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Case Report

An atypical case of giant intradiploic epidermoid tumor

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ABSTRACT

Intradiploic epidermoid tumors are uncommon and giant epidermoid with dural involvement is scarcer. We report a unique case of a giant frontal epidermoid tumor presenting without typical features of swelling or bulge in scalp. A 61-year-old male presented with the complaints of forgetfulness and headache. Contrast magnetic resonance imaging brain revealed a large left frontal epidermoid tumor. A tumor measuring 13 × 11 × 4 cm, involving the dura but sparing the brain parenchyma, was excised through left frontal craniotomy. Such a presentation of giant epidermoid tumor with dural involvement is highly unusual. Complete surgical excision is the final aim and vigilant follow-up for recurrence is a must.

Keywords: Diploic, epidermoid, frontal, intracranial

Introduction

Epidermoid tumors are benign in nature and can be intracranial or spinal. They can be intradural or extradural and occur usually near the cerebellopontine or para sellar cisterns.[1-3] The intradiploic epidermoids are rarely reported in the literature and the stated frequency of involvement is 46% in both tables, 31% in outer table, 10% in both tables and dura, 7% in inner table, 3% in inner table and dura and 3% in inner table, and dura and brain.[4]

We present a unique case report of a 61-year-old male patient with an unusual presentation of giant epidermoid tumor of intradiploic origin with dural involvement.

Case Report

A 61-year-old male presented with the complaints of off and on forgetfulness for three months and left sided headache for 15 days. He was conscious and fully oriented with a GCS of E4V5M6. No focal neurological deficit was observed and there was no similar past history. Contrast-enhanced magnetic resonance imaging (MRI) of brain was suggestive of the left frontal space occupying lesion, features of which were consistent with epidermoid tumor [Figures 1 and 2] (Extra-axial lesion hypointense on T1W image and hyperintense on T2W and ADCC image showing diffusion restriction).

Tumor was approached by performing left frontal craniotomy. The tumor was found to be enveloped with a thin capsule and originating from frontal diploe. It perforated the dura and compressed the underlying brain, thereby displacing the superior sagittal sinus toward right. Pearly white in appearance, the tumor was, avascular, non-suckable, firm, and waxy, like cholesterol crystals with a well-defined plane of cleavage. The overlying frontal bone was found eroded with multiple osteolytic lesions [Figure 3]. The tumor was excised, taking care to remove its capsule along-with. To prevent any recurrence, duraplasty was done using pericranium patch and involved bone flap was removed, cranioplasty was undertaken using titanium mesh. Subdural and a subgaleal drains were used. Postoperatively, the patient recovered well and had no neurological deficits. Drains were removed on the 2nd post-operative day. On the 3rd post-operative day, the patient was discharged on antibiotics and analgesics. The patient has been on regular follow-up for the 14 months. The patient has had no significant complaints. Brain MRI done at 6 months of follow-up showed no recurrence.

Histopathology

Tumor tissue, grossly included multiple gray white and pearly white soft tissues measuring (13 × 11 × 4) cm and bone fragment/flap, included single hard tissue measuring (7 × 6 × 4) cm. On microscopic examination, sections showed wall of a cyst lined by stratified squamous epithelium, keratinous material, and cholesterol crystals. No features of malignancy were seen. The bone fragment/flap showed signs of erosion [Figure 4].

