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Rare case of interhemispheric epidermoid in relation to corpus callosum: A case report

KEYWORDS: Corpus callosum, Interhemispheric fissure, epidermoid

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ABSTRACT *Intracranial epidermoids are developmental lesions reported in literature as case reports only. Of intracranial epidermoids interhemispheric epidermoids are still rarer lesions and only 34 cases have been reported in literature till date. We report a case of interhemispheric epidermoid in relation to corpus callosum in a 50 year old male who had presented with generalized tonic clonic seizures of twenty years duration. Neuroimaging revealed the lesion in relation to posterior third of corpus callosum. The lesion was completely removed through posterior interhemispheric approach. Post operatively patient's anti epileptic medications have been stopped and he is seizure free for the last 27 months.*

INTRODUCTION

Epidermoid tumors are benign, slow growing congenital lesions and the most common intracranial embryonal tumors.[10] The most common location of epidermoid tumors are cerebellopontine angle, followed by the suprasellar cisterns, other locations include Sylvian fissure, brainstem, intraventricular, pineal regions, intradiploeic space of skull, and spinal cord. [10] Interhemispheric epidermoids are rare, and only 34 cases have been reported. [13, 4] In the series of 15 patients of Bhat DI et al, [4] the oldest patient was 45 years and longest duration of symptomatic patient was of 15 years. We report a case of large epidermoid cyst of anterior and middle interhemispheric fissure who was 50 years old and was symptomatic for over 20 years.

CASE REPORT

A 50 year old male was referred to our center with 20 year history of generalized tonic clonic seizures, 5 years history of headache and 1 year history of memory disturbances. Neurological examination revealed bilateral early papilloedema and no other focal neurological deficits. The patient had continued to have seizures despite being on antiepileptic drugs for the last 15 years. We optimized the patient's antiepileptic drugs and Magnetic Resonance Imaging (MRI) was performed. The lesion was located in the interhemispheric region on MRI. On T1- weighted images (T1 WI) axial (figure 1A) and saggital (figure 1C), lesion was hypointense to gray matter and mildly hyperintense to cerebrospinal Fluid (CSF) (figure 1A, 1C). On T2-weighted images (T2 WI), axial (figure 1B) it appeared uniformly hyperintense to both gray matter and CSF. Post contrast T1 WI Saggital (figure 2A) and coronal (figure 2B) showed only mild enhancement of the capsule. A preoperative diagnosis of epidermoid tumor of the anterior and middle interhemispheric fissure was made. The patient underwent a right frontoparietal craniotomy and near total excision of the tumor [Figure 3A, 3B]. Per operatively we found a large well encapsulated lesion in the anterior and middle interhemispheric region with cystic component containing clear fluid & suckable whitish cheesy material [Figure 3C]. The histopathological diagnosis was a epidermoid tumor. In the post operative period patient had left hemiparesis which gradually improved and patient was discharged on the fourteenth post operative day. Patient has been on regular follow up and is seizure free, ambulant without support and antiepileptic medications have been stopped.

DISCUSSION

Epidermoid tumors are benign congenital lesions and account for 0.5% to 1.8% of all intracranial brain neoplasms. [16, 7] Intracranial epidermoid tumors arise from displaced epithelial tissue between the third and fifth week of gestation during the closure of the neural tube. During in folding, if the surrounding ectoderm (which later forms the cutaneous structures) does not separate from the neural ectoderm completely, nests of these cells may be entrapped along with the neural ectoderm. These nests of cells later grow within the central nervous system (CNS), resulting in the formation of a spectrum of lesions, viz., epidermoids, dermoids and dermal sinuses. [7, 12, 2] Epidermoids grow slowly and become symptomatic during the third to fifth decades of life (17 to 45 years, in our series), unlike dermoids, which are faster growing and which become symptomatic in the second decade. [12] The epidermoids have a growth pattern similar to that of the skin. Like skin cells, these epidermoid cells also desquamate but into a closed cavity. Slowly as the keratin, cholesterol and desquamated cells accumulate; the lesion grows and becomes symptomatic either due to the mass or rupture. [10, 2, 3]

