

Case Report

A Primary Ossifying Intracranial Myxoma Arising from the Ethmoid Sinus

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Myxomas are rare benign tumors that originate from mesenchymal tissue. They usually develop in the atrium of the heart, the skin, subcutaneous tissue, or bone. Involvement of the skull base with an intracranial extension is very rare and not well-described in the literature. We report a rare case of primary intracranial ossifying myxoma arising from the anterior skull base and mimicking a huge chondrosarcoma, and we review the relevant literature.

Key Words : Primary · Myxoma · Skull base · Neoplasm.

INTRODUCTION

Myxomas are rare histologically benign tumors of mesenchymal origin, usually found in the heart. The left ventricle of the heart is the most frequent location, with the thigh and shoulder comprising other frequent soft tissue sites³⁾. These tumors are locally aggressive and can be recurrent if en bloc resection is not possible. Intracranial myxomas may have an embolic origin from underlying cardiac myxomas²⁾. Primary intracranial myxoma with huge calcification arising from skull base is extremely rare and not well-described in the literature. Small cases of intracranial myxoma have been recorded (Table 1)^{6-8,11)}. In this article, the histological and radiological findings as well as treatment options for a giant primary ossifying intracranial myxoma mimicking a chondrosarcoma will be briefly discussed, together with a review of the relevant literature.

CASE REPORT

A 50-year-old man presented with spontaneous abrupt onset of headache and vomiting, as well as difficulty in standing because of dizziness. This was preceded by a month-long history of progressive hyposmia and feeling generally unwell. Neurological examination revealed only right abducens nerve palsy without signs of other cranial nerve palsies or pyramidal signs. The patient was taking no medication and the rest of his exami-

nation was unremarkable. He had been referred to our hospital with a pre-diagnosis of huge intracranial bony neoplasm, such as osteochondrosarcoma. Computed tomography (CT) studies revealed a well-defined low density mass lesion with huge dense calcification in both frontal lobe and right ethmoid sinus (Fig. 1). Sphenoid and maxillary sinuses were intact. Magnetic resonance imaging (MRI) scans revealed a giant neoplasm, 8.0×6.7×5.4 cm in size, a well-defined multi-lobulated cystic mass lesion with large popcorn-shaped dense calcifications on the frontal lobe and right ethmoid sinus. After gadolinium contrast enhancement, multifocal enhancing foci could be seen in the cystic mass (Fig. 2). CT of chest, and electrocardiogram were normal.

The patient underwent extensive resection of the mass via a right subfrontal and transcortical approach with both-sided frontotemporoparietal craniotomy. The calcified bony part of the tumor was present in the frontal base. The mass was extradural but firmly attached to the dural surface at the frontal base with significant displacement of the falx. Part of the right frontal dura was defect and appeared to contain tumor spreading from the frontal base. The calcified neoplasm was removed piece by piece with a drill and rongeur. Jelly-like neoplasms that appeared to be cystic radiologically were dissected and enucleated en bloc. No underlying brain involvement was evident during the surgery. The defective dura was repaired with galeal tissue to prevent cerebrospinal fluid (CSF) rhinorrhea. The gross tumor was totally removed along with the adjacent dura except for the

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Table 1. Literatures review of primary intracranial myxoma

Authors	Age (years), sex	Symptoms	Location of tumor	Prognosis
Oruckaptan et al. ⁷⁾	34, M	Right facial weakness	Right temporal, middle fossa	Local recurrence, 3rd-years follow up
Nagatani et al. ⁶⁾	38, M	Deterioration of visual acuity	Posterior fossa	No recurrence, 4th-years follow up
Osterdock et al. ⁸⁾	17, M	Hearing loss, left	Left petrous bone	No mention
Yin et al. ¹¹⁾	27, M	Epistaxis, visual impairment	Right temporal, ethmoid	No recurrence, 6th-months follow up

