Intradiploic Epidermoid Cyst Mimics a Bone Tumor: A Case Report

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Abstract

Intradiploic epidermoid cysts are cysts are a rare condition rare condition, located in the diploe of skull, caused by remnants of embryonic ectoderm. Most of intradiploic epidermoid cysts presented with small asymptomatic mass at head. We described the importance of considering such lesions in the differential diagnosis, especially when dealing with patients presenting with chronic sinusitis and airway obstruction. A 66-year-old male patient presented with history of chronic sinusitis and airway obstruction for few years. CT and MRI evaluation revealed an extra-dural well-defined mass at right temporal area of skull. Surgical resection was performed and histological examination has shown a cystic structure lined by squamous epithelium and containing laminated keratin material, which are consistent with epidermoid cyst. Because of variable radiological image findings, distinguishing epidermoid cyst from bone tumor and tumor-like lesion is difficult. Due to the rarity of this lesion, clinicians should be familiar with this disease. The total excision of the cyst along with its capsule is crucial to prevent the recurrence and potential complications.

Keywords: Intradiploic epidermoid cyst, scalp mass, cholesterol cleft

Epidermoid cysts are an uncommon lesion of intracranial territory which account for 1% of all cranial tumors.[1,2] The more common sites are cerebellopontine angle, posterior fossa and sylvian fissure. The less common type (intradiploic) is located in the skull. It is caused by a remnant of embryonic ectoderm within the skull bone or secondary to trauma.[3,4] This type commonly occurs in patients aged between 20-60 years old. The patients can present with long-standing, painless skull mass, headache, seizure, and focal neurological deficit. The imaging studies show bony defect with erosion of both inner and outer table of skull. The signal intensity is variable due to proportion of the keratin and cholesterol content. Histologic examination mainly reveals keratinized squamous epithelium with keratin lamellae, debris and cholesterol clefts, but devoid of skin appendages. The gold standard of treatment is complete surgical resection, given the varieties of differential etiologies in this region, it is essential to diagnose early and accurately. Here, we report a case of intradiploic epidermoid cyst that mimics bone tumor.

Case Report

A healthy 66-year-old man was referred to our hospital due to an incidental finding of right scalp mass by Computerized tomography (CT) scan. He had a history of chronic sinusitis and airway obstruction for a few years and had been treated with Fluimucil, Bromhexine and Cetirizine. No clinical history of fever, headache, visual disturbance, or neurological deficit. No history of cancer in the family or head injury. Physical examination revealed painless firm mass at right temporal area of skull. Reviewed CT scan showed bony defect of the mass involving both tables of temporal bone (Fig.1). The mass was an extra-dural well-defined mass, 3.8x4.3x4.5 cm in size, which showed mixed hypo-to-hyperintensity on T2-weighted magnetic resonance imaging (MRI) without gadolinium enhancement. There was a hemorrhagic component with solid enhancing component. Other intracranial structures such as nasopharynx and paranasal sinuses were not remarkable. The pre-operative differentials were aneurysmal bone cyst and giant cell tumor of

bone. The right fronto-temporal craniectomy with cranioplasty was performed and the intra-operative findings revealed firm to rubbery mass with a pearly white to yellow content. The histological examination demonstrated flat, thin squamous epithelial lining and dense flake of keratin without other skin appendages (Fig.2), which was consistent with an epidermoid cyst. Post-operative course was smooth and the patient was discharged without any complication 5 days after operation.

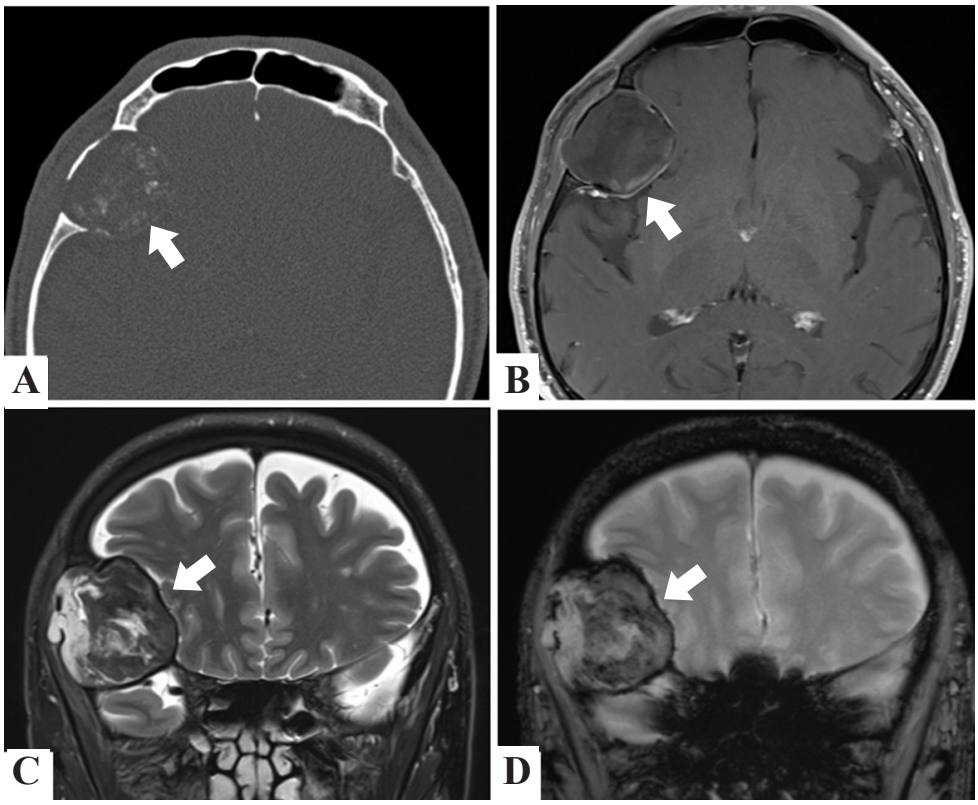

Figure 1: (A) CT scanning revealed a sharply demarcated bony defect(asterisk) and zones of calcification at the diploic space of the right sphenoid bone. (B) T1-weighted MRI with gadolinium demonstrated no contrast enhancement. (C) T2-weighted image showed mixed hypointense and hyperintense lesion. (D) Atypical internal hemorrhage on susceptibility image.

