



A Case of Giant Extracalvarial Epidermal Cyst with Intracranial Extension

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Abstract

Extracalvarial epidermal cysts are benign, uncommon type of tumors. Their incidence is around 0.2 to 1%. They are usually asymptomatic except for the absurd swelling over the scalp, which is associated with headache and rarely neurological deficits. Proper radiological diagnosis with accurate surgical approach and excision with capsule gives a good prognosis and prevents recurrence. Here we present the case of a 75-year-old gentleman with a large swelling over the scalp with mild pain. Surgical excision of the tumor with capsule was done followed by cranioplasty. The histopathology was consistent with an epidermal cyst. Case reports pertaining to such conditions are few.

Keywords

- bony erosion
- extracalvarial epidermal cyst
- giant swelling

Introduction

An extracalvarial epidermal cyst is a benign, uncommon type of tumor. Its incidence is around 0.2 to 1%.^{1–3} Epidermoid cysts of the skull of primary origin are rare. The first reported case dates back to the 19th century by Muller.⁴ In most cases, they are slow growing, benign, and congenital, and derive from the ectodermal remnants misplaced during embryogenesis.⁵ A few can also occur following a trauma.⁶ Malignant changes in such tumors may occur rarely.⁷ These cysts vary in size, location, and rate of progression. The most common symptom is a long-standing, asymptomatic lump on the head. Headache, focal tenderness, seizures, and traumatic rupture can occur. Rarely, very large lesions may be associated with focal neurological deficits.⁸ Radiological investigations include skull X-ray, computed tomography (CT), and magnetic resonance imaging (MRI). Histopathological analysis is confirmatory for diagnosis.

Case Report

A 75-year-old gentleman presented to us in the outpatient department with complaints of a large lump over the head for the past 40 years, which was slowly progressing in size

over the years. It was the first time in four decades that he consulted a medical professional for the swelling as he developed mild but tolerable pain for the last 1 month with mild itching over the swelling (► **Fig. 1**). On examination, the size of the swelling was 15 cm × 13 cm × 7 cm extending over the biparietal and posterior frontal regions. It was irregular in shape with a smooth surface. The swelling was soft in consistency and skin over the swelling was pinchable and the swelling was mobile. There was no engorged vein, no visible pulsation, no localized rise in temperature, no cough impulse, and no other skin changes.

Magnetic resonance imaging (MRI) revealed a large expansile lytic lesion of measuring 14 cm × 11 cm × 5 cm in size arising from the parietal bone causing erosion of the bone bilaterally (► **Fig. 2**). The lesion was limited to the dura, causing mass effect on the left parietal lobe with effacement. There was no contrast enhancement.

The patient was taken for surgery under general anesthesia. Posterior bifrontal skin flap was planned and raised. Tumor tissue was covered by a pale yellow-colored capsule, which was dissected off from the scalp layers. The capsule was densely adhered to the pericranium. Yellow-colored pultaceous cheesy material was drained (► **Fig. 3**). The capsule with tumor tissue was excised in toto. The

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Fig. 1 Giant swelling present over the frontoparietal region.

