Fast-Growing Intradiploic Epidermoid Cyst with Osteolytic Features in the Parietal Bone of an Adult: A Case Report

Peiran Xu1, Chengxian Yang1, Xueyuan Li1, Yayan Bi2, Yunxiao Meng3, Wenbin Ma1, Renzhi Wang1 and Xinjie Bao*

1Department of Neurosurgery, Peking Union Medical College Hospital, China
2Department of Pathology, Peking Union Medical College Hospital, China

Abstract

Epidermoid cysts are rare but slowly-growing congenital lesions of central nervous system that mainly reside in the subdural space. Diploic epidermoid cysts are occasionally encountered in clinic, often with a slowly enlarging size and majorly in children. The present study reports a case of fast-growing intradiploic epidermoid cyst with osteolytic features in the parietal bone of an adult. A 37-year-old male presented with a rapidly-growing mass in his left frontoparietal region without subjective complains. Brain CT and MRI showed a well-defined intradiploic mass in the left parietal bone, which had homogenously low T1 signal, high T2 signal, but no visible enhancement. The lesion was totally removed with a left frontoparietal craniotomy. Histological examination revealed an epidermoid cyst with osteolytic features. One-year follow-up CT found no recurrence. This case shows one fast-growing intradiploic epidermoid cyst with osteolytic features in the parietal lobe of an adult with a good recovery after surgery.

Keywords: Epidermoid cysts; Histological examination; Frontoparietal craniotomy

Introduction

The epidermoid cyst is a rare lesion of benign kind, representing between 0.2% and 1% of all intracranial tumors. 75% are located in the intracranial space and 25% within the diploic spaces [1-3]. Mostly the intradiploic epidermoid cyst is congenital tumors which derive from ectodermal remnants misplaced during embryogenesis [4]. Therefore children constitute the major part of patients. However in a few cases the posttraumatic etiology was also considered. In 1838, it was first described by Muller [5]. Since then, only about 120 cases have been reported and less than 5 cases were involved the frontoparietal area [6,7]. We present a report of a rare intradiploic frontoparietal epidermoid, with skull and cerebral dura mater invasion, in a 37-year-old male. We believe it is the first report such a lesion arising near the pterion in the adults.

Case Presentation

Patient presentation and diagnosis

A 37-year-old male was admitted to Peking Union Medical College Hospital (Beijing, China) for complaining with a rapidly growth mass in his left frontoparietal region. The patient had one-year history of a mass that was initially 1 cm in diameter without pain even under pressing. Because of no headache, dizziness, epilepsy and movement disorder of limb, he did not refer to hospital. The mass grew rapidly and the patient went to our hospital due to elastics of the mass. Medical records report no head traumatic history, disorders in other systems, and related family diseases. Physiological examination found no neurological signs but a visible 5-cm-diameter mass that felt soft, elastic and smooth without pain. The overlying skin showed no pathologic signs like defects and inflammatory secretion. Blood tests were within normal limits. Head Computed Tomography (CT) revealed an intradiploic lesion of 5.9 cm × 2.6 cm with well-defined margins and osteolytic depression in the frontoparietal bone (Figure 1). Magnetic Resonance Imaging (MRI) showed a heterogeneous mass having a low T1 and high T2 signal without notable enhancement on Gd contrast (Figure 2). He was diagnosed as a bone cystic tumor and scheduled to surgical operation.

Treament

A U-shaped craniotomy was performed on the frontoparietal region. The scalp and temporal muscle totally were cut, separated from the bone and turned to the cranial base to expose the mass. A careful observation showed that cranial outer table was partially eroded by cheese-like materials.
contained in the mass. After a complete exposure of the tumor, the mass was drilled from the skull at 1cm distance. There was adhesion between the tumor and the cerebral dura mater. We separated the tumor from the cerebral dura mater completely and carefully. The inner table was eroded and the tumor’s capsule was unbroken. The tumor measuring 5 cm × 5 cm × 2.5 cm was soft. Cutting open the capsule, we obtained large amounts of white pearly material. The tumor was removed completely, including its capsule. The cerebral dura mater was not involved and left intact. Cranial repaired with an inner table graft and titanium mesh. After that we closed the scalp layer by layer and returned the patient to his ward. His postoperative course was uneventful and neurological examination was the same as pre-operation. Pathological diagnosis and prognosis: Macroscopic evaluation disclosed an irregular, oval piece of friable, pale, pearly tissue measuring 5 cm in its greatest dimension. Histologic examination revealed that the cyst wall consisted of disintegrated keratinizing epithelium layer, keratinizing epithelium layer, and stratified squamous epithelium layer from inner to outer, underlying collagenous connective tissue containing adnexal structures. According to these pathological findings, epidermoid cyst was considered as the diagnosis (Figure 3). A postoperative CT one year post-surgery revealed the correct placement of the implant (titanium mesh) without evidence of a residual tumor (Figure 4).

**Discussion**

Epidermoid cysts of the skull are rare, benign tumors, accounting for less than 1% of all intracranial tumors. These tumors originate from ectodermal remnants within the neural tube during its closure.
at 3 and 5 weeks of gestation [1,2,4]. Intradiploic epidermoid cysts are derived from ectodermal remnants that stay within the cranial bones. Patients who get the tumors always have painless subcutaneous scalp swelling covered with normal skin, like the patient in this case. A few patients may have headache, nausea, dizziness or develop a sudden grand mal seizure because the brain lobes are compressed by tumors. Signs and symptoms depend on tumor location. For the radiological diagnosis, CT and MRI allow for the good assessment of both skull involvement and intracranial extension. The typical CT aspect is a large homogenous hypodense mass, with or without calcifications [1]. The characteristic MRI findings of intradiploic epidermoid cysts include well-demarcated osteolysis, high signal intensity on T2-weighted images, and varied signal intensity on T1-weighted images [8]. The low intensity signals are probably the result of cellular debris and solid cholesterol crystals within the cyst. As for pathologic findings, epidermoid cysts have a thin capsule of stratified squamous epithelium filled by keratin, cellular debris and cholesterol, and do not contain hair or other dermal elements [5,9-13]. The differential diagnosis includes dermoid cysts, eosinophilic granuloma, fibrous dysplasia, osteomyelitis, and metastatic lesions. Dermoid cysts usually occur in relation to the periorbital area, suture lines and the midline [10,11]. Osteomyelitis and fibrous dysplasia may present with some degree of sclerosis but epidermoid cysts appear mostly with a definite and sclerotic border. And lesions of eosinophilic granuloma and metastatic are not usually rimmed. According to the literature, observation versus surgical treatment of intradiploic epidermoid cyst depends on whether or not the lesion is symptomatic. Surgical resection is recommended in patients with large symptomatic lesions to relieve mass effect. In smaller, asymptomatic lesions, close observation with serial imaging studies is a reasonable and accepted option [14]. Malignant changes can occur in the epithelial lining of intradiploic epidermoid cysts, especially in the relapsing tumors or tumors with inflammation [15]. Therefore complete removal of the cysts should be accomplished with considering the patient’s age, co-morbidity, potential neurological damage and facilities of pre-operative assessment of the tumor.

Conclusion

In conclusion, we present the case of a rare intradiploic epidermoid cyst. Intradiploic epidermoid cysts of the skull are rare, slow growing, mostly benign tumors. CT scan and MRI are complimentary methods in determining the nature of these types of lesions. We recommend the prophylactic removal of these tumors through surgery and primary skull repair when needed. Adequate exposure of the epidermoid cyst is needed to allow complete excision with the goal of preventing recurrences. The prognosis is good when the cyst is totally resected with its capsule.

References