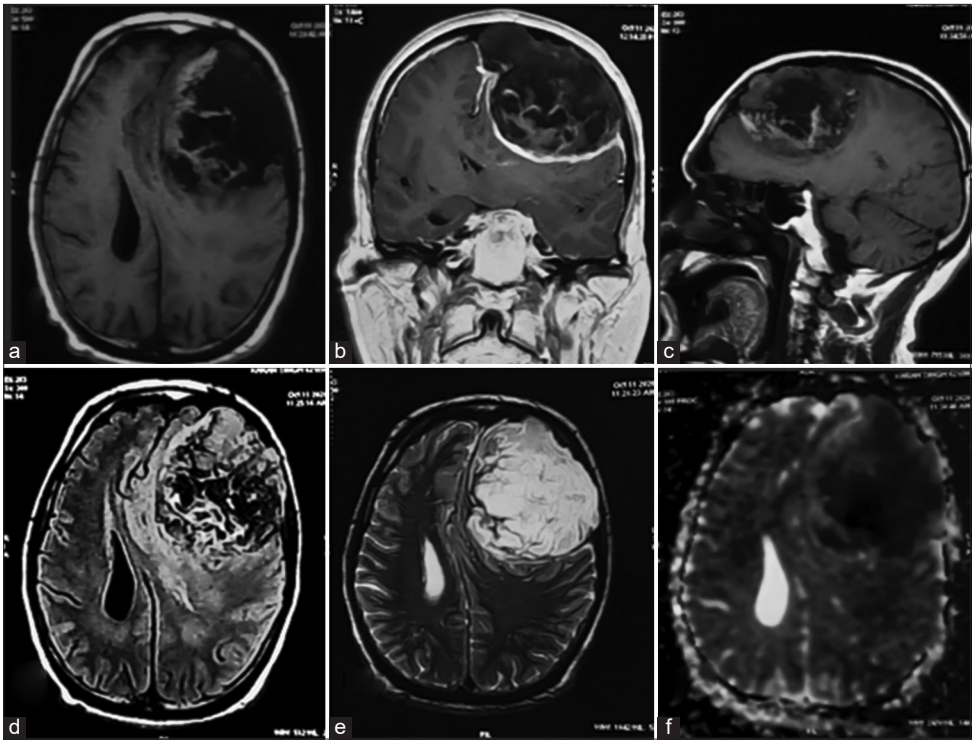

Figure 1: Pre-operative magnetic resonance imaging Axial T1W image showing hypointense extra axial lesion with mass effect on the underlying parenchyma in the left frontal region (a). Coronal T1W contrast image demonstrating the peripheral contrast enhancement (b). Sagittal T1W contrast image illustrating the minimal contrast enhancement with significant mass effect (c). Axial flair image showing minimal peripheral peritumoral edema (d). Axial T2 image illustrating hyperintense extra-axial lesion in frontal region with mass effect (e). ADCC image showing diffusion restriction, characteristic of epidermoid (f)

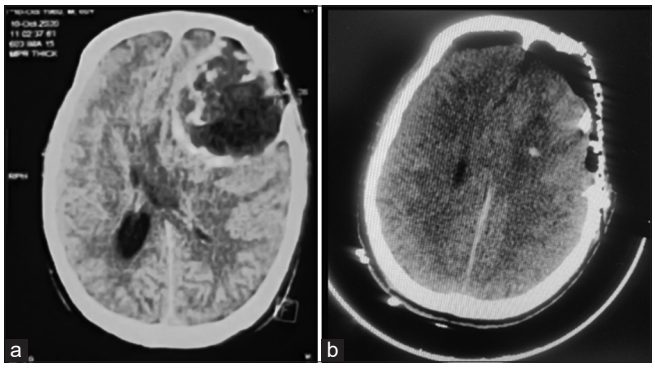

Figure 2: NCCT demonstrates a mixed density lesion in the left frontal region causing compression of the underlying parenchyma (a). Post-operative NCCT brain illustrated no residual tumor and absence of a mass effect, no midline shift, no ventricular compression, cranioplasty done with titanium mesh (b)

Discussion

Epidermoid is slow growing congenital tumors. Cruveilhier, a French pathologist regarded them as the "most beautiful tumors of all the tumors" based on their pearly appearance.[5] They grow at a rate similar to the epidermal cells of skin, multiplying along the cisternal spaces barring a few of those extending into the parenchyma.[6] These tumors are known to occur through ectopic inclusions of epithelial cells at the time of the closure the neural tube.[7] On the other hand, Dias and Walker considered gastrulation dysembryogenesis to be the offending event.[8]

Typical computed tomography appearance is that of a homogeneously non-enhancing hypodense mass in the subarachnoid space sans peritumoral edema. At times, epidermoid tumors present as significantly more dense lesions (known as "white epidermoids"), hence confounding the diagnosis.[9,10] MRI appearance includes a spectrum of appearances, varying from hypointense to hyperintense. Multiloculated appearance is quite common. More commonly, the tumor is heterogeneous and hypointense on T1-weighted images and hyper intense on T2-weighted images.[10] On histopathological examination, the tumor capsule is typically thin, consisting of stratified, keratinized squamous epithelium. Also accumulation of desquamated epithelial cells were seen with cholesterol and keratin. Epidermoid tumors may rarely give rise to squamous cell carcinoma.[11]

Epidermoid tumor may occur anywhere in the neuroaxis, more commonly in the cerebellopontine angles (40–50% of the cases) and the parasellar region.[10] Atypical locations, like

intra-axial, constitute <1.5% of all intracranial epidermoid lesions[12] and intradiploic epidermoid tumor consist of <3% of such tumors.[13] Among the intraparenchymal epidermoid tumors, most occur in the frontal and temporal lobes.[12] Such tumors are scarce in the pineal gland[14] or the brainstem.[15]

Intradiploic epidermoid tumors have been mentioned in only as case reports or case series.[7,16-18] A PubMed central search done by the keywords "frontal intradiploic epidermoid cyst," produced 27 results including case reports [Table 1], case series, and review studies [Table 2].[19-41].

