

CASE REPORT

Rare occurrence of intracranial epidermoid tumor in an unusual location with atypical radiological features

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ABSTRACT

Intracranial epidermoid tumors are benign rare tumors that arise due to dysembryogenesis. Though rare, they have characteristic radiological features such as their magnetic resonance imaging signal intensities and a typical location. We present a rare case of intracranial epidermoid tumor in a young female patient that was unusually located within the left frontal lobe and had certain atypical radiological findings. Preoperatively, this lesion was erroneously presumed to be a low grade glioma or a dermoid. Therefore, this case report prompts us to include atypical epidermoid cyst in the list of differential diagnosis for nonenhancing lobar tumors.

Key words: Atypical, epidermoid, frontal lobe

Introduction

Intracranial epidermoid tumors are rare tumors. They constitute 1% of all the intracranial tumors.^[1] Commonly, they are located within the vicinity of the cisternal spaces with characteristic imaging features.^[2] However, rarely, they may occur in an unusual location with atypical radiological features. Such unusual variants have been reported to account for 5-6% of all epidermoid tumors.^[1,3,4] In addition, nearly half of such atypical epidermoid tumors are misdiagnosed as some other pathological entity preoperatively. We present an unusual and rare case of an epidermoid tumor in the left frontal lobe. Preoperatively, this lesion was presumed to be a low-grade glioma or a dermoid.

Case Report

A 20-year-old female patient presented with one episode of generalized tonic clonic seizure 1-month ago. Since then,

she had been having nonspecific headache not suggestive of raised intracranial pressure. The fundus examination was normal, and there were no focal deficits. Magnetic resonance imaging (MRI) showed a large nonenhancing mass in the left frontal region that was seen surfacing. There was mass effect on ipsilateral frontal horn of the lateral ventricle. The brain tumor interface was good with no signal changes within the adjacent brain. The lesion was densely hypo to isointense on T1-weighted and homogenously hypertense on T2-weighted imaging [Figure 1]. Fluid attenuation inversion recovery imaging showed hyperintense signal changes within the lesion and diffusion weighted imaging showed restricted diffusion [Figure 2]. Spectroscopic examination was not done. Preoperative diagnosis of low-grade glioma or dermoid tumor was considered. Intraoperatively, the dura was normal. Beneath it there was an encapsulated, yellowish, flaky, avascular tumor [Figure 3]. The tumor was capsulated, and it was adherent in a certain location to the underlying pia. However, the entire lesion could be removed without gross violation of pial margins [Figure 4]. Histopathological examination showed a cystic lesion lined by keratinized stratified squamous epithelium containing lamellated keratinous debris suggestive of an epidermoid cyst [Figure 5]. The patient made an uneventful postoperative recovery and stays asymptomatic until the last follow-up.

Discussion

Epidermoid tumors arise from the ectodermal elements and are slow growing benign tumors. They are rare tumors accounting for 1% of all intracranial tumors.^[1,4] Vast majority of them share certain typical characterizing features such as their location

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DOI:

10.4103/1793-5482.162718

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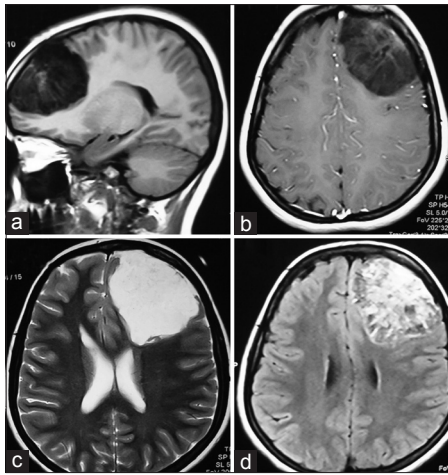


Figure 1: Magnetic resonance imaging brain (a) T1-weighted sagittal section showing predominantly hypointense surfacing lesion with mass effect. (b) Gadolinium axial image showing no significant enhancement pattern. (c) T2-weighted axial image shows homogenous hypointensity. (d) Fluid attenuation inversion recovery axial image reveals hyperintensity within the lesion



Figure 2: Diffusion weighted imaging shows restricted pattern

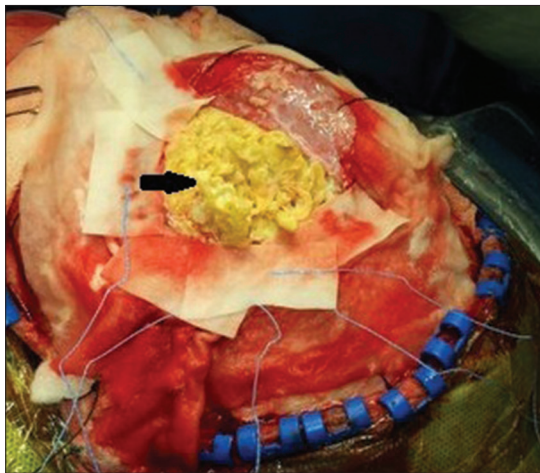


Figure 3: Intra operative photograph showing tumor (black arrow) consisting of typical yellowish, cheesy, flaky material under the dura