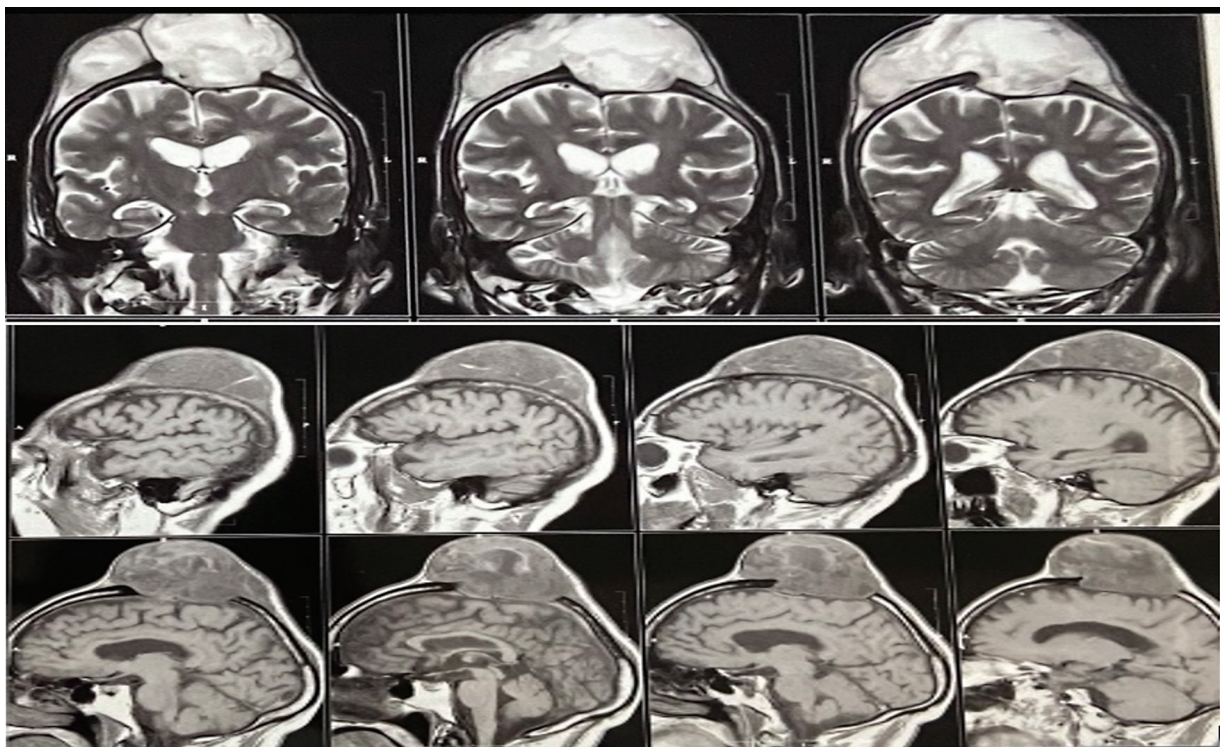


Fig. 2 Magnetic resonance imaging (MRI) showing large expansile extradural lesion in the parietal region causing mass effect and effacement on the left parietal lobe.

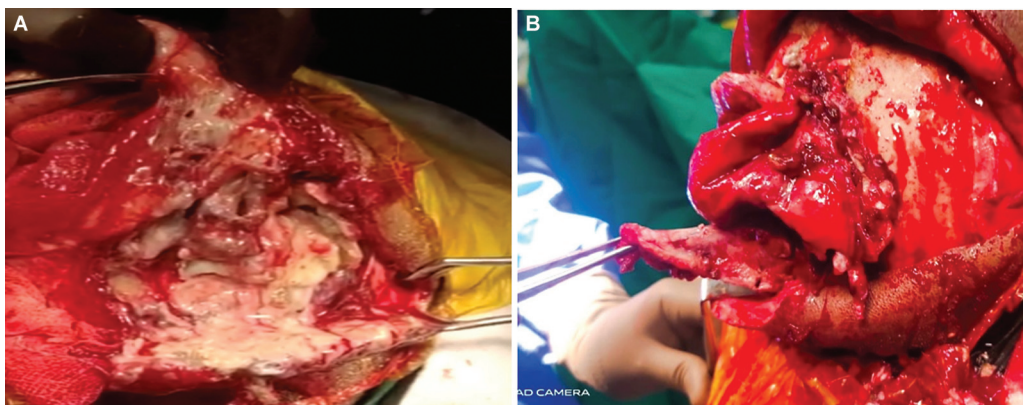


Fig. 3 (A) Pultaceous cheesy material draining out and (B) pale yellow capsule of the tumor.

