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Case Report

Myxoid chondrosarcoma of maxilla: A rare case report

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ABSTRACT

Chondrosarcomas are malignant tumors arising from cartilaginous tissues. Myxoid chondrosarcoma is rare histological variant of chondrosarcoma that usually occurs in limbs. Head and neck is an extremely rare site for this kind of tumor with very few cases reported till date. We hereby report a case of myxoid chondrosarcoma involving the maxilla of an adult patient.

1. Introduction

Chondrosarcoma is an uncommon malignant neoplasm of cartilaginous origin devoid of tumor osteoid with the evidence of fully developed cartilaginous structures. It is an extremely rare primary tumor of head and neck with less than 10% of the cases occurring in craniofacial region. The chondrosarcoma of the craniofacial region may arise from any bone, cartilage or soft tissue structures. In head and neck, the most common sites of origin are the maxilla, mandible, nasal septum, sphenoid sinus and the ethmoid sinus [1]. Myxoid chondrosarcoma is a rare histologic variant of chondrosarcoma, and is characterized by abundant chondroid matrix and malignant chondroblastic cells arranged in cords resembling chordoma [2,3]. Myxoid chondrosarcomas are typically located in the limbs in older patients, and only rarely originate in the head and neck [2]. We hereby report a case of myxoid chondrosarcoma involving the maxilla of an adult.

2. Case report

A 24-year-old male patient reported with fast enlarging bony hard swelling on the left maxilla. Extraoral examination revealed 3 cm × 4 cm firm lesion involving the left maxillary region with obliteration of nasolabial fold. Intra-oral examination revealed a 4 cm × 3 cm firm, lobulated lesion involving the left maxillary premolar and molar area with extension to both buccal and palatal side (Fig. 1). Marked mobility and displacement of upper left molars were also noted.

Contrast enhanced computed tomography was suggestive of a well-defined expansile lesion involving the entire left maxilla with spotty areas of radiopacity suggestive of calcification. Marked destruction of the floor of left maxillary sinus with extension into oral cavity and buccal space was also noted (Fig. 2).

Incisional biopsy of the lesion was taken. Histopathological examination of the hematoxylin and eosin stained section revealed superficial stratified squamous epithelium and underlying lamina propria. The deeper portion of connective tissue revealed multiple lobules of tumor cells in abundant myxoid stroma, separated by dense fibrocollagenous tissue. The section also shows areas of calcification. The tumor cells were polygonal, spindle as well as stellate shaped. In the center of lobules, the cells were loosely arranged excepting few discrete areas where they were arranged in nests or clusters (Fig. 3).

High power resolution revealed polygonal, spindle and stellate shaped cells with pleomorphic, hyperchromatic nuclei, scanty to moderate amount of deeply eosinophilic neoplasm. Cytoplasm was vacuolated at places, pushing nucleus to one side. Highly

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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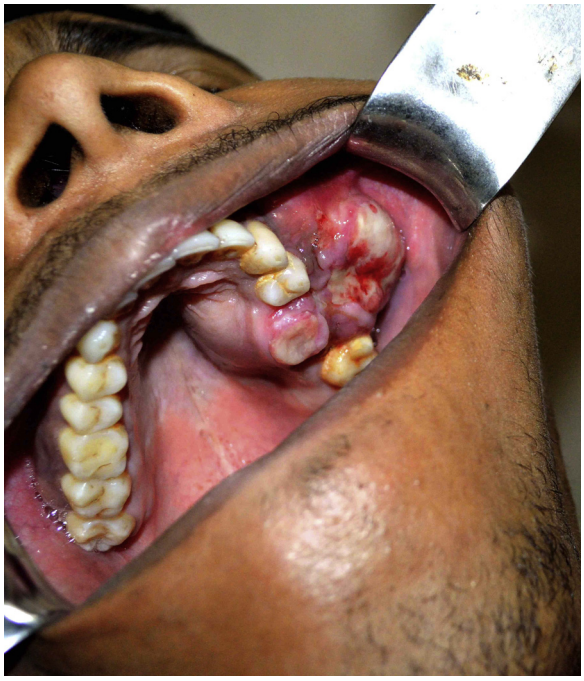

Fig. 1. Intra-oral view showing the extent of the lesion.

pleomorphic tumor cells with large bizarre nuclei and increasing mitotic figures were present adjacent to uncalcified matrix (Fig. 4).

Immunohistochemistry was done for confirmation of diagnosis. Tumor cells revealed strong positivity for S-100 and Vimentin (Figs. 5 and 6).

The case was diagnosed as myxoid chondrosarcoma as multiple malignant chondrocytes with cellular and nuclear pleomorphism, nuclear hyperchromatism were found in myxoid stroma.

