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Chondroblastoma with secondary aneurysmal bone cyst in the anterior skull base

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ABSTRACT

Chondroblastoma with secondary aneurysmal bone cyst (ABC), especially in the anterior skull base, is an extremely rare condition. A 5-year-old boy presented with a large space-occupying lesion in the anterior skull base along with a left sided-epistaxis, proptosis and decreased vision. Radical excision of the lesion was performed by an endoscopic transnasal and transethmoidal approach. The patient recovered without any recurrence during a follow-up period of up to 28 months. Here, we review this rare case and discuss the clinical presentation and surgical treatment.

Introduction

Chondroblastoma is a rare lesion that typically presents in the epiphysis of the long bones. These tumors make up less than 1% of all primary bone tumors and 9% of all benign bone tumors [1]. Approximately 20% of these tumors occur in either the calcaneus or talus, and they occur almost exclusively in the epiphysis, and may extend into the diaphysis of long bones. About 10%–15% of chondroblastomas have an associated aneurysmal bone cyst (ABC). Chondroblastoma with secondary ABC in the skull and face has been rarely reported [2].

The diagnosis of ABC is difficult, because it is similar to other ossifibrous lesions such as ameloblastoma, giant cell tumor, myxoma, traumatic bone cyst and odontogenic keratocyst. Definitive diagnosis can be made only after biopsy. The usual methods of treatment are curettage, resection, intracystic injection of sclerotherapy and embolization [3]. It is still a considerable matter of debate which treatment option is the best choice, particularly in children.

We report a rare case of chondroblastoma with secondary ABC involving the anterior skull base in a five year-old boy, presenting with proptosis and headache. In this case, the tumor was resected completely by an endoscopical transnasal transethmoidal approach, under an image guidance system (IGS). There were no complications and the boy recovered well during a follow-up period of 28 months.

Case report

A five-year old boy was hospitalized because of nasal obstruction, rhinorrhea and headache for 1 month as well as a left sided epistaxis, proptosis and decreased vision for about 10 days before admission. There was no history of trauma. Physical examination showed the left sided proptosis, while the movement of the eye was normal. Endoscopic examination revealed a mass in the left nasal cavity, which easily hemorrhaged. Other neurological parameters following examination were normal.

Computed Tomography (CT) of the nasal sinus showed a soft tissue density lesion with multiple cystic cavities involving the ethmoid and sphenoid sinuses. The lesion extended to the bilateral orbit and anterior and middle skull base, where it led to bone erosion. Magnetic Resonance Imaging (MRI) of the nasal sinus showed that the lesion consisted of multiple cystic cavities extending to the medial orbit, the internal wall of the cavernous sinus as well as occupying the middle and upper meatus in the left nasal cavity. The lesion appeared as a "soap-bubble" (particularly in T2 weighted images) with contrast enhancement of the septa. The MR signal of the mass was low signal intensity on T1-weighted MR images and high signal intensity on T2-weighted MR images (Fig. 1).

The patient underwent surgery and the tumor was resected under hypotensive general anesthesia. A transnasal transethmoidal approach using rigid endoscopes, 4 mm in diameter and with a 0° lens was performed. The image guidance system (IGS) was set up before

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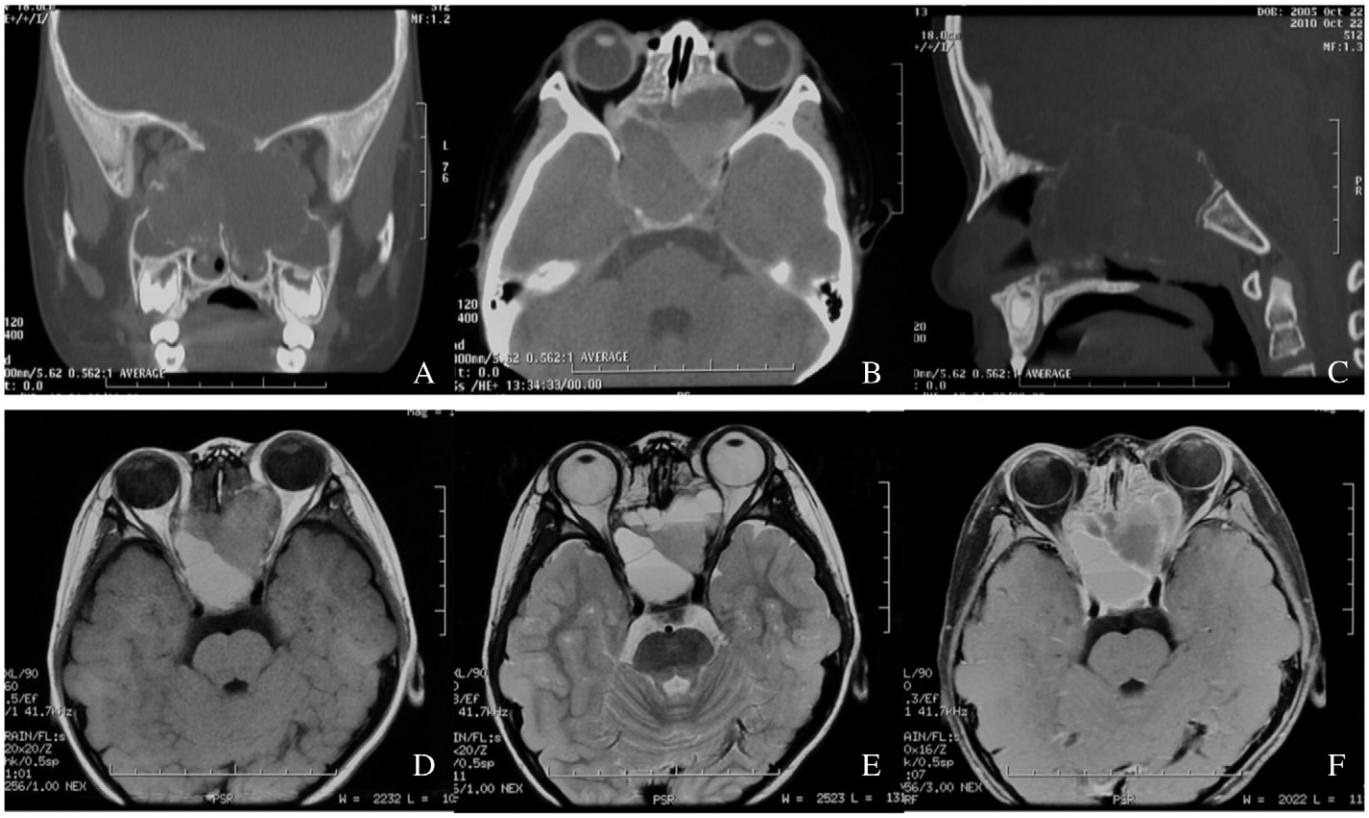

