Craniofacial epidermoid cysts are rare tumors representing 0.2%–1% of all the intracranial tumors. Intradiploic variants account for 25% of these cysts. These cysts are benign, slow-growing, congenital tumors derived from ectodermal remnants misplaced during embryogenesis. Mean age at presentation of these lesions is 40 years; epidermoid cysts are typically asymptomatic (Toglia JU, Netsky MG, Alexander E Jr. Epithelial (epidermoid) tumors of the cranium. Their common nature and pathogenesis. J Neurosurg 1965;23:384-93). Giant extradural epidermoid cysts with profound deformation of the brain and extensive lytic skull lesions may allow a normal life without any significant neurological deficits. Computed tomography scan and magnetic resonance imaging play an important role in the diagnosis of the lesion and management protocol. Histologically, epidermoid cysts are lined by stratified squamous epithelium and are filled with anucleatic keratin (Hao S, Tang J, Wu Z, Zhang L, Zhang J, Wang Z. Natural malignant transformation of an intracranial epidermoid cyst. J Formos Med Assoc 2010;109:390-6). Complete removal of the cyst along with its capsule is the treatment of choice. Recurrences of intradiploic epidermoid cyst have been reported. Here, we report a case of 14-year-old girl presenting with a giant frontal intradiploic epidermoid cyst with an intracranial and extracranial extension without any neurological deficits. Bifrontal craniotomy was performed and cyst was excised in toto.

**Keywords:** Epidermoid cyst, extracranial extension, frontal, giant, intradiploic, intracranial extension

---

**INTRODUCTION**

Epidermoid cysts are quite uncommon in the craniofacial region, more so in the intradiploic region of the skull. They are slow-growing, benign cystic lesions. Intracranial extensions of the tumors occur rarely. Epidermoid cysts with both extracranial and intracranial extension are extremely rare. Accurate radiological diagnosis followed by excision of cystic lesion is an ideal treatment.

**CASE REPORT**

A 14-year-old girl presented with complaints of swelling over the forehead for the past 5 years, gradually progressive, not associated with pain, and no history of trauma. On examination, the patient was found to be clinically stable with a 5 × 5-cm, smooth, nontender, hard swelling on the forehead in the midline extending onto the left side.

Computed tomography (CT) scan revealed a 5.1 × 5.2-cm cystic lesion in expanded left frontal sinus extending into the anterior cranial fossa. Magnetic resonance imaging (MRI) revealed a 5.1 × 5.2-cm T1 heterogeneously hypointense and T2 hyperintense...
cystic lesion arising from the frontal bone in the midline with thinning of inner and outer tables of frontal bone associated with cortical buckling of bilateral anterior frontal lobes showing minimal peripheral enhancement postcontrast and restriction on diffusion-weighted imaging (DWI) [Figure 1].

Bifrontal craniotomy was performed and the cyst was excised in toto [Figure 2] followed by cranialization of the frontal sinus. Excised specimen was sent for histopathological examination, which revealed a cyst lined by keratinized squamous epithelium with loose keratin tissue in the cystic cavity [Figure 3].

Postoperative course was uneventful with good aesthetic results.

**DISCUSSION**

Epidermoid cysts popularly known as “Tumeur perles” (pearly tumors) were first described by Cruveilhier in 1835. Intradiploic epidermoid cyst was first described by Cushing in 1922. They represent 0.04%–0.7% of intracranial neoplasms. They are considered pseudotumors as they occur due to sequestration of ectodermal elements within the cranial cavity during the neural tube formation because of defective separation of neuroectoderm. Posttraumatic etiology was considered in a few cases. Enlargement of the intradiploic epidermoid cysts occurs by desquamation of the normal cells into the cystic cavity and not by
active cell division, thus explaining the slow-growing, benign nature of the cyst; however, they may rarely undergo malignant transformation into a squamous cell carcinoma.[2]

Mean age at presentation of these lesion is 40 years; epidermoid cysts are typically asymptomatic.[9] Giant extradural epidermoid with profound deformation of the brain and extensive lytic skull lesions may allow a normal life without any significant neurological deficits.[10]

CT scan plays an important role in the assessment of the skull involvement and intraparenchymal extension. Typically, epidermoids are hypodense on CT scan. Thinning and expansion of the cortex of outer and inner tables on CT scan indicates a slow-growing intradiploic cyst. MRI is useful in assessing the intracranial extension of the tumor. It appears hypointense on T1-weighted and hyperintense on T2-weighted MR sequences.[3] High signal intensity on DWI and lack of signal suppression on fluid-attenuated inversion recovery sequences of MRI are additional features of epidermoid cyst on MRI.

Histologically, epidermoid cysts are lined by stratified squamous epithelium and are filled with keratin.[9]

Complete removal of the cyst along with its capsule is the treatment of choice. Recurrences of intradiploic epidermoid cyst have been reported.[7]

**Conflicts of interest**
There are no conflicts of interest.

**REFERENCES**

1. Jimenez DF, Savage JG, Samuelson M. Developmental anomalies: Arachnoid cysts, dermoids and epidermoids. In: Ellenbogen RG, Abdulrauf SI, Sekhar LN, editor. Principles of Neurological Surgery. 3rd ed. Philadelphia: Elsevier Saunders; 2012. pp. 129-35.
2. Hao S, Tang J, Wu Z, Zhang L, Zhang J, Wang Z. Natural malignant transformation of an intracranial epidermoid cyst. J Formos Med Assoc 2010;109:390-6.
3. Hassaneen W, Sawaya R. Epidermoid, dermoid, and neuroenteric cysts. In: Winn HR, editor. Youmans Neurological Surgery. 6th ed. Elsevier Saunders; 2011. pp. 1523-8.
4. Locatelli M, Alimehmeti R, Rampini P, Prada F. Intradiploic frontal epidermoid cyst in a patient with repeated head injuries: Is there a causative relationship? Acta Neurochir (Wien) 2006;148:1107-10; discussion 1110.
5. Mehra S, Chandra GU, Kumar S. Intradiploic epidermoid tumor of temporal bone X-ray, CT, MR Imaging. Indian J Otolaryngol Head Neck Surg 2012;18:98-102.
6. Sirin S, Gonul E, Kahraman S, Timurkaynak E. Imaging of posterior fossa epidermoid tumors. Clin Neurol Neurosurg 2005;107:461-7.
7. Toglia JU, Netsky MG, Alexander E Jr. Epithelial (epidermoid) tumors of the cranium. Their common nature and pathogenesis. J Neurosurg 1965;23:384-93.
8. Vega RA, Hidlay DT, Tye GW, Fuller CE, Rhodes JL. Intradiploic dermoid cyst of the lateral frontotemporal skull: Case report and review of the literature. Pediatr Neurosurg 2013;49:232-5.
9. Wolfgang K, Alexander H, Heidrun H. Giant intradiploic epidermoid cyst with large osteolytic lesions of the skull: A case report. J Med Case Rep 2012; 6:85.
10. Zavanone M, Guerra P, Rampini PM, Crotti F, Vaccari U. A cervico-dorsal intramedullary epidermoid cyst. Case report and review of the literature. J Neurosurg Sci 1991;35:111-5.