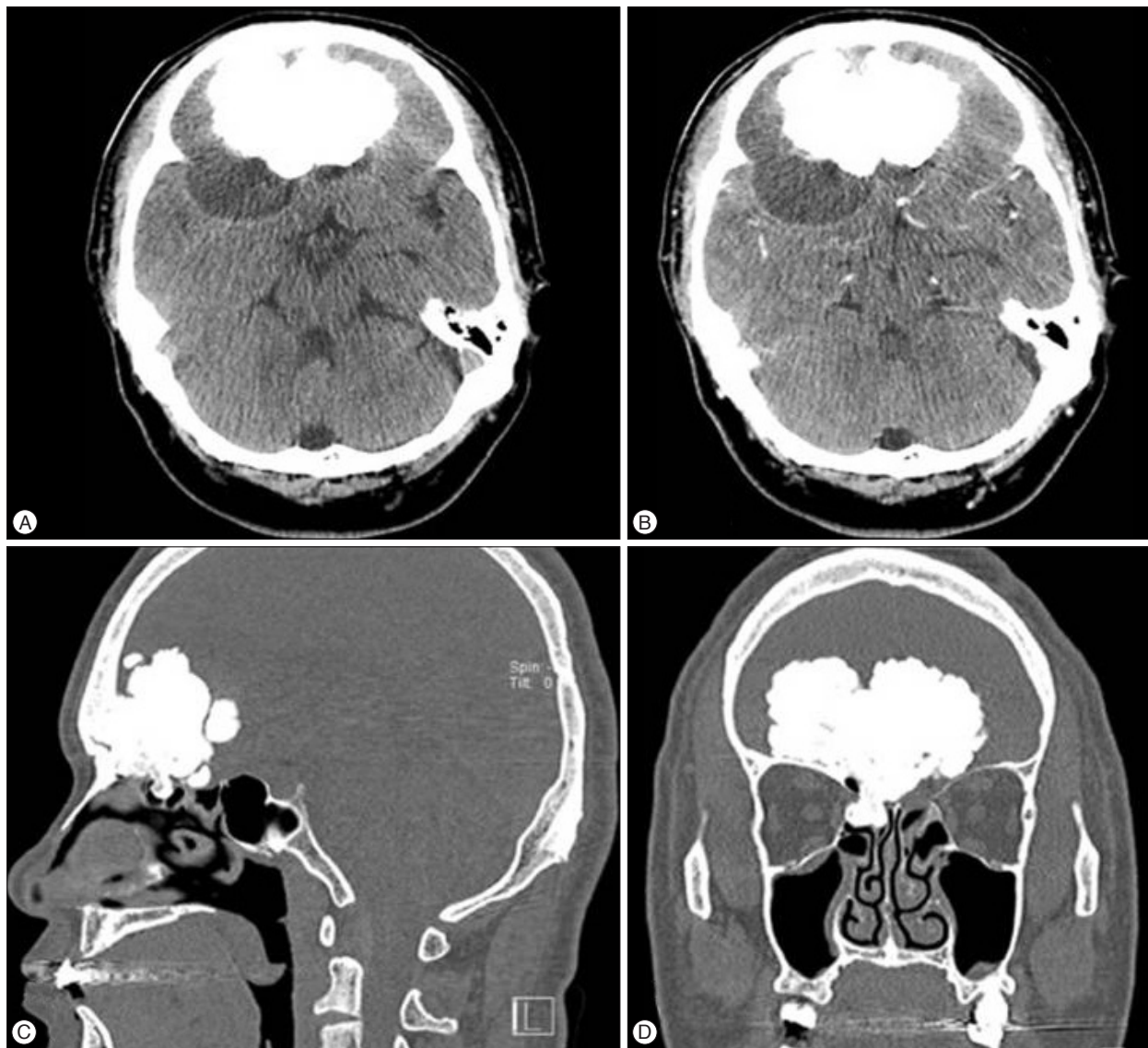


Fig. 1. Preoperative brain (A and B) facial CT (C and D) scans showing giant bony lesions arising from the right ethmoid sinus (C and D), and a low density, multi-lobulated mass with slight peripheral enhancement (A and B).

bony component inside right ethmoid sinus (Fig. 3).