Table 1: Review of case reports

S. No.	Country	Year	Gender	Age	Location	C/F	Size (cm)	Dura involvement	Remarks
1.	Brazil	2019	M	23	Frontal (L)	Proptosis diplopia	N/A	No	Only intradiploic, extending to orbit
2.	India	2019	F	42	Frontal (L)	Headache , seizures	2.4×3	No	History of meningioma surgery. Initial diagnosis? mets. FDG – PET done
3.	India	2018	F	14	Frontal (midline)	Pain, Swelling	5.1×5.2	No	Extending from frontal sinus to ACF
4.	India	2018	F	46Y	Occipital (R)	Headache, swelling	4×7 × 6.7	No	Giant epidermoid
5.	China	2018	M	54	Frontal (L)	Headache, confusion	N/A	No	Concurrent chronic epidural hematoma
6.	USA	2016	F	47	Frontal (L)	Seizure	2.5×3	No	Post traumatic
7.	Mexico	2015	M M	42 46	Occipital Frontal	Headache Delirium with ICH	N/A	No	Intracranial hypertension syndrome+
8.	Turkey	2014	F	14 m	Frontal	Asymptomatic	N/A	No	Craniosynostosis +
9.	India	2013	F	24	Frontal (L) + Orbital	Headache, diplopia, swelling, ocular movements decreased	N/A	No	Post traumatic
10.	Turkey	2013	M	69	Frontal	Bulge, headache	8×5	No	–
11.	Germany	2012	M	81	Frontal + Temporal + parietal	Swelling	15×12×10	No	Only biopsy done, surgery refused by patient
12.	Turkey	2011	M	4	Frontal + orbital (L)	Ulcer of left eyelid	N/A	No	Fistulisation of eyelid
13.	USA	2010	M	69	Frontal (L)	Headache , diplopia	1.8×2.8×4.2	No	–
14.	South Korea	2006	M	69	Frontoparietal (L)	Swelling , mass, seizure	N/A	Yes	Parenchymal invasion +
15.	Italy	2005	M	23	Frontal- Midline	Swelling	N/A	No	Post traumatic
16.	Italy	2002	M	24	Frontal (R)	Swelling followed by rupture	N/A	No	Traumatic rupture of epidermoid cyst
17.	Spain	2001	F	22	Frontal + sphenoid + orbital	Proptosis	N/A	No	–
18.	Switzerland	1997	F	52	Frontal (R)	Left sided hemiparesis , anisocoria	N/A	No	Traumatic pneumocephalus

Table 2: Case series and review studies

S. No.	Country	Year	Gender	Age	Location	C/F	Size	Dura involvement	Remarks
1.	Italy	2018	M 112 F 122	26.99m ± 32.7	47/237-frontal	N/A	NA	No	2/237 epidermoid. 22/237 intrdiploic 7 partial thickness bone erosion 15 full thickness erosion. (Epidermoid-partial thickness erosion)
2.	USA	2016	Male: 60.5% Female 39.5%	Mean age of presentation was 38.1	30.5% frontal	Swelling, neurological deficits, headache	N/A	30/167– dural involvement +.10/30 were frontal	30% Frontal
3.	Turkey	2004	F F F F	46 19 55 35	Occipital (L) Frontal (R) Frontal (R) Occipital (L)	Headache, dizziness Lump/ swelling Headache Headache	N/A N/A 2.2×2.0×1.2, 1.2×0.7 N/A	No No No No	2 lesions
4.	Spain	1995	M 26 F 11	29.6Y (mean)	13 Parietal 10 Frontal	Swelling	0.7–14 cm (avg 2.9cm)	No	
5.	Netherlands	1991	F F M F	18 23 52 26	Frontal (L) Sphenoid bone Sphenoid Temporal (R)	Proptosis Proptosis Proptosis Proptosis	N/A 3×4 cm N/A N/A	No No No No	Operated 4 times
6.	U.K	1989	M M F	52 29 68	Occipital (L) Frontal Frontal	Headache, vision impairment. Swelling Headache	N/A N/A N/A	Yes Yes Yes	Intradural extension

