

A Rare Case of Giant Intradiploic Epidermal Cyst of the Frontal Bone with Intracranial Extension

Abstract

Calvarial intradiploic epidermal cyst are very rare neoplasms which can have intracranial extension. These cysts can attain a large size and can cause lytic destruction of the calvarium resembling malignancy. Since these lesions are benign complete excision is curative. We report the case of a 77-year-old male patient who presented with a large swelling on the forehead. Computed tomography and magnetic resonance imaging showed an expansile lytic lesion with intracranial extension. Complete excision of the lesion with cranioplasty was done. Histopathology was consistent with the epidermal cyst. These cysts attaining gigantic proportions are very few in literature.

Keywords: Calvarial, intracranial, intradiploic epidermal cyst

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Introduction

Epidermoid cyst is a benign inclusion cyst formed from epidermal elements and contains keratin. They account for <1% of all intracranial neoplasms.^[1,2] These cysts arising from the calvarial diploe and growing to such large size with intracranial extension is extremely rare.^[3]

Case Report

A 77-year-old male patient presented with gradually progressive swelling of the scalp of 6 years duration. There was no history of headache or any discomfort pertaining to the lesion. The swelling gradually increased in size, and the patient tried to hide the swelling by wearing a turban. However, for the past 1-month patient had on and off headache with difficulty to sleep and neck pain due to the giant size of the tumor. Furthermore, he could not hide the tumor using a turban as the size of the tumor was huge.

On examination

A swelling of size 19 cm × 12 cm × 7 cm extending fully over the bifrontal scalp noted. The lesion was extending posteriorly up to the middle of parietal convexity, and laterally, the lesion was seen overhanging the temple [Figure 1].

The skin over the swelling appeared smooth, and there was loss of hair over the swelling. No visible engorged veins or visible pulsations present. The swelling was soft in consistency, and the skin over the swelling was pinchable. Mobility and fixity could not be assessed due to the large size of the swelling. The swelling was nonpulsatile, and there was no impulse on coughing. The patient was further evaluated with routine blood investigations, which were within the normal limits. The X-ray of the skull showed midline erosion of the calvarium of the frontal and parietal bones [Figure 2]. Magnetic resonance (MR) imaging showed a large expansile lytic lesion of size 18 cm × 9 cm × 6 cm arising from the intradiploic region of the frontal bone causing destruction of both tables of the frontal bone, the lesion was limited by the dura causing mass effect on the frontal lobes and effacement of the frontal horns. There is no contrast enhancement and MR venogram showed decreased filling of the superior sagittal sinus in the anterior one-third [Figure 4].

The lesion was excised completely. Pale yellow capsule was dissected free from the scalp layers. The capsule was adherent to the bone with destruction of the bone. The capsule was opened, and yellow-colored pultaceous material was removed. The

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capsule was excised completely. The area of the bone where the capsule was adherent was nibbled. There was no intradural extension or dense attachment to the superior sagittal sinus. Cranioplasty was done with a titanium mesh implant [Figure 3]. Histopathology was consistent with epidermal cyst.

Discussion

Intradiploic epidermoid cysts are very rare lesions. They are inclusion cysts formed due to the incorporation of epithelial islands into the developing bone.^[4] Usual sites of intradiploic epidermoid cysts include skull and phalanx. It is characterized by expansile growth, characterized by thinning of the bone, and can mimic a lytic bone

malignancy.^[5] The intradiploic epidermoid of the skull can attain huge sizes and are primarily extradural.^[6] Our lesion was one among the largest of such lesions reported. From the literature review in PubMed and google scholar, we could find only one case report of such a giant lesion.^[7] Usual sites of origin of these are in the frontoparietal and occipital region.

The most common site of occurrence of an intradural epidermoid cyst is the cerebellopontine angle (40%) followed by the suprasellar cistern (18%).^[8] Other less common sites include Sylvian fissure, basal cisterns, pineal region, petroclival region, and the lateral and fourth ventricles.^[9] These lesions usually present with cranial nerve involvement according to the site of origin and mass effect due to such lesions are rare due to the tendency to grow along the cisterns. Cerebellopontine epidermoid cysts can present with hemifacial spasms or trigeminal neuralgia, whereas suprasellar epidermoid cysts can cause decreased visual acuity as well as visual field defects. Rupture of these cysts, especially during excision can result in chemical meningitis. The intradiploic epidermoid cysts usually present with mass effect and local headache due to the extradural location and very slow progression. Embryologically the intradural epidermoid cysts are formed due to the trapping of ectodermal cells before the closure of the neural tube, while the intradiploic epidermoid cysts are formed in a similar manner, after the neural tube