Grossly, the surgical pathology specimens consisted of glistening yellowish soft-to-solid tissue with a large amount of bone (Fig. 4). Microscopically, they had a myxoid appearance with focal yellowish spots and a fibrous membrane. Immunohistochemically the tumor cells were positive for vimentin and negative for GFAP, S-100, EMA, CD34, and cytokeratin (Fig. 5). These findings are consistent with a pathological diagnosis of myxoma. The postoperative course was uneventful except for

transient frontal lobe syndrome due to a frontal lobe hematoma of the left hemisphere. After three-month the patient returned to work and will be followed-up every 3–6 months by MRI.

DISCUSSION

Myxomas are benign tumors originating in the mesenchyme. They occur equally in males and females and at any age⁶⁾ and in diverse organs including heart, skin, bone, and subcutaneous

tissues. Myxomas involving the bone of the base of the skull may originate from the primitive mesenchyme in the mastoid, sphenoid or ethmoid cells of embryos and newborns, and rare cases have been reported in the anterior cranial fossa^{6,9}.

Myxomas typically manifest as painless, very slow growing

masses in soft tissues or bones, and they may go undetected for several years¹. If they involve the skull base, as in our case, they frequently develop the signs and symptoms of cranial nerve palsies by the time of diagnosis¹². Myxomas and chondrogenic tumors usually arise from cartilaginous synchondroses at the