Epidermoids are mostly cerebellopontine angle in location followed by the suprasellar cistern, the other sites being the Sylvian fissure, brainstem, pineal region, petrous apex, intra-fourth ventricular.[10, 12, 14, 6, 5] Interhemispheric location is rare, and only 19 cases have been reported. [13, 11] These lesions become symptomatic due to either the pressure effects on the surrounding neural elements or irritation of the nerves or cortex, presenting with ataxia, nystagmus, hemiparesis, hydrocephalus, neuralgia or seizures. Rupture of these cysts spontaneously or spillage of contents during surgery can cause aseptic chemical meningitis. [14, 5, 17] In our case patient had presented with episodes of generalized tonic clonic epilepsy.

Radiologically, epidermoids may be seen as extra-axial lesions in the basal cisterns growing along the CSF spaces encasing vessels and nerves and causing a disproportion-

Volume : 5 | Issue : 1 | Jan 2015 | ISSN - 2249-555X

ately less distortion of the surrounding brain. On CT scan, the lesions appear as hypodense lesions with attenuation similar to or lower than that of CSF. There is usually no enhancement on contrast. [14, 9] On T1-weighted images, epidermoid appears hypointense to the gray matter and slightly hyperintense to CSF. On T2-weighted images, the lesion is hyperintense to gray matter and similar to that of CSF. However, heterogeneity of the signals is seen commonly. [8] Diffusion-weighted images show restricted diffusion making the lesion hyperintense. CSF and arachnoid cysts appear hypointense as there is no restricted diffusion [15]. The radiological features were similar in our case.

The interhemispheric location of the lesion and its proximity to the anterior cerebral arteries, its branches, motor cortex and supplementary motor cortex make surgical excision technically challenging. Preservation of the bridging veins must be given utmost priority. Despite this, safe radical excision can be achieved in most of these cases. In our case complete excision could be achieved with preservation of adjacent vital structures. Chemical meningitis is a well-known complication after epidermoid surgery. [14, 17] Up to 40% incidence of chemical meningitis following surgery has been reported. [5] In our case patient recovered well didn't had chemical meningitis following surgery.

These slow-growing epidermoids are known to recur during the long-term, especially when partially removed. However, as the risk of recurrence is low for small residues, it is better to try to achieve total excision safely and when not possible leave behind small residues attached to vital structures. [14, 17] Because of the slow growth of the tumor, the chance of symptomatic recurrence during one's lifetime following total excision may be negligible or practically absent. [14, 1]

In our case patient has been under regular follow up for last 27 months, and there has been no recurrence as of now. Patient has also become seizure free. Patient has no fresh deficits as compared to pre-operative status.

Figures and legends to Figure:

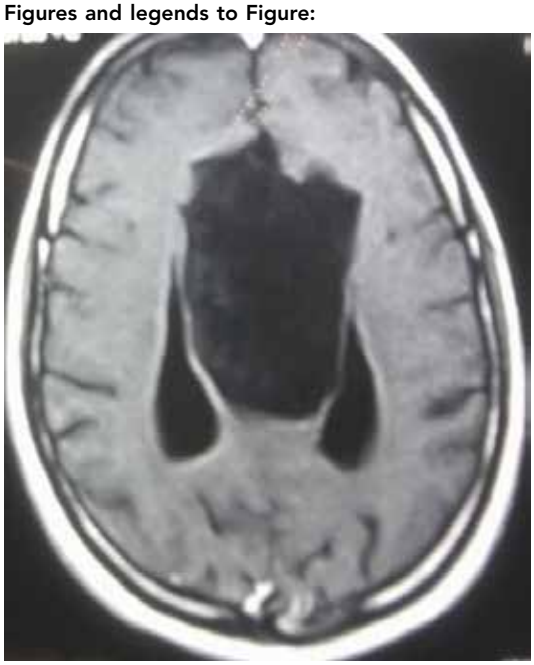

Figure 1A: T1 weighted axial image showing a large hypointense lesion in interhemispheric region.