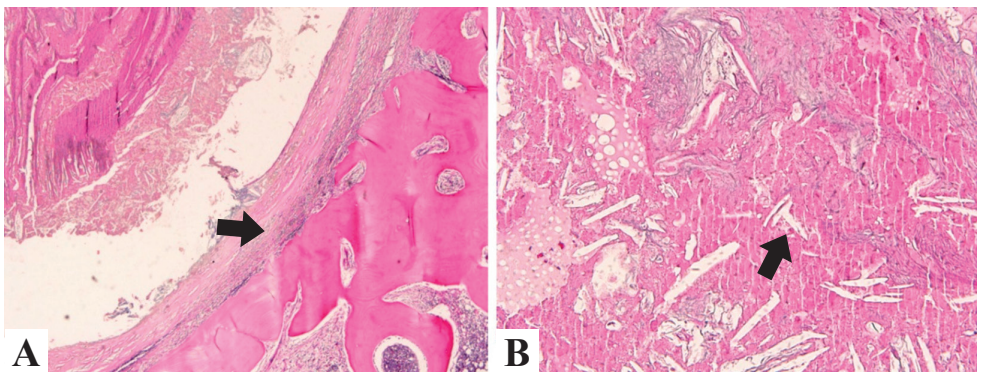

Figure 2: (A) A skull showed well-demarcated cyst lined with flat squamous epithelium(arrow). (B) The cystic content was mixed cellular debris, keratin flakes and cholesterol crystal(arrowhead).

Discussion

Intradiploic epidermoid cyst was first described by Muller in 1838.[5,6] These lesions were typically slow-growing masses found within the skull. They can occur at various locations on the skull and are more commonly observed in male patients.[7,8] However, malignant transformation into squamous cell carcinoma has been reported but in very rare cases.[9,10]

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Characteristic finding on CT scan were bony defect, expansion of both inner and outer tables of skull and sharp sclerotic border. MRI findings are variable from hypo to hyper intensity for T1 with mild enhancement, which depends on ratio between keratin content and cholesterol clefts.[11]

The definite diagnosis is typically established through pathological examination. For cholesterol granulomas, the key histological features include a cystic structure lined by keratinized squamous epithelium without skin appendages. The differential diagnoses are dermoid cyst, cholesterol granuloma, osteoma, aneurysmal bone cyst, giant cell tumor. The dermoid cyst has skin appendages such as sebaceous gland, sweat duct and hair follicle. On MRI, epidermoid cysts usually have well-defined borders, and they may not cause as much bony erosion as cholesterol granulomas. The histology of cholesterol granuloma typically reveals aggregation of foamy macrophages with or without hemosiderin pigments. The skull osteoma consists of thick hyperostotic lamellar bone. The aneurysmal bone cyst shows multiloculated blood-filled spaces intervened by fibroblasts, giant cells and woven bone. Osteomas typically appear hypointense on both T1-weighted and T2-weighted images. This is because they are composed of dense, compact bone. Unlike epidermoid cysts, osteomas are solid lesions composed of bone tissue and do not contain cystic or fluid-filled components. The giant cell tumor rarely occurs in the skull. MRI of giant cell tumor usually shows enhancement with contrast material.[12] The microscopic findings are tightly-packed osteoclast-like giant cells and neoplastic mononuclear cells with pale eosinophilic cytoplasm. It's commendable that the patient sought a second opinion and decided to undergo surgery for the skull mass. Complete surgical resection of the mass along with its capsule is indeed a crucial goal in the treatment of lesions like epidermoid cysts to prevent recurrence. Achieving a complete and meticulous surgical excision is essential for minimizing the risk of recurrence. Postoperative follow-up and imaging studies are crucial components of the management plan after the surgical resection of a skull mass, such as an epidermoid cyst. These follow-up measures are essential for several reasons. Regular and thorough monitoring, along with appropriate medical care, will contribute to the long-term well-being of the patient. Choi et al, reported that the recurrence rate of epidermoid cyst was between 1-54%.[13] Our patient was free of residual tumor by MRI at the 1-year follow-up.

Determining the exact cause of an intradiploic epidermoid cyst can be challenging, and in many cases, the etiology remains unclear.[14] Given the lack of a clear history of head trauma in this patient, it may be challenging to attribute the cyst to an acquired cause. The absence of a traumatic event does not rule out the possibility of an acquired lesion, as the exact mechanisms leading to the development of these cysts are not fully understood.

Conclusion

The diagnosis of intradiploic epidermoid cysts can be challenging due to varied radiological findings. The cysts can mimic bone tumors or tumor-like lesions, making it important for clinicians to consider this possibility when evaluating patients with skull masses. Surgical removal should be performed carefully to ensure complete removal of the cyst and to minimize the risk of complications. Given the rarity of intradiploic epidermoid cysts, it is important for clinicians to be familiar with this condition to ensure accurate diagnosis and appropriate management. Additionally, long-term follow-up may be necessary to monitor for any signs of recurrence.

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