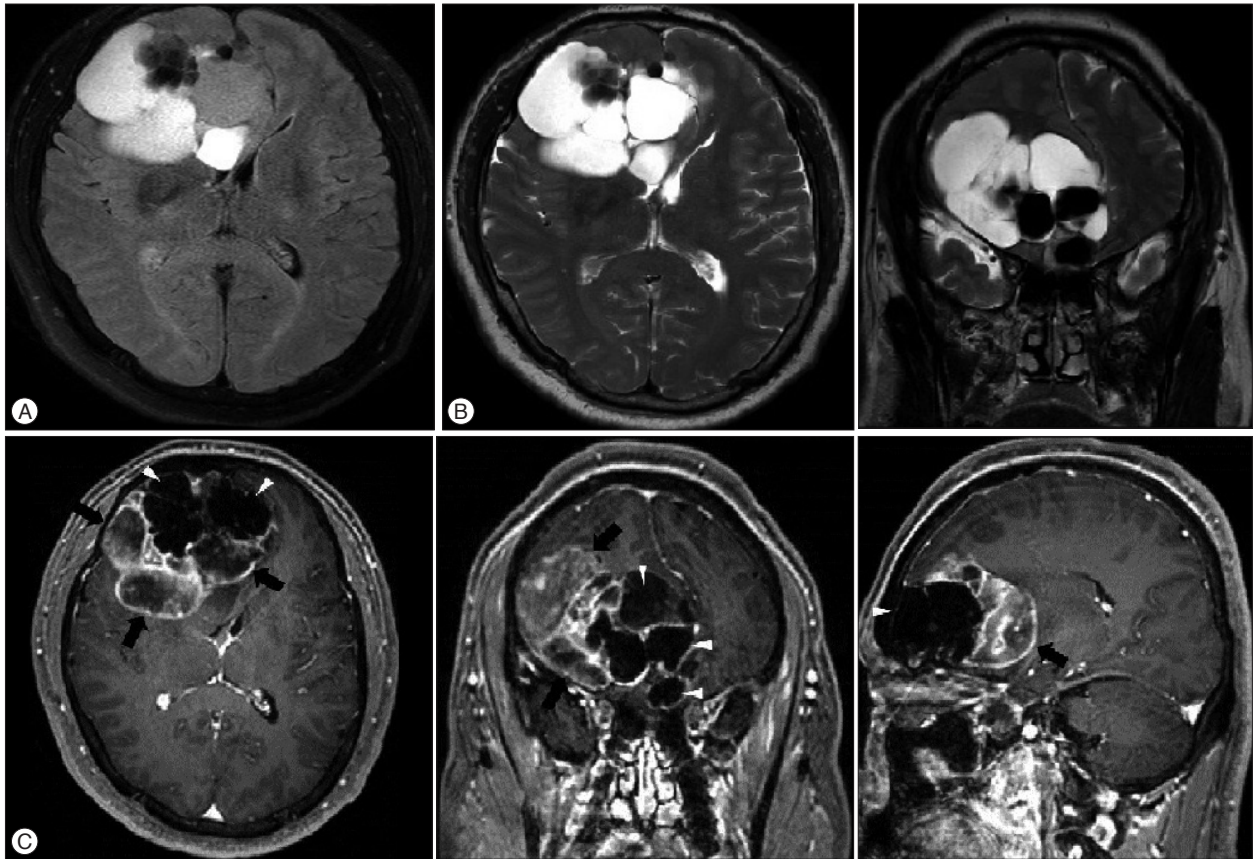


Fig. 2. Preoperative brain magnetic resonance (MR) images. A : Axial Flare MR image showing peripheral high signal lesion and multiple mixed iso- and low signal mass. B : Axial and coronal T2 weighted MR images demonstrating a high signal lesion and central low signal mass. C : Illustrating the giant low-signal lesion (white arrow-heads) and multi-lobulated hypo- or iso-signal mass (black arrows) with mild uneven peripheral gadolinium enhancement.

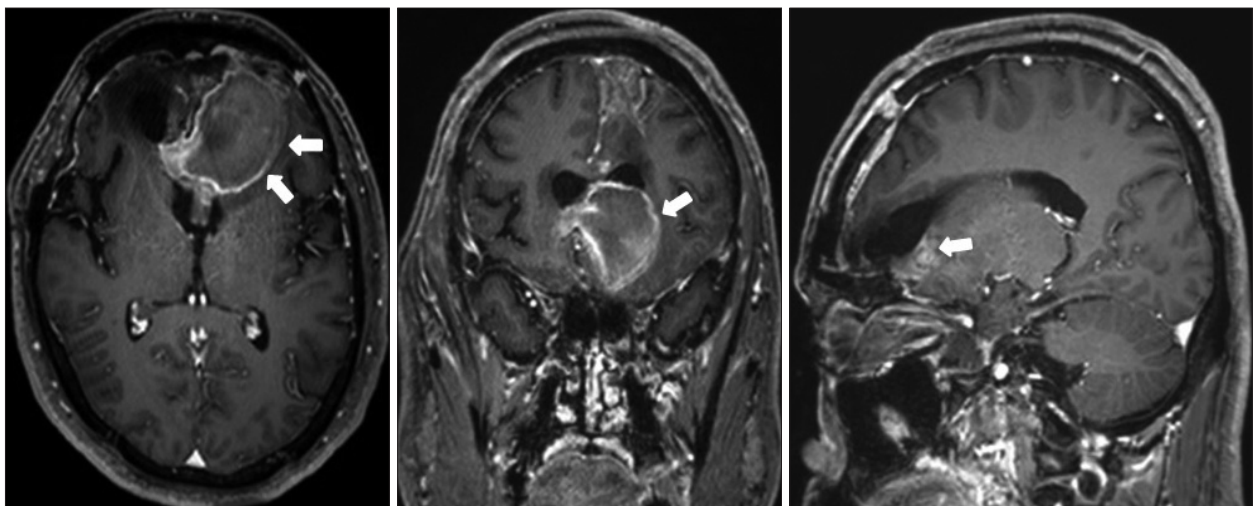


Fig. 3. Postoperative T1-weighted MR images with gadolinium enhancement in the axial, coronal, and sagittal plans show subacute hematoma at left frontal area (white arrows), total resection of the cystic lesion.

skull base. Unlike myxomas, chondrosarcomas are expansile, lobulated, soft tissue masses with endosteal bone resorption. In our case, the tumor was thought to be a chondrosarcoma radiologically. Intraosseous meningiomas and intradiploic epidermoid cysts are rare but do occur, and should be kept in mind in differential diagnosis⁷.