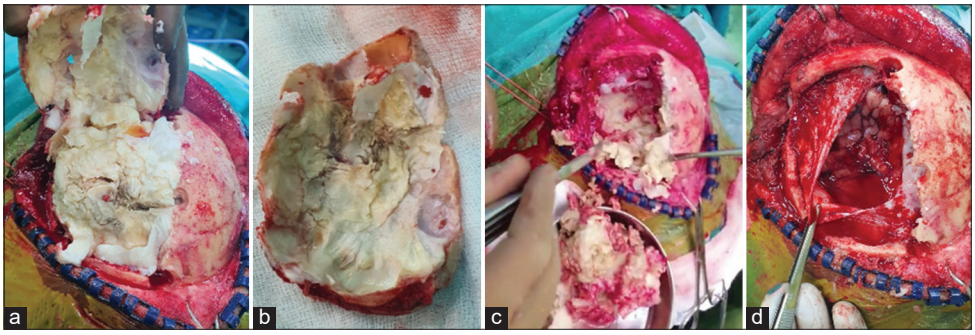

Figure 3: Intraoperative findings-epidermoid with thin capsule, invading the bone (a). Skull bone with areas of erosion (b). Pearly white epidermoid tumor being excised (c). Total excision of the tumor tissue with the involved dura, underlying brain parenchyma is compressed under pressure from tumor (d)

In the cases with lesion of more than 5 cm, swelling is a common presenting feature.[21,28] On the contrary, in our case, a swelling or a bulge in the scalp was not present, which is highly unusual for a tumor of such large dimensions.

Such large tumors have been reported to be adherent to dura, although dural perforation has not been mentioned.[21] In a review done by Arko *et al*. out of 167 tumors, 30 were found to have dural involvement. Ten tumors out of these 30 were found to be in frontal region.[24] Overall, in 434 cases reviewed in the literature, only 34 were seen to involve dura [Tables 1 and 2]. In our case, the dura was observed to be invaded causing extensive perforations at mandating a pericranial patch cranioplasty.

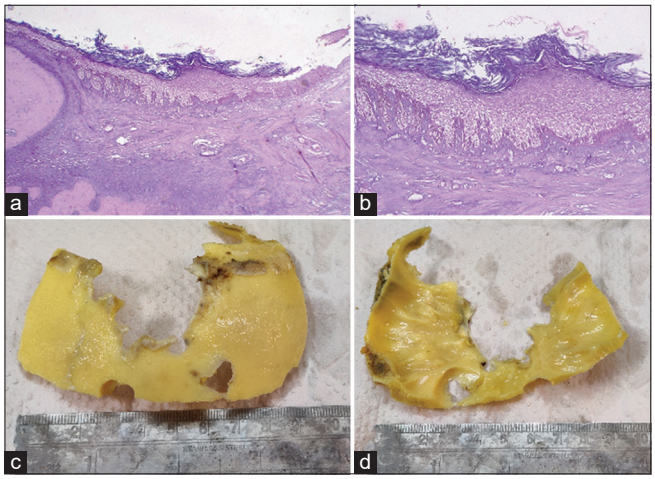

Figure 4: H and E section ×10 magnification (a) and ×40 magnification (b) showing epidermoid inclusion cyst lined by stratified squamous epithelium and cyst is filled with keratin flakes. (c and d) show single hard tissue (bone flap) measuring 7 × 6 × 4 cm

Conclusions

Epidermoid is a congenital tumor occurring in cranial as well as extracranial locations. This tumor usually restricts itself to certain common locations. As exemplified by this case, it needs to be borne in mind that a large intradiploic epidermoid may not present with a typical swelling or a bulge in the scalp. Dural invasion and perforation are needs to be anticipated in such a large sized tumors. In cases of dural involvement, it is imperative that the surgeon excise the involved dura and do duraplasty, preferably autologous. Such a measure may also decrease recurrence. Furthermore, a pre-operative preparation for mesh cranioplasty must be done. Treatment aim should be total surgical excision of the tumor without causing neurological deficit and minimizing the chances of recurrence.

Authors' Declaration Statements

Patients consents

Written informed consent has been taken from the studied patient**.**

Data availability statement

The data used to support the findings of this study are included within the article.

Competing interests

The authors declared no have conflicts interest.

Funding statement

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Authors' contributions

Dr Sanjeev Dua, Dr Anil Dhar – Conceptualization and review; Dr Hershdeep Singh- Writing, reviewing, and preparation of manuscript; Dr Roomba Ambastha- performed histopathology; and Dr Vikrant Katyar, Dr Aditi Shukla – performed data collection.

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