Figure 1: (a and b) Clinical images showing the swelling (photos taken after obtaining the informed written consent from the patient)

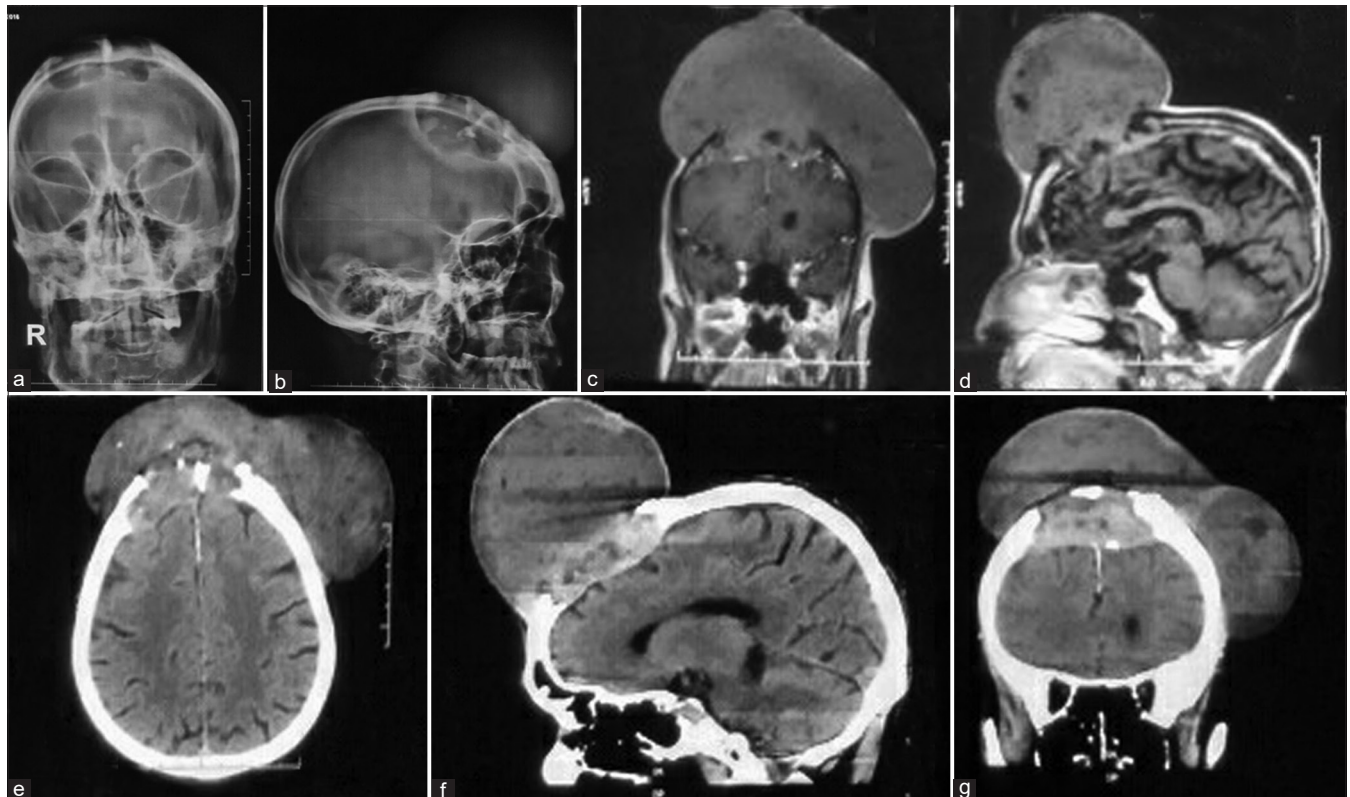


Figure 2: Imaging of the patient: (a and b) X-ray skull showing calvarial erosion; (c and d) magnetic resonance imaging coronal and sagittal images; (e-g) computed tomography axial, sagittal, and coronal images, respectively