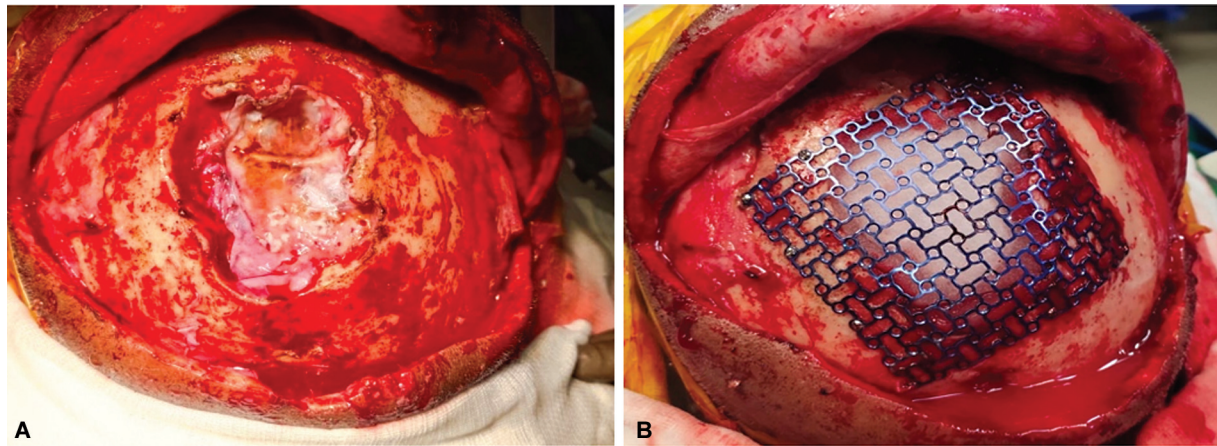


Fig. 4 (A) Large bony defect. (B) Cranioplasty done with titanium mesh.

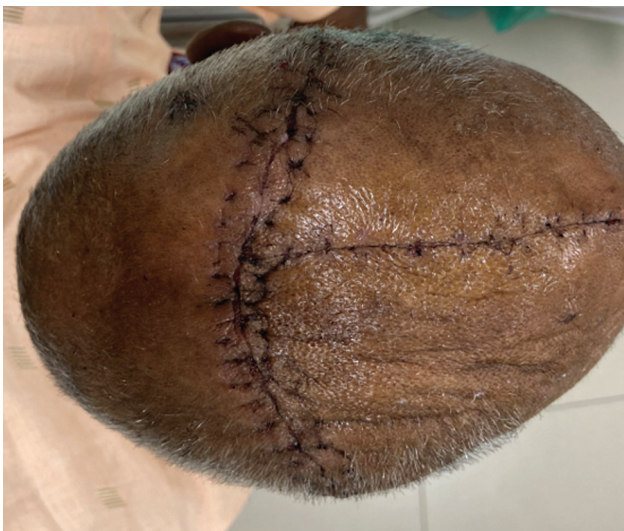


Fig. 5 Postsuture removal and Y-plasty flap.

bony defect (►Fig. 4) was identified and the cyst wall from the dura was gently scraped. There was no intradural extension of the tumor. Cranioplasty was done with a titanium mesh. Extra skin was trimmed and Y-plasty was

done and skin was closed in layers. The postoperative period was uneventful. Sutures were removed on day 10 (►Fig. 5).

Histopathology suggested laminated keratin material and cholesterol crystals with cellular debris, which were consistent with the diagnosis of epidermal cyst (►Fig. 6). The patient has been followed up for the past 6 months without any sign of recurrence.

Discussion

Epidermal cysts can occur in most parts of the body but are most commonly seen in the trunk, face, neck, and genitals. They may be infected or enlarged. These cysts are usually mobile and vary in size depending upon the time of presentation. The cause of the bony defect is hypothesized to be persistent pressure by the expanding lesion.⁹

Intradiploic cysts constitute 25% of extracalvarial tumors, whereas the rest are intradural.² Differential diagnosis includes dermoid cyst, hemangioma, fibrous dysplasia, and eosinophilic granuloma. Atypical epidermoid cysts may be difficult to distinguish from other lytic lesions of the calvaria.¹⁰

Cushing stated that the aim of surgery was complete excision of the tumor along with its capsule and thorough

Table 1 Similar cases in the literature

Sl. no.	Article title	Year of publication	Authors
1	Giant intradiploic epidermoid tumor of the occipital bone: case report	1990	Guridi et al ¹
2	Intradiploic epidermoid cysts of the skull: report of 10 cases and review of the literature	1990	Ciappetta et al ²
3	Intradiploic primary epithelial inclusion cyst of the skull	2006	Kalgutkar et al ⁵
4	Intradiploic frontal epidermoid cyst in a patient with repeated head injuries: is there a causative relationship?	2006	Locatelli et al ⁶
5	A giant intradiploic epidermoid cyst with perforation of the dura and brain parenchymal involvement	2007	Cho et al ⁹
6	Giant intradiploic epidermoid cyst with large osteolytic lesions of the skull: a case report	2012	Krupp et al ¹⁴