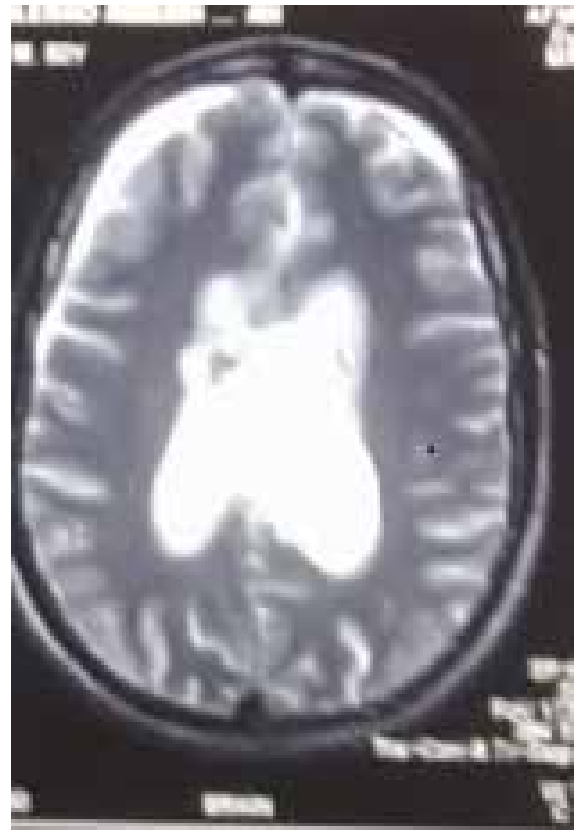

Figure 1B: T2 weighted axial MRI image showing hyperintense lesion in interhemispheric region

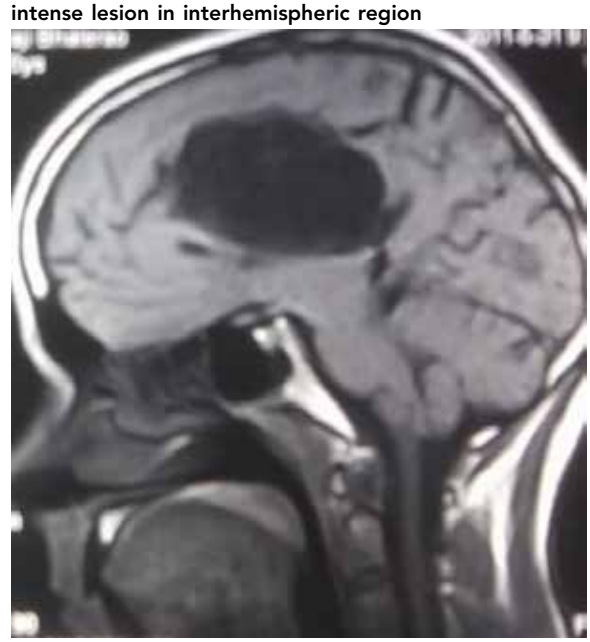

Figure 1C: T1 saggital image showing hypointense lesion compressing the corpus callosum

