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 $13 \times 11 \times 4$ 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 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 $(13 \times 11 \times 4)$ cm and bone fragment/flap, included single hard tissue measuring $(7 \times 6 \times 4)$ 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].
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. 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. These tumors are known to occur through ectopic inclusions of epithelial cells at the time of the closure the neural tube. On the other hand, Dias and Walker considered gastrulation dysembryogenesis to be the offending event.

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. 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. 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.

Epidermoid tumor may occur anywhere in the neuroaxis, more commonly in the cerebellopontine angles (40–50% of the cases) and the parasellar region. Atypical locations, like
Dua, et al.: An atypical case of giant intradiploic epidermoid tumor

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.\(^{(12,16-18)}\) 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      | 42  | 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  | 26.99m ± 32.7 | 47/237-frontal   | N/A          | NA                 | No               | 2/237 epidermoid. 22/237 intradiploic 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 122  | F 46     | Occipital (L)     | Headache, dizziness | N/A              | N/A              | No                                                                   | 2 lesions |
| 4.     | Spain   | 1995 | M 26   | F 18     | Frontal (L)       | Proptosis     | N/A              | No               | Operated 4 times                                                       |
| 5.     | Netherlands | 1991 | M 23   | F 52     | Sphenoid bone     | Proptosis     | N/A              | No               | Intradural extension                                                  |
| 6.     | U.K     | 1989 | M 52   | M 29     | Occipital (L)     | Headache, vision impairment. | N/A              | Yes              | Intradural extension                                                  |

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 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)
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’ contributions

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

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Dua, et al.: An atypical case of giant intradiploic epidermoid tumor

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