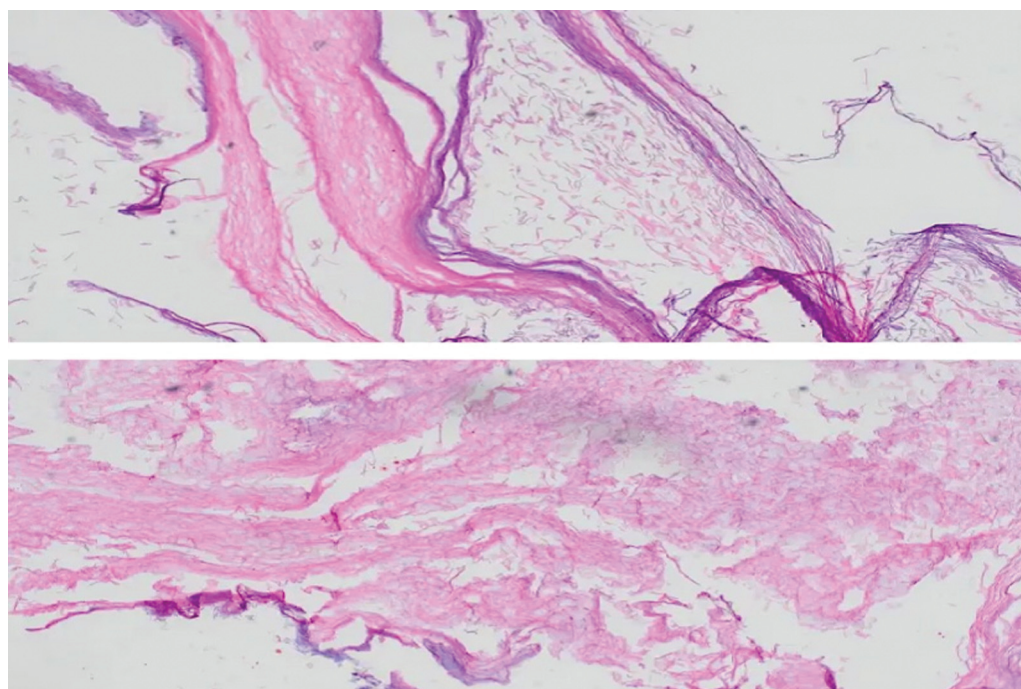


Fig. 6 Histopathological slide showing cellular debris and laminated keratin with cholesterol crystals.

curettage of the dura.¹¹ Incomplete removal causes a recurrence rate of 8.3 to 25%.¹²

To our knowledge, very few cases of giant intradiploic epidermoid cysts with extensive bony defects and deformation of brain have been described in the literature (► **Table 1**). The time interval between the appearance of the lump and first consultation for it is about four decades, which is uncommon and corresponds only to few other case reports, and is the first case in which a slowly progressive, painless proptosis of an eye was described.¹³ In the other case report, the patient presented with dizziness and focal retinal detachment.¹⁴ Among the others cases, either the presentation was within few years or the pathology was purely intracranial.

Our patient experienced no restrictions of his daily life activities or social contacts pre- or post-op. Post-op his head felt light and he no longer felt different from the rest of the people around physically. Surprisingly our patient never experienced problems laying down on either side preoperatively. Cranioplasty was done in our patient to avoid postoperative complications and for good cosmesis.

Conclusion

Extracalvarial epidermal cysts are benign, slow-growing tumors. Unfortunately, even in 2024 there are patients living with such large resectable tumors. Despite their large size, they do not cause debilitating symptoms. If the patient is willing, surgery should be advised to them, as complications occur very rarely postoperatively. Cranioplasty should be performed in large bony defects.

Complete removal of the capsule is a must while dealing with such tumors.

Conflict of Interest

None declared.

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