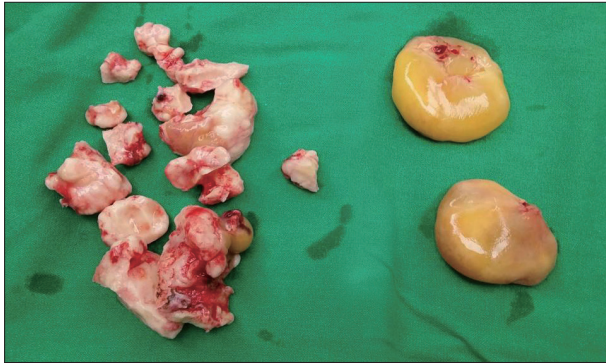


Fig. 4. Gross specimens showing the bony structure (left) and jelly-like glistening nature (right) of the resected myxoma.

Radiological examination is a decisive tool in the diagnosis of myxoma. CT scans can reveal bone destruction and relatively clear tumor margins. Myxomas may be hypodense to isodense in CT scans, and show variable enhancement patterns¹¹, and bone window imaging reveals the degree of bony destruction and the expansile pattern of the mass. More extensive bony growth than bony destruction was observed in our case, but this is a rare finding in myxoma.

MRI yields an intermediate or low signal intensity on T1-weighted images and a high signal intensity on T2-weighted images, and heterogeneous enhancement is a frequent finding^{2,12}. Radiologically, chondrosarcoma, chordoma, metastatic tumors of the skull, meningioma and epidermoid cysts of the dura and skull base are frequently encountered in differential diagnoses^{6,8,11}. Histologically, myxomas consist of characteristic hypocellular areas of satellite and spindle cells with a mucoid intercellular substance. The spindle and stellate cells are fibroblasts and myofibroblasts, and absence of nuclear pleomorphism is typical. Therefore, immunohistochemical staining can play a key role in differentiated diagnosis. Characteristically,