Fig. 1. Preoperative nasal sinus Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) scans showing a soft tissue density lesion with multiple cystic cavities involving the ethmoid and sphenoid sinuses, with an intraorbital and intracranial extension. The presence of the lesion is indicated by the "soap-bubble" features (particularly in T2 weighted images). (A) Coronal CT scan highlighted the mass as invasive as well as destruction of the orbital and arterial skull base. (B) Axial and (C) sagittal CT scans of skull. (D) Mass depicting two different signal intensities on the T1-weighted MR images. (E) T2 weighted MR images scan show the mass with "soap-bubble" features. (F) T1 weighted MR images with contrast show that the septum of multiple cystic cavities was enhanced.

the operation. As a first step, the nasal cavity was examined carefully using the endoscope before resection, to estimate the anatomic structures around the mass. Subsequently, a peripheral ostectomy of the bone around the mass was performed. A solid soft-tissue tumor was found. The fluid levels detected in the MRI and CT scans were confirmed to be blood sinuses and each of the cystic components contained a large amount of venous blood. The lesion was separated by pseudomembranes and bone trabeculae between cavities. To prevent blurring of the endoscope gauze packing was used to stop the bleeding. The mass was removed in a piecemeal fashion. When most of tumor was removed, the bleeding reduced. During the operation, IGS helped localize the critical anatomical structures such as the lamina papyracea, the skull base and the optic canal. The dura at the skull base and internal carotid artery canal were not involved but closely adjacent to the tumor. Finally, with the assistance of IGS, complete excision of the mass was achieved, leaving the meninges, the lamina papyracea and internal carotid artery intact.

Histological analysis showed that multiple cyst-like spaces were filled with erythrocytes separated by connective septa and infiltrated with multinucleated giant cells, spindle cells and fibroblasts (Fig. 2). These findings are characteristic of an aneurismal bone cyst. Scattered proliferating cartilage cells and chondroblasts were also observed. The pathological diagnosis was chondroblastoma with secondary aneurismal bone cyst.

On the first day after the surgery, the left sided proptosis was resolved and the headache relieved gradually. Postoperative MRI of the nasal sinus showed complete removal of the lesion (Fig. 3A). The patient was discharged from the hospital eight days after the operation. The patient recovered well during regular follow-ups spanning a 28 month period. CT of the nasal sinus showed no evidence of recurrence and normal rebuilding of the skull base (Fig. 3B and C).

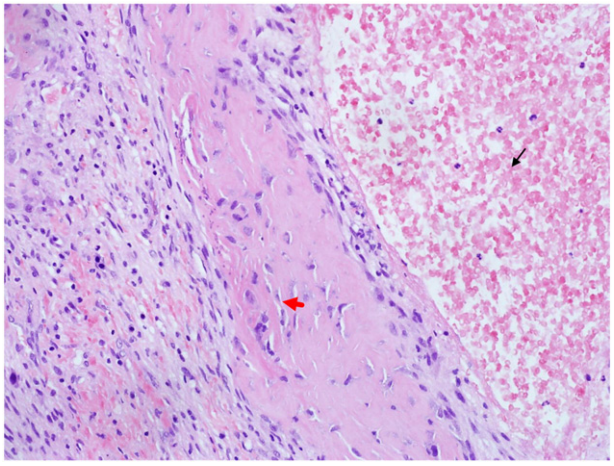

Fig. 2. Histopathological images of the lesion with multilocular cyst-like spaces filled with erythrocytes (black arrow) separated by connective septa and infiltrated with multinucleated giant cells, spindle cells and fibroblasts (red arrow) (H&E, ×400).