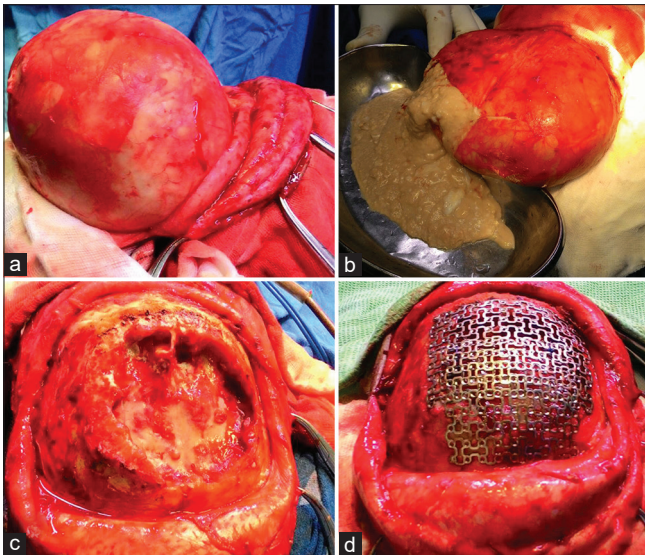


Figure 3: Per operative images; (a) cyst dissected out from the scalp; (b) cyst opened and pultaceous material evacuated; (c) eroded calvarium; (d) titanium mesh cranioplasty done

closure. Being extradural, complete excision and a better outcome is feasible with intradiploic epidermal cysts. But in intradural epidermal cysts recurrences are common since the cyst wall is adherent to vital structures which will hamper the complete excision.^[8,9]

These lesions are characterized by a pearly white cyst wall containing cheesy material. The wall is formed of stratified squamous epithelium and the cheesy content is formed by desquamated keratin and cholesterol crystals. These cysts, when formed in the skull, causes scalloping of the skull bone and are present in the subgaleal space.^[10] These cysts are attached to the scalloped bony edges. Similarly, in our case also the cystic tumor was adherent to the bone edge of the eroded calvarium which was nibbled to prevent the recurrence. These are slow-growing lesions and cause symptoms such as headache due to mass effect on the dura and underlying brain. Very rarely they can have a secondary infection or malignant change.^[11]

These lesions have a very good prognosis since the complete excision of the cyst wall leads to cure.^[11] In our patient since the cyst wall was adherent to the bony edges, we curetted out the cyst wall completely and nibbled the bone edge.^[12] Usually, cranioplasty can be done in the same instance if the defect is large and there is no evidence of infection. If there is intradural extension complete excision of the lesion is difficult since there may be multiple pseudopod-like extensions of the lesion through the arachnoid.^[13]

Conclusion

Intraosseous epidermoid is a differential diagnosis for osteolytic lesions of the skull with swelling attaining giant proportions. Such lesions are seen in developing countries such as India even in the 21st century. Even though the

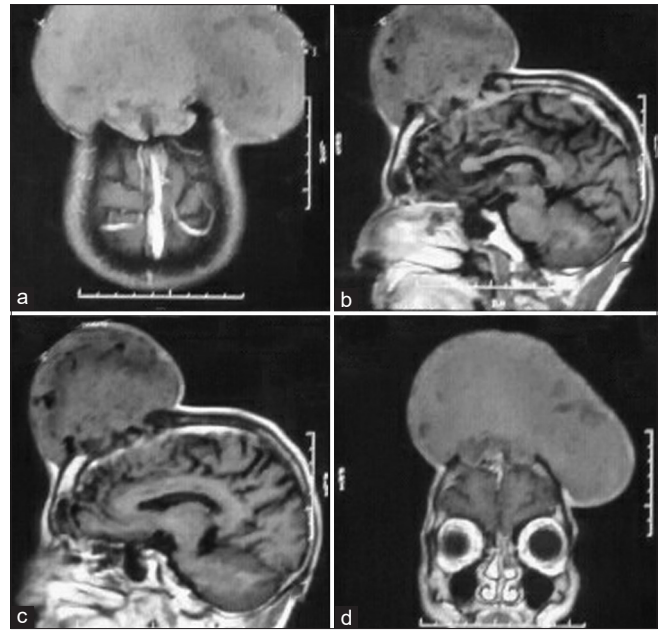


Figure 4: Magnetic resonance imaging of the brain with contrast; (a) axial (b and c) sagittal and (d) coronal sections showing partial occlusion of superior sagittal sinus

lesion was giant with an erosion of the calvarium, the complete excision with cranioplasty could be accomplished.

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Conflicts of interest

There are no conflicts of interest.

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