The patient underwent left total maxillectomy after gaining access through the Weber–Fergusson incision with

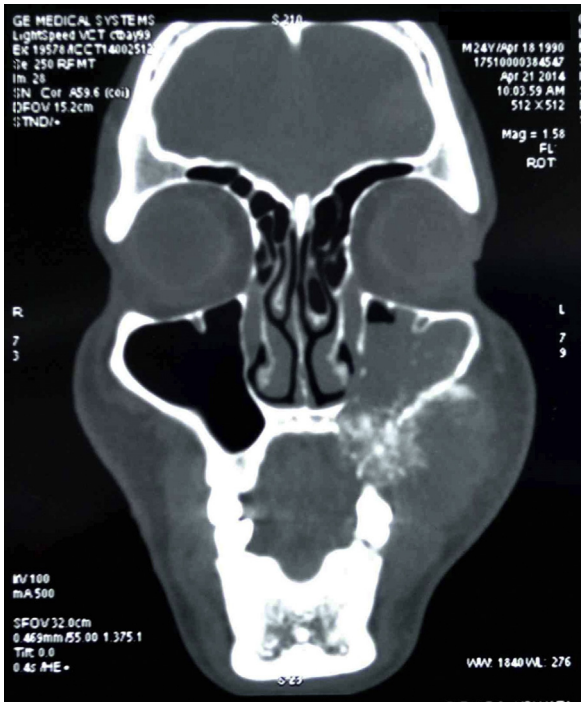

Fig. 2. Coronal computed tomogram showing the extent of lesion with spotty areas of calcification.

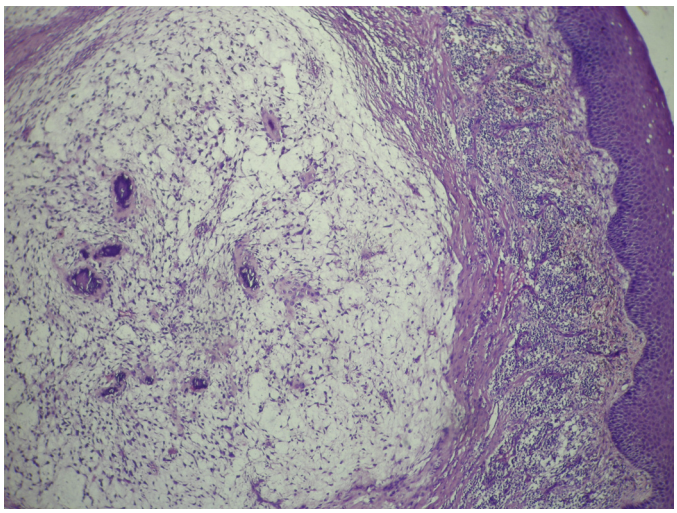

Fig. 3. Photomicrograph showing stellate shaped tumor cells in abundant myxoid stroma (H&E stain 100×).

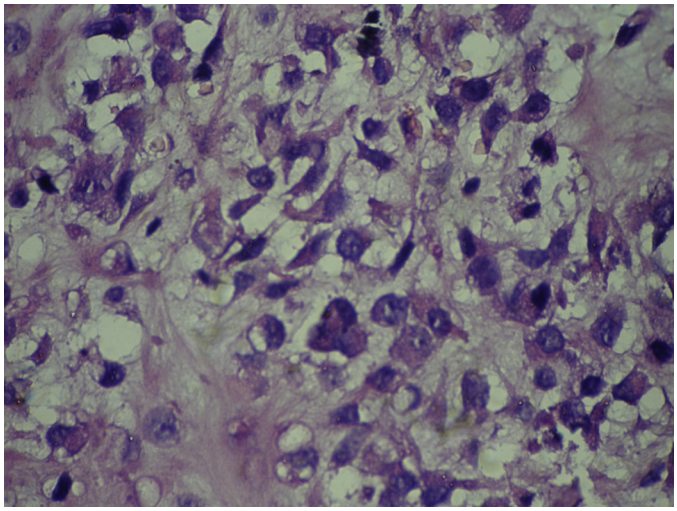

Fig. 4. Photomicrograph showing cytoplasmic vacuolation, abnormal mitosis and chondroid matrix (H&E stain 400×).

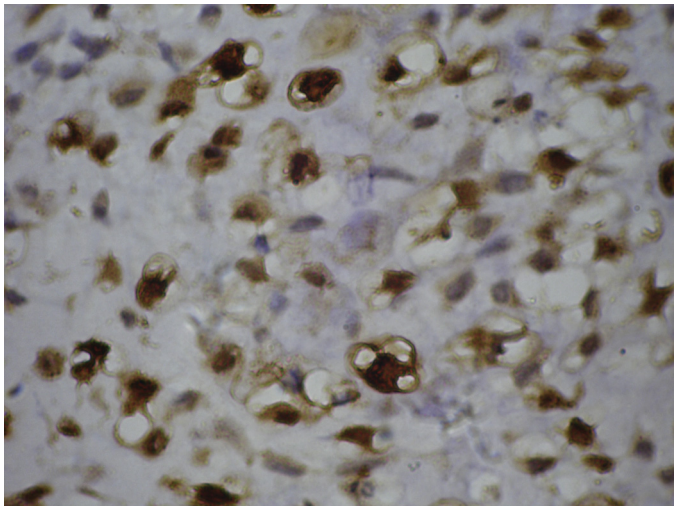

Fig. 5. Tumor cells were immunoreactive for S-100 protein.