
Case report

Intradiploic epidermoid cyst - a pearl within the skull

V Lakshmi¹, KS Thirumurthy², S Rabia¹, V Chaitra¹

Introduction

Epidermoid cysts are quite uncommon in the craniofacial region and more so in the intradiploic region within the skull. They are benign and slow-growing. Some attain great size, producing major neurological signs. A high index of suspicion with an accurate radiological assessment followed by complete removal of the tumour and its capsule are essential for good long-term prognosis. We report an interesting case of an epidermoid cyst occurring in the skull of a young male patient with a short review of the relevant literature.

Case report

A 31-year-old male patient presented to the outpatient unit of the neurosurgery department with complaints of giddiness. The symptoms were present intermittently and no relevant past medical history or family history could be elicited.

On examination, the patient had a GCS (Glasgow Coma Scale) of 15/15 and his vital signs were stable. Computed tomographic scan (CT Scan) showed an intradiploic expansile cystic lesion with a sclerotic rim in the left temporo-parietal region. The patient underwent craniotomy for excision of the cyst. A small piece of the lesion was sent for frozen section study, which showed anucleate squames and keratin debris. The cyst was excised and the residual bone defect was covered with a titanium mesh, fixed with titanium screws. Bone cement was applied to solidify. Post-operatively, the patient remained stable. A course of antibiotics and anticonvulsants were prescribed.

The specimen was sent to the histopathology laboratory in 10% neutral buffered formalin. An elliptical piece of bone with the inner surface showing a fleshy mass was received. Cut surface of the growth appeared friable with gritty areas. (Fig. 1)

¹Department of Pathology, ²Department of Neurosurgery, PSG Institute of Medical Sciences and Research, Coimbatore, India.

Correspondence: Vidhya Lakshmi, Department of Pathology, PSG Institute of Medical Sciences and Research, Coimbatore - 641004, India. Email: drvidhyalakshmi@yahoo.co.in

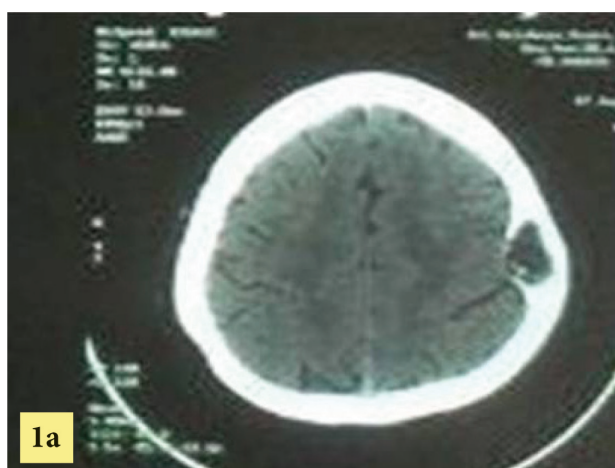


Fig. 1. 1a. Computed tomographic (CT) scan showing an intradiploic expansile cystic lesion with a sclerotic rim in the left temporo-parietal region. 1b. Complete excision of cyst with cranioplasty. 1c. Gross image of inner surface of skull showing a fleshy mass causing ulceration of the dura.

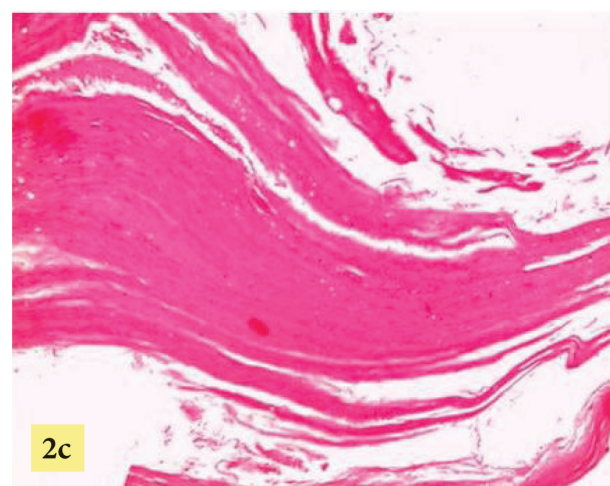
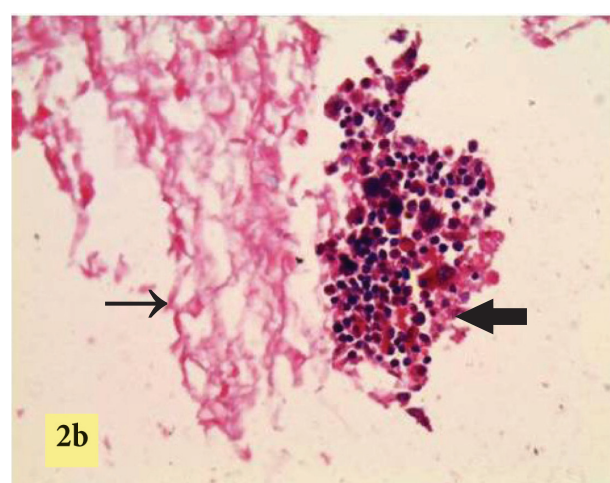
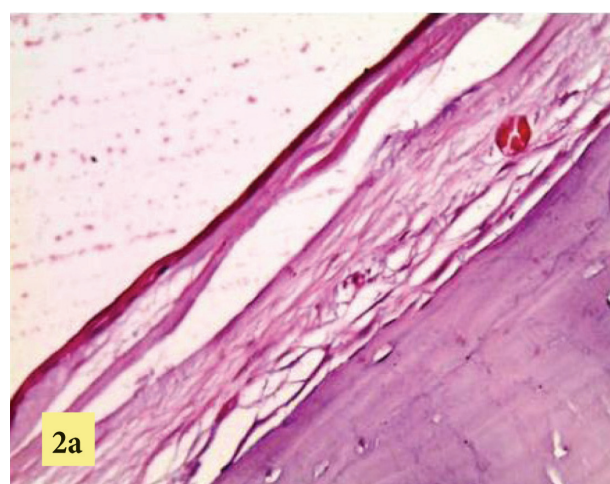


Fig.2. 2a Degenerated bony spicules with a cyst lined by stratified squamous epithelium H&Ex400. 2b. Anucleate squames (arrow) and hematopoietic elements (block arrow). 2c. Keratin flakes; H&E x400.