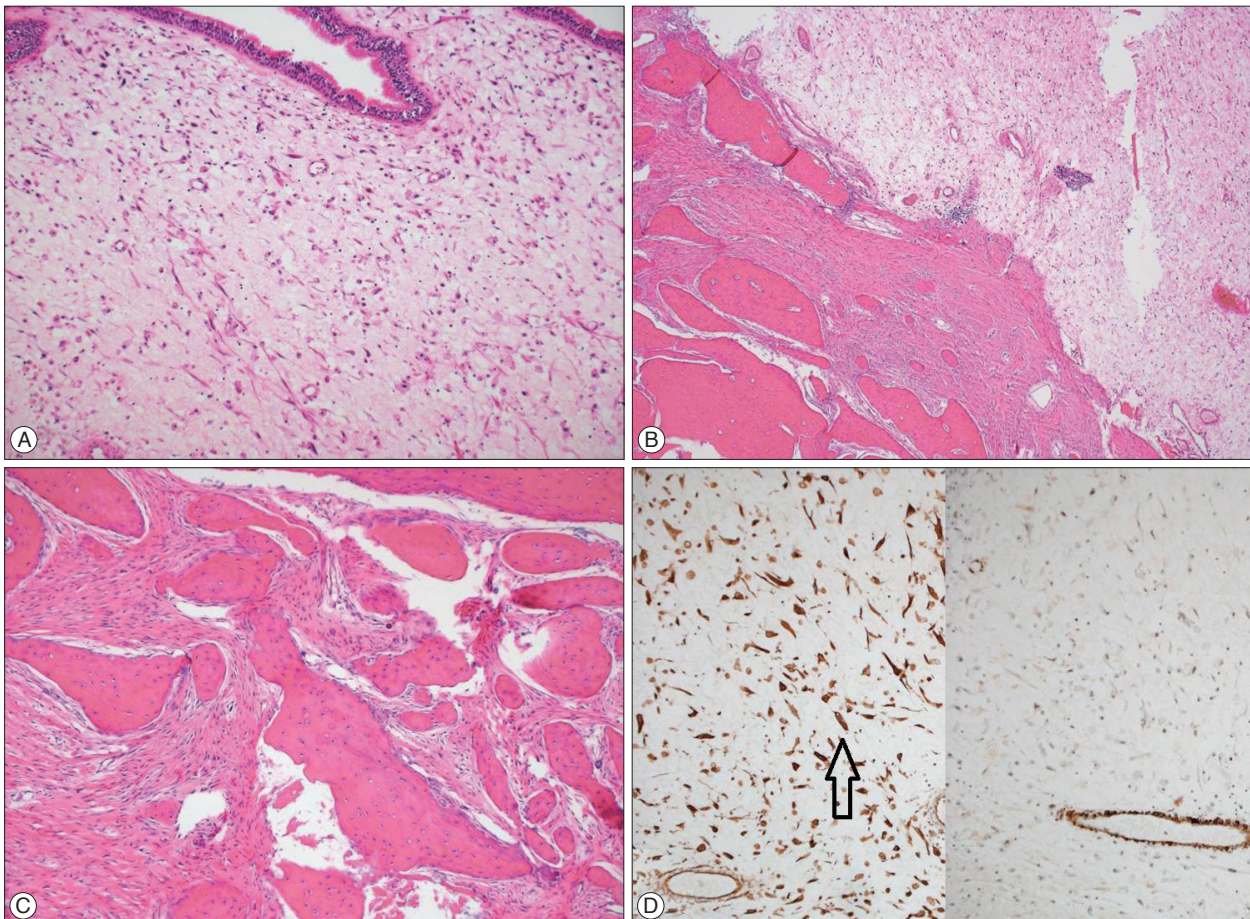


Fig. 5. Light microscopic and immunohistochemical findings. A : Tumor tissue is largely composed of haphazardly scattered stellate cells embedded in the loose myxoid stroma and is continuously lined by pseudostratified respiratory epithelium (H&E stain, $\times 100$). B : Tumor base shows prominent ossification with adjacent fibrous stromal reaction, which is abutting on myxoma area (H&E stain, $\times 40$). C : High power view of ossified fibrotic area (H&E stain, $\times 100$). D : Myxoma cells are positive for Vimentin (Left, Vimentin, $\times 100$, black arrow) but negative for SMA (Right, SMA, $\times 100$). SMA : smooth muscle actin.

myxomas stain positively for vimentin and negatively for S-100 protein, neuron-specific enolase, neurofilaments, glial fibrillary acid protein, keratin^{1,5,8,10}.

The treatment of choice for primary intracranial myxomas is radical surgical removal because they are generally insensitive to radiation therapy^{1,4,8}. However, a huge primary myxoma of the skull base is still a very challenging tumor, and very difficult to remove totally from the cranial base bone. After gross total removal of such tumors, reconstruction of the cranial base is also a problem. When the bones of the skull base are infiltrated deeply or widely, en bloc total resection is generally impossible. Even if it were feasible, the morbidity and mortality rate would be too high. In our case we opted for subtotal resection of the bony part of the tumor that had infiltrated the ethmoid sinus and it was removed by piecemeal resection, even though that approach was not very satisfying.

Most authors consider myxomas insensitive to radiation therapy^{1,4,8}. However, Zhang et al.¹³ reported one instance in which shrinkage of a myxoma of the skull base was seen at a seven month follow-up after gamma-knife stereoradiotherapy. Total removal of a myxoma originating from a bone of the skull base may be impossible, and radiotherapy may be a salvage treatment, though definitive evidence is not available. Recurrence is common, with 25% of tumors recurring if they cannot be removed radically. Recurrence has been noted as early as 3 months after surgical resection and as late as 10 years after surgical resection⁸.

We describe here a rare case of giant primary intracranial ossifying myxoma of the ethmoid sinus, review its histopathology, and emphasize that gross total resection, when possible, is the only definitive treatment.

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