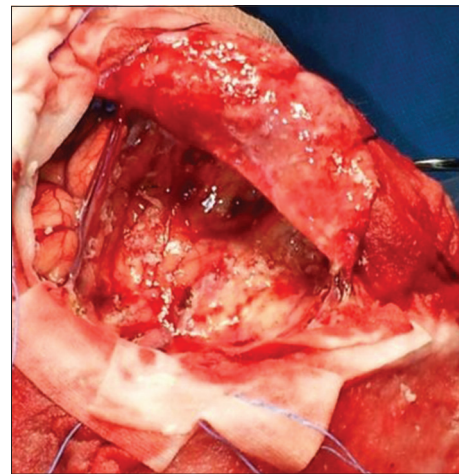


Figure 4: Tumor bed after resection

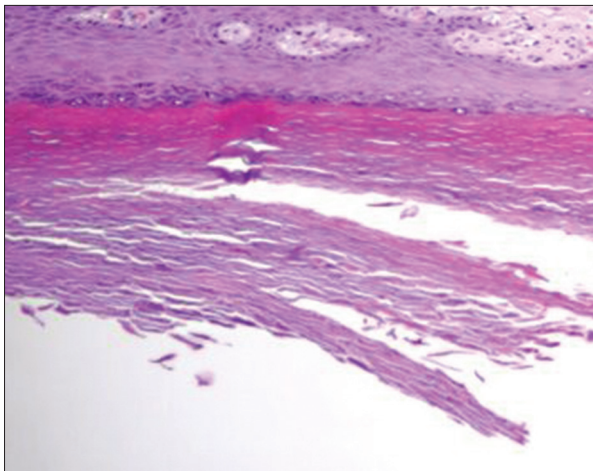


Figure 5: Hematoxylin and eosin sections staining of the keratinized stratified squamous epithelium containing lamellated keratinous debris

and MRI signal intensities.^[2] They are known to arise in the vicinity of a cisternal space, often in the cerebellopontine angle, suprasellar and ambient cisterns.^[2,3] It is very rare for an epidermoid to be present within the parenchyma of the brain. Second, one of the important radiological finding of a typical epidermoid is homogeneously long T1-weighted and T2-weighted signals.^[1] The T1-weighted, proton density, and T2-weighted sequences generally demonstrate heterogeneous signal intensity that is hyperintense to cerebrospinal fluid (CSF).^[1] However, in exceptional situations like in our case, variations have been reported.^[1,3,4]

Hu *et al.* reported series of intraparenchymal epidermoid tumors in six patients, out of which only one (Patient 2) was located in the frontal region.^[1] This tumor was a giant tumor with extension into the temporal lobe as well with mixed intensities on MRI. Ren *et al.* in a large series of 404 patients with epidermoid cyst reported only 24 cases with unusual signals (5.6%).^[4] The tumors with such anomalous signals were defined as “atypical.” Out of this only two patients (patient 3 and patient 18) had tumors that were located in the

Table 1: Characteristics of atypical frontal intraparenchymal epidermoid tumors as described in the literature

Study and year	Number of cases	Intraoperative findings			MRI findings				Pathology		Preoperative diagnosis
		Adhesion	Extent of resection	Contents	T1	T2	DWI	Gado	Granulation	Hemorrhage	
Hu <i>et al.</i> 2008	01	Nil	Total	Whitish yellow	Mix	Mix	Restricted	Nil	Nil	Nil	Nil
Lian <i>et al.</i> , 2012	01	Nil	Total	Tough, rubbery	Mix	Hypo	Not restricted	Nil	Nil	Nil	Oligodendroglioma
Ren <i>et al.</i> , 2012	02	Patient 3: Yes, Patient 18: No	Total	Yellow, cheesy	Mix	Hyper	Not reported	Faint	Patient 3: No, patient 18: Yes	Yes	Patient 3: Teratoma, Patient 18: Glioma
Our case	01	Yes	Total	Yellow cheesy	Hypo	Hyper	Restricted	Faint	Nil	Nil	Dermoid/glioma

MRI - Magnetic resonance imaging; DWI - Diffusion-weighted imaging

frontal lobe. Recently, Lian *et al.* have reported a similar case of epidermoid tumor within the frontal lobe having atypical MR findings.^[3] This illustrates the rarity of epidermoid tumor to be located intra axially within the frontal lobe, as seen in our case.

On MRI, there were certain unusual features associated with the tumor in our case. Apart from the unusual location, the lesion showed mixed signal intensities with visually striking hypo intensity on T1-weighted images that were indeed very peculiar. It was iso to mildly hyperintense with respect to CSF. None of the atypical epidermoid tumors in Ren's study showed gross hypo intensity on T1-weighted as witnessed in our case. Various characteristic features of these atypical pure intraparenchymal frontal epidermoid tumors have been summarized in the Table 1.

Hence, a preoperative assumption of a low-grade glioma or a dermoid tumor for an epidermoid tumor was made. Interestingly, appearance of the lesion was so lucrative to be erroneously termed as a meningioma by some, even though; the classical radiological features of a meningioma were absent.

Therefore, the combination of unusual MRI signal findings and an extremely rare location of this tumor make this report a unique case. The literature is sparse in reporting of unusual epidermoid tumors with handful publications in this regard.

Though, the reasons are unclear; it would be really interesting to know the cause for such an embryonal tumor arising within the parenchyma of the brain and the basis for MR signal variations. Emerging imaging modalities such as spectroscopy

may help in the future to further characterize the tumor resulting in accurate preoperative diagnosis. Further studies and reports would also be important to assess the clinical behavior of such atypical epidermoid tumors.

Conclusions

In the list of differential diagnosis for nonenhancing lobar tumors with atypical MR signals that are predominantly hypo intense on T1-weighted images, "atypical" epidermoid tumor should be considered along with other lesions such as astrocytomas, arachnoid cysts, dermoid cysts, and cavernomas. Long-term studies are required to assess the clinical behavior of such lesions.

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How to cite this article: Sarkar H, Vijaya G, Mitta S. Rare occurrence of intracranial epidermoid tumor in an! unusual location with atypical radiological features. *Asian J Neurosurg* 2015;10:319-21.

Source of Support: Nil, **Conflict of Interest:** None declared.