Volume : 5 | Issue : 1 | Jan 2015 | ISSN - 2249-555X

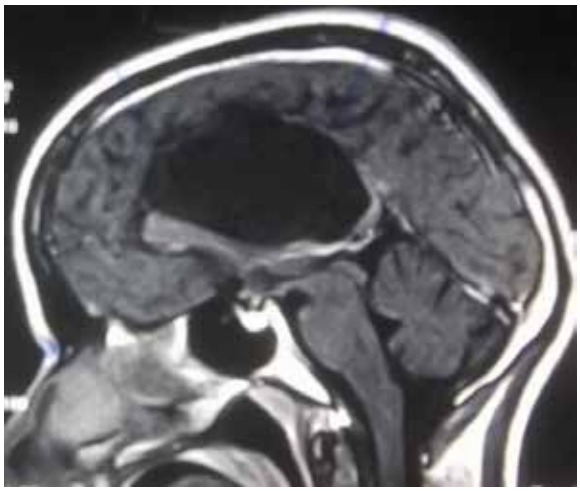

Figure 2A: T1 –weighted saggital contrast image showing only mild enhancement of the capsule.

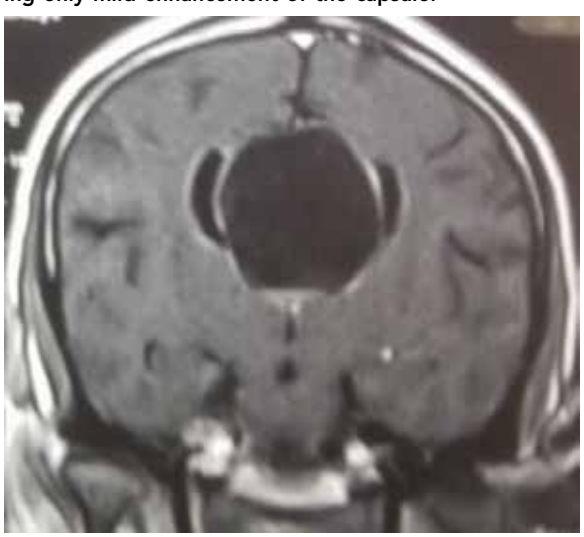

Figure 2B: T1 – weighted coronal contrast image – showing capsule enhancement

Figure 3A: Intraoperative image showing planned right parasaggital craniotomy

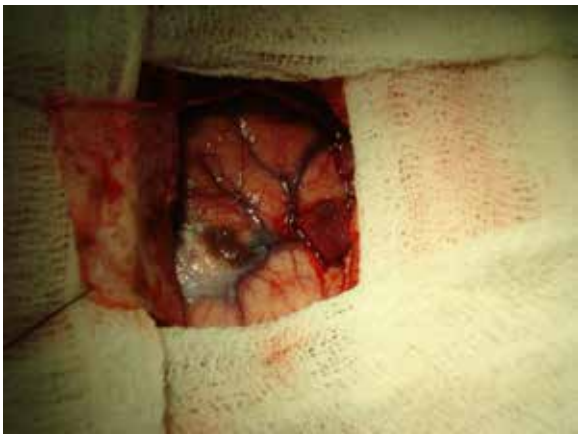

Figure 3B: Intraoperative image after durotomy revealing evidence of the lesion over right parasaggital and interhemispheric region.

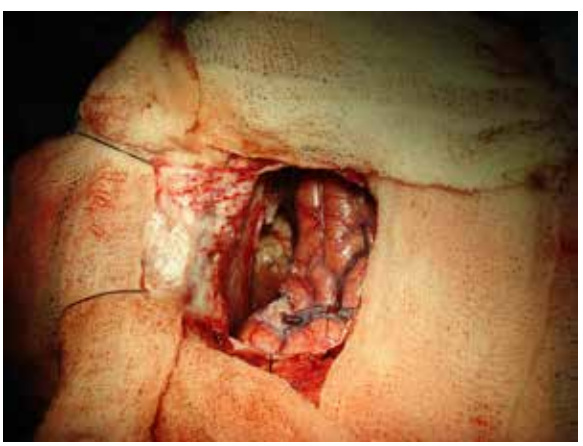

Figure 3C: Intraoperative image showing white cheesy material suggestive of epidermoid during excision of tumor.

Volume : 5 | Issue : 1 | Jan 2015 | ISSN - 2249-555X

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