CASE REPORT

Chondrosarcoma of maxilla

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Received: 02-04-2013
Accepted: 31-12-2014

ABSTRACT
Chondrosarcoma (CHS) is a rare malignant neoplasm of the jaws. Based on the morphologic features alone, a correct diagnosis of CHS may be difficult. Therefore, correlation of radiological and clinicopathological features are mandatory for the diagnosis of CHS. A case of CHS of the maxilla is reported. A brief discussion on the etiopathogenesis, radiologic and histologic presentation of the tumor and the treatment modalities of this unusual tumor is discussed.

Key words: Chondrosarcoma, maxilla, mesenchymal chondrosarcoma

INTRODUCTION

Chondrosarcoma (CHS) is a rare malignant tumor that produces cartilagenous matrix. The overall incidence of CHSs is 1 in 200,000 per year.[1]

Mesenchymal chondrosarcoma (MCHS) is one of the types of CHS. It was first described in 1959 by Bernstein and Lichtenstein as a distinct variant of CHS.[2] It usually appears in the second and third decades of life. It makes up less than 2–13% of all primary CHSs. This affects females more frequently than it does males (F/M = 1.4/1).[3] One of the most affected regions is the facial skeleton, especially the jaw. These neoplasms are characterized by sheets or clusters of highly undifferentiated, small, ovoid cells that alternate with small zones of neoplastic cartilage. The prognosis for patients with MCHS is unpredictable. This type of neoplasm shows aggressive local behavior as well as a high metastatic potential.[2]

In this report, we present a case of MCHS primarily involving the maxilla.

CASE REPORT

A 40-year-old female patient reported with a chief complaint of swelling in right side of face since 3 months. The swelling was initially pea-sized in upper right posterior region of jaw which gradually increased extending extraorally to a present size of 4 × 4.5 cm. Patient had initially consulted a local dentist about two and half months back. Medications were prescribed and radiograph was taken. Reports were normal with no evidence of bony erosion. Hematologic investigations were also done which were within normal limits. Patient gave history of paresthesia with right upper lip and right ala of nose since 2 months. Patient underwent biopsy from right maxillary posterior region 2 months back and histopathologic report was suggestive of a inflammatory lesion. Patient also gave a history of cataract of the right eye for which she had visited ophthalmologist 6 months back.

Extraorally, facial asymmetry was noted due to diffuse swelling on right side of face extending supero-inferiorly from right infra-orbital region to lower border of mandible [Figure 1]. Antero- posteriorly, the swelling extended from midline causing obliteration of right nasolabial fold to 2 cm anterior to tragus of right ear. The swelling was round to oval, same color as that of surrounding mucosa, smooth and tensed with well-defined margins and measured 6 × 4.5 cm in size approximately. The temperature was raised, it was soft to firm in consistency and non-tender. Lower right submandibular lymph node was palpable and enlarged to the size of 0.5–1 cm; was tender, hard in consistency, mobile and single in number.

Intraoral examination revealed expansile fleshy swelling on right maxillary posterior region of size 4 × 4.5 cm in relation with 13 to 18 involving right buccal mucosa, extending upto upper right gingivo-buccal sulcus involving the attached gingiva extending to the palatal gingiva and hard palate upto the midline. Obliteration of upper gingivo-buccal sulcus and labial vestibule was noticed on right side [Figure 2]. The swelling was yellowish white with red inflammatory marks representing the indentation of lower anterior teeth over labial and palatal mucosa. The swelling was dome-shaped over the

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Quick Response Code:
Website: www.jomfp.in
DOI: 10.4103/0973-029X.151338

Figure 1: Extraoral profile of the patient showing a large swelling on the right side of the face.

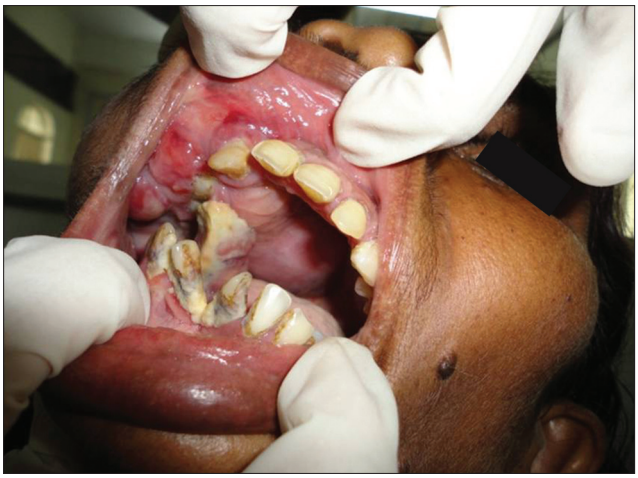

Figure 2: Intraoral photograph of the patient showing the lesion involving the right posterior gingivobuccal sulcus and the palate with displacement of teeth.