On histological examination, degenerated fragments of bone were seen with a lesion composed of a lining stratified squamous epithelium overlying lamellated flakes of keratin, keratinous debris and occasional anucleate squames. (Fig.2) Other adenexal structures were not seen. On follow up the patient showed a significant improvement of his symptoms.

Discussion

Epidermoid cysts are common in routine histological practice and they are usually located in hair-bearing areas of the skin. Location within the cranial cavity are rare. They represent 0.04 to 0.7% of intracranial neoplasms (1). Intradiploic dermoid cyst was first described by Cushing in 1922.

During neural tube formation, sequestration of ectodermal elements can be seen within the cranial cavity due to defective separation of the neuroectoderm (2). This results in the formation of epidermoid cysts which can remain asymptomatic even after attaining a large size. The presence of cheesy, keratinous debris within the cyst lumina imparts a “pearly” appearance and hence these tumours have also been referred to as “pearly tumours” in literature (3). The intradiploic location is far less common than the intradural location (4).

CT imaging (5) is of greater value than plain radiography as CT allows better

assessment of both skull involvement as well as intracranial extension. Intraparenchymal extension of intradiploic epidermoids is known to occur. Imaging appearance depends on the ratio of keratin to cholesterol in the lesion. Typically epidermoids are hypodense on CT.

Although they remain asymptomatic for long periods, erosion of the bone and involvement of the brain parenchyma can occur. Rupture of the cyst with release of contents can occur into the subarachnoid space. Chronic granulomatous arachnoiditis can result from irritation of the meninges due to released keratin.

Epidermoid cysts are mostly seen in the third and fourth decades of life with a male predominance. The lesions may extend and present as a subcutaneous swelling of the scalp. In contrast, dermoid cysts are typically associated with the suture lines, commonly occur in orbital region and in the midline and usually present in childhood. (6). Histology of an epidermoid cysts shows a keratin-filled cyst lined by stratified squamous epithelium (7). The cyst wall characteristically lacks dermal elements.

Other differential diagnoses of epidermoid intradiploic cysts include bone-destroying lesions affecting the orbit, eosinophilic granulomas, giant cell granulomas, cholesterol granulomas, cavernous hemangioma, and osteolytic intradiploic metastases (8,9).

Giant extradural epidermoids with profound deformation of the brain and extensive lytic skull lesions may allow a normal life without any significant restrictions (10). Individual risk-benefit assesment is mandatory in the discussion of a specific treatment. In our case, the surgical excision was done because the patient was symptomatic and the size of the lesion was significant, causing compressive symptoms.

We report this case because of its characteristic radiological and histopathological appearance and stress the importance of considering this entity in the differential diagnosis of skull lesions.

References

1. Vega R.A., Hidlay D.T., Tye G.W., Fuller C.E., Rhodes J.L., Intradiploic Dermoid Cyst of the Lateral Frontotemporal Skull: Case Report and Review of the Literature. *Pediatric Neurosurgery* 2013; 49:232-235
2. Mehra S, Chandra GU, Kumar S. Intradiploic epidermoid tumor of temporal bone X-ray, CT, MR Imaging. *Indian Journal of Otolaryngology and Head and Neck Surgery* [serial online] 2012 [cited 2014 Nov 11]; 18:98-102.
3. S. Gaivas, D. Rotariu, G. Dumitrescu, B. Iliescu. Intradiploic epidermoid cyst of the skull. Case report. *Romanian Neurosurgery XVIII* (2) 2011.
4. Brandon Miner, DO; Paul Aanderud; Steven K. Grekin. Giant Intradiploic Epidermoid Cyst of the Skull: A Case Report and Management of Large Cystic Scalp Masses. *Cosmetic Dermatology* 2009, vol. 22(7)
5. Shibani Mehra, Garga Umesh Chandra, Suresh Kumar. Intradiploic epidermoid tumor of temporal bone X-ray, CT, MR Imaging. *Indian Journal of Otolaryngology and Head and Neck Surgery* 2012; 18:98-102.
6. Narlawar R S, Nagar A, Hira P, Raut A A. Intradiploic epidermoid cyst. *Journal of Postgraduate Medicine* 2002;48:213
7. Bikmaz K, Cosar M, Bek S, Gokduman CA, Arslan M, Iplikcioglu. Intradiploic epidermoid cysts of the skull: a report of four cases. *Clinical Neurology and Neurosurgery*. 2005 Apr;107(3):262-7

8. Aldo F. Berti, MD, FACS, FAAP, Gregory C. Lovaas, MD, FACS, Alejandro Santillan. Primary Intradiploic Pterional Epidermoid Cyst. *Southern Medical Journal*. 2010; 103(1):87-89.
9. Russel DS, Rubinstein LJ. Tumors and tumorlike conditions of maldevelopmental origin. Russel DS Rubenstein LJ, editors. Pathology of tumors of nervous system. 5 th ed. London: Edward Arnold; 1989.p. 693-5.
10. Wolfgang Krupp, Alexander Heckert, Heidrun Holland. Giant intradiploic epidermoid cyst with large osteolytic lesions of the skull: a case report. *Journal of Medical Case Reports* 2012, 6:85