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Intracranial Epidermoid Cyst in a Middle Aged Female: A Rare Case Report

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Abstract

Intracranial epidermoid cysts are uncommon congenital lesions which account for about <1% of all intracranial tumors. They are the consequence of ectodermal materials being included during neural tube closure, and because of the mass effect on surrounding tissues, they usually manifest in middle life. The material they contain is derived from desquamated epithelial cells. Epidermoid cysts are primarily congenital; however, they might take years to manifest since they grow extremely slowly. We present a rare case of intracranial epidermoid cyst extending into the frontal bone in a 57 years old female. Bifrontal craniotomy with excision of cyst was performed safely without any neurologic deficit. Subsequent histopathological analysis identified the lesion as an epidermoid cyst. **Keywords:** Intracranial epidermoid cysts; CT Scans; MRI Scans

Introduction

Intracranial epidermoid cysts are rare congenital diseases that account for around 1% of all intracranial tumors. They are caused by the incorporation of ectodermal components during neural tube closure and usually appear in middle age due to the bulk effect on adjacent tissues. Although epidermoid cysts are mostly congenital, they are often very slow developing and take many years to manifest. Patients often range in age from 20 to 40. There may be an increase in male predominance [1,2]. An uncommon association exists with anorectal anomalies, sacral anomalies and presacral mass, and is known as the Currarino triad [1].

Epidermoid cysts can be congenital (the most frequent, resulting from ectodermal inclusion after neural tube closure) [3] or acquired (after surgical or traumatic implantation). They are distinct from dermoid cysts, which have both epidermal and cutaneous appendages such as hair and sebaceous cysts, and mature teratomas, which have all three layers. We present a rare case of intracranial epidermoid cyst extending into the frontal bone in a 57 years old female. Bifrontal craniotomy with excision of cyst was performed safely without any neurologic deficit. Subsequent histopathological analysis identified the lesion as an epidermoid cyst.

Case Report

A 57 years old female presented with painless swelling over the forehead since last 3 months. She had similar swelling over the same region since last 15yrs and she underwent excision in a local hospital with a diagnosis of dermoid cyst 4 months before presenting to our institute. There was no history of any neurological symptoms and she had no comorbidities. Her routine blood workup was normal. She underwent CECT brain which revealed hypodense lesion measuring 7 x 4 cm in the frontal lobe extending into the cortex of adjacent bone [Figure 1,2]. He was referred to our neurosurgical unit.

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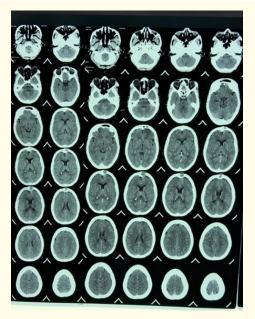


Figure 1

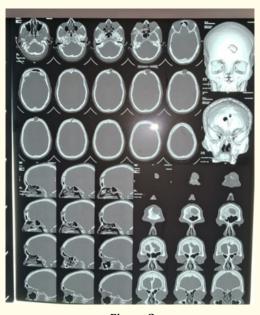


Figure 2

After thorough clinical examination and reviewing the scan, patient underwent Bifrontal craniotomy with excision of the lesion with part of frontal bone under general anesthesia [Figure 3]. The defect was closed with titanium mesh [Figure 4]. Specimen was sent for histopathological examination. She had uneventful postoperative outcome without any neurologic deficit. Subsequent histopathological analysis identified the lesion having fibrocollagenous cyst wall lined by stratified epithelium with lumen containing lamennated keratin suggestive of epidermoid cyst. Cyst wall is seen to rest on underlying bone [Figure 5, 6].

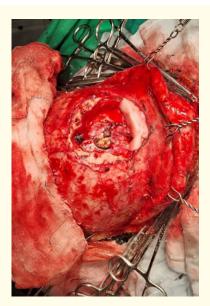


Figure 3

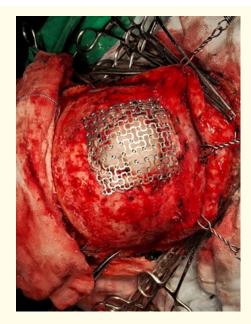
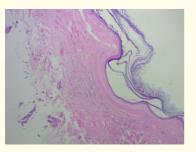


Figure 4

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Figure 5





Discussion

Epidermoid cysts are slow-growing benign and congenital inclusion cysts that can arise in both extracranial and intracranial locations in individuals of all ages. Intracranial epidermoid cysts, which account for up to 1.8% of all primary intracranial tumors [4], are often seen in the sellar and suprasellar areas, the middle fossa, the cerebellopontine angle, and off-center in the posterior fossa [5,6]. Meanwhile, extracranial epidermoid cysts develop elsewhere in the body when embryonic components combine. According to previous research, epidermoid cysts are mostly found in the ovaries and testicles (80%), with 7% in the head and neck and 1.6% in the oral cavity [7]. Epidermoid cysts in the head are uncommon but often create cosmetic issues.

Signs and symptoms of intracranial epidermoid cysts are due to gradual mass effect, with presentation including headaches (most common), cranial nerve deficits, cerebellar symptoms, seizures, raised intracranial pressure. Recurrent aseptic meningitis is uncommon but recognized, similar to the less common dermoid cyst [2]. Most epidermoid cysts on CT scans are hypodense and lack contrast due to the presence of debris, keratin, water, and cholesterol within them. MRI scans often show hyper intensity on T1WI, variable appearance on T2WI, and absence of contrast enhancement with gadolinium [8]. Few findings confirm that cerebral epidermoid cysts have an atypical look on CT and MRI scans due to calcification, albumin, iron, copper, and elevated protein content [9-11]. Histopathologically Epidermoid cysts often include a thin capsule of squamous epithelium and an interior cystic component with desquamated epithelial cells and cholesterol crystals on histologic inspection. Epidermoid cysts have a glossy, mother-of-pearl look with a nodular outer surface [12]. If the patient is symptomatic or incidentally diagnosed with imaging, surgical excision is the recommended therapy. However, total resection is challenging since not all tissue can be removed, particularly around the bones, cranial nerves, and veins [9,10]. Recurrence is thus not unusual, though development is normally modest, and many years may pass without new symptoms [10].

In this report, we present an intracranial epidermoid cyst extending into frontal bone and presenting as a dermoid. One has to keep in mind the differentials before going ahead with the excision of the swelling on an outpatient basis.

Conclusion

Intracranial epidermoid tumors are benign, slowly growing lesions that may be treated. With sophisticated microsurgery methods, total resection ought to be the aim of care. With slow and meticulous piece meal removal, capsular bits can be teased from the surrounding structures in most cases. Intracranial epidermoid tumors are difficult clinical entities despite their benign biology because of their high rates of postoperative morbidity, especially in patients with more sparing partial resection, their high recurrence statistics, and the absence of systemic therapy options.

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