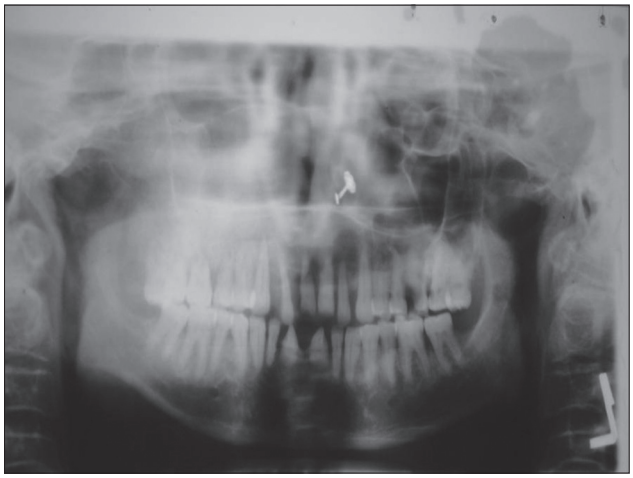

Figure 3: Orthopantomogram of the patient showing hazy radiopacity in the right maxillary posterior and maxillary antrum area.

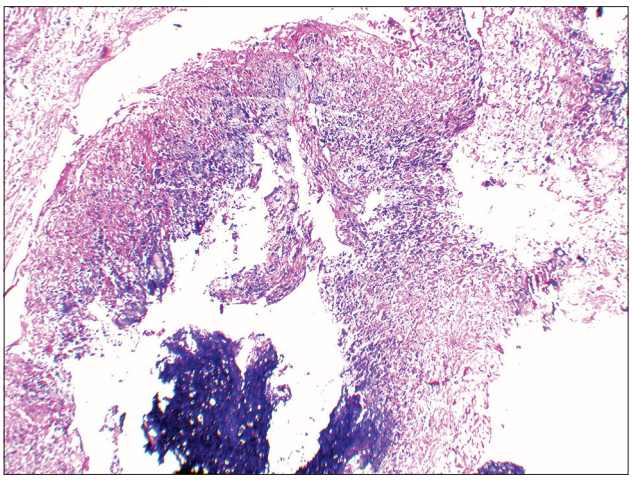

Figure 4: Photomicrograph showing cellular areas along with chondroid tissue (H&E stain, x40)

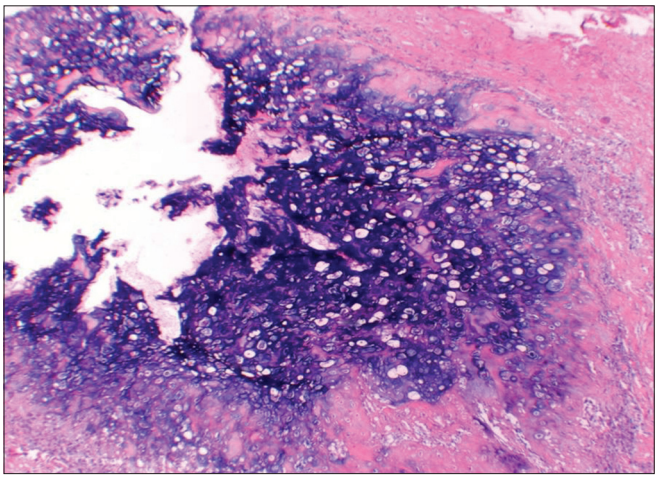

Figure 5: Photomicrograph showing areas of chondroid tissue (H&E stain, x200)

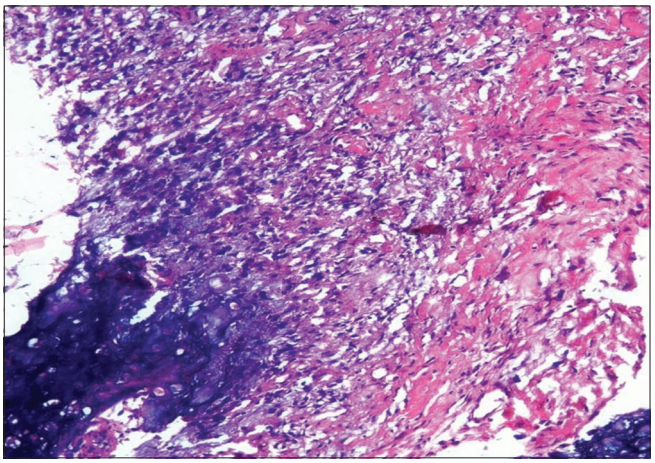

Figure 6: Photomicrograph showing a bimorphic pattern composed of lobules of cartilage and sheets of mesenchymal tissue (H&E stain, x200)

palate with smooth surface, ill-defined margins and was soft to firm in consistency.

Radiological examination by orthopantomogram showed severe inter-dental bone resorption with 11 to 16, cloudy

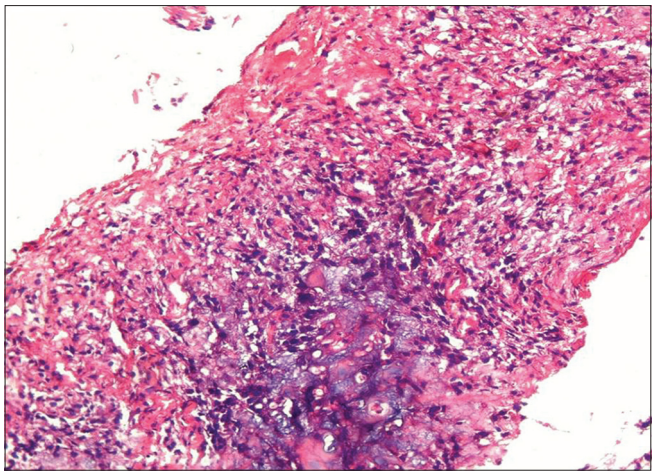

Figure 7: Photomicrograph showing connective tissue stroma with malignant chondrocytes and areas of highly cellular tissue composed of small round or spindle-shaped cells along with areas of calcification (H&E stain, x200)

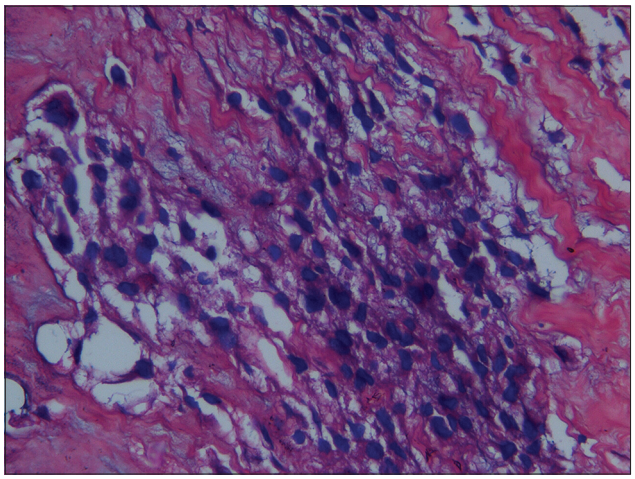

Figure 9: Photomicrograph showing small round or spindle-shaped cells (H&E stain, x400)

and hazy radio-opacity over right maxillary antrum with loss of demarcation of posterior wall and anterior tuberosity [Figure 3]. On the basis of clinical and radiological evidence, we made the provisional diagnosis of osteosarcoma, CHS, Ewing's sarcoma.

Under all aseptic conditions, biopsy from the respective site was taken and sent for histopathological examination. On gross examination, specimen consisted of multiple bits of tissue, soft to firm in consistency that were creamish in color. Microscopic examination revealed connective tissue stroma showing a bimorphic pattern consisting of sheets of small round or spindle-shaped mesenchymal cells, interspersed with islands of hyaline cartilage. The chondrocytes showed features of malignancy like nuclear hyperchromatism, pleomorphism, altered nuclear-cytoplasmic ratio and abnormal mitotic figures [Figure 4-9]. Also, areas of mineralization was seen which was confirmed by performing Goldner's stain [Figure 10].

