microsurgical era the operative mortality ranged from 22-57%. But now-a-day's surgical mortality and morbidity have come down very much with tremendous improvement in neuro imaging, microsurgical skills and surgical techniques as well as conservative radical approach in intracranial epidermoid excision.<sup>14,15</sup> Partial tumour excision leads to recurrence at a very slow pace. Immediate post-operative CT scan confirm total excision of lesion but the hypodensity in the tumour bed persists for quite a longer period even after the complete removal of the tumour possibly because of the long standing deformation of the neural structures<sup>15</sup>. MRI is helpful in diagnosing early recurrence /residual tumour. Re-operation is indicated again if patient is symptomatic. In our series the follow-up is very small and we did not find any recurrence or symptomatic recurrence in residual tumours.

## CONCLUSION

Intracranial epidermoids are rare benign tumours but have a potential for recurrence if resected sub totally. With the availability good imaging techniques and modern micro neurosurgical techniques it is possible to achieve total or near total excision where ever possible to avoid recurrence of the lesions, resulting in total cure of the patient.

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