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Imaging characteristics of the chondromyxoid fibroma of maxillary sinus

ABSTRACT

Chondromyxoid fibroma (CMF) is an unusual benign tumor of cartilaginous derivation. We present a young patient with resected and histologically proved CMF of the left maxillary sinus. Clinicopathological and radiological features with computed tomography (CT), magnetic resonance imaging (MRI) and digital subtracted angiography (DSA) images are presented.

Key words: Chondroma; Maxillary sinus; Magnetic Resonance Imaging

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INTRODUCTION

Chondromyxoid fibroma (CMF) is a rare, benign cartilaginous tumor that often occurs in the metaphyses of proximal tibia, proximal and distal ends of the femur and small bones of the foot. Tumors of the craniofacial bones are extremely rare and most often involve the mandible and the maxilla (1, 2).

Patients who have this very rare tumor are usually males between the age of 10 and 35 years. The tumor is slow growing and accompanied by mild symptoms (slowly progressive local pain, swelling, restriction of motion).

Radiographs show a lytic ovoid lesion with sharp sclerotic margins and pseudoloculated pattern resembling that of a bone cyst, eccentrically located.

Histologic findings include a strange mixture of fibrous, myxomatous, and chondroid tissues, of which the myxoid component is characteristic for chondromyxoid fibromas. Giant cells are commonly present. The conversion of CMF

to chondrosarcoma is extremely rare (3).

CMF is quite aggressive locally, especially in children. It has a tendency to recur locally unless completely excised, otherwise with simple curettement and bone grafting, the recurrence rate is about 25% (3).

CASE REPORT

A 9-year-old boy was admitted to the Ear-Nose-Throat department with painless swelling of the left cheek and signs of the fracture of the anterior wall of the left maxillary sinus. A month before that, he had suffered trauma of the left cheek during playing. Clinical examination showed partial fracture of the left maxillary tuber followed by intraoral fluctuation in the region of fornix. Control examination showed no signs of fracture healing with moveable maxillary tuber, which turned out to be pathologic fracture. Successive aspirations of the left maxillary sinus were performed and venous blood was aspirated hence tumor of vascular origin was clinically presumed diagnosis.

Initial CT examination, with limited scanning protocol for evaluation of sinusitis showed 5cm expansive lesion of the left maxillary sinus with destruction of its upper and medial wall. Tumor extended into nasal cavity with initial destruction of lower nasal concha and eroded ipsilateral pterygoid processes. MR images were obtained with 1.5-T Siemens Magnetom SP63-

4000 (Siemens, Erlangen, Germany). The MR examination demonstrated polycystic tumour mass of smooth margins with epicenter at the level of upper pre-molars. Content of cystic components demonstrated fluid-fluid level with signs of precipitation.

DSA revealed irrigation vessels of the tumor originating from maxillary artery and venous drainage through the facial vein.

Hemi-resection of the left maxilla with gross total excision of the CMF was performed and endoprosthesis was successfully implanted.

Clinical follow-up during four months revealed no signs of tumour recurrence with undisturbed nutrition.

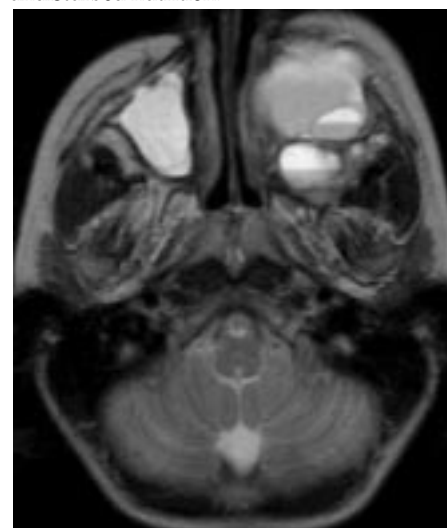


Figure 1. MR axial T2-weighted image with polycystic lesion expanding left maxillary sinus. Fluid-fluid levels can be seen within cysts

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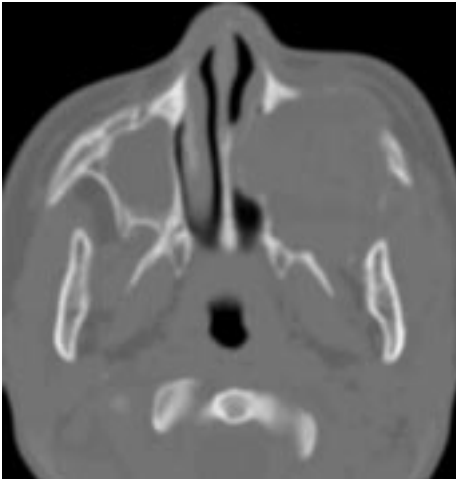


Figure 2. Correspondent CT axial image showed preformed thin layer of the anterior wall of the left maxillary sinus, destroyed medial wall and partially eroded pterygoid process



Figure 3. MR coronal T1-weighted postcontrast image with enhanced expansive mass involving the floor of the orbit, nasal cavity and cheek soft tissue displacement

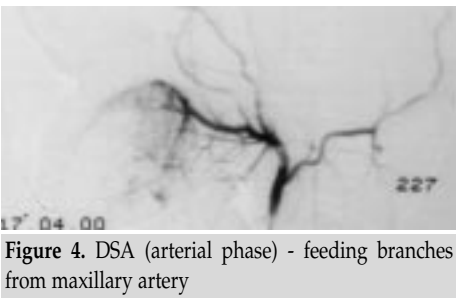


Figure 4. DSA (arterial phase) - feeding branches from maxillary artery



Figure 5. DSA (venous phase) - drainage through the facial vein

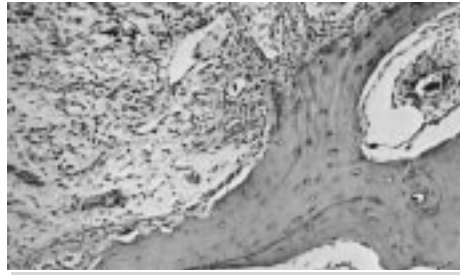


Figure 6. A photomicrograph of a CMF shows a vascularized periphery with stellate cells and a myxomatous background. A rather bland relationship between the tumor and the bone is seen. Haematoxylin-eosine staining, x112

DISCUSSION

Chondromyxoid fibroma (CMF) is the least common benign cartilaginous tumor, comprising less than 0.5 to 1% of all skeletal neoplasms with slight male predilection. About 75% occur in bones of lower extremities (4). The lesion is often greater than 5 cm in diameter at presentation. Differential diagnosis is wide and includes simple or aneurysmal bone cyst, giant cell tumor, nonossifying fibroma, fibrous dysplasia, enchondroma, chondroblastoma, eosinophilic granuloma and fibrous cortical defect (5, 6).

At the operation, the lesion filled the intracortical defect with myxoid soft tissue, bulging into the adjacent soft tissue. Microscopically, it showed typical features of chondromyxoid fibroma composed of mainly myxoid nodules and peripheral fibrous elements with focal chondroid differentiation. Because of pleomorphic appearance they can be confused with chondrosarcomas, giant cell tumors or osteosarcomas. The risk of a sarcomatous change independent of therapy is very infrequent. Chondromyxoid fibroma can, and should be distinguished from chondrosarcoma and chordoma, two tumors, which more commonly arise in the skull base and which have the potential to metastasize.

Our case demonstrates the uncommon occurrence of maxillary sinus CMF with nasal, pterygoid and orbital infiltration. When faced with an intracranial chondrocytic tumor, it is important to distinguish this neoplasm from enchondroma and chondrosarcoma.

MR images allow a detailed assessment of the soft tissue masses of the craniofacial region while CT offers superior analysis of bone struc-

ture involvement. The present case underlines the importance of MR examination in evaluation of soft tissue masses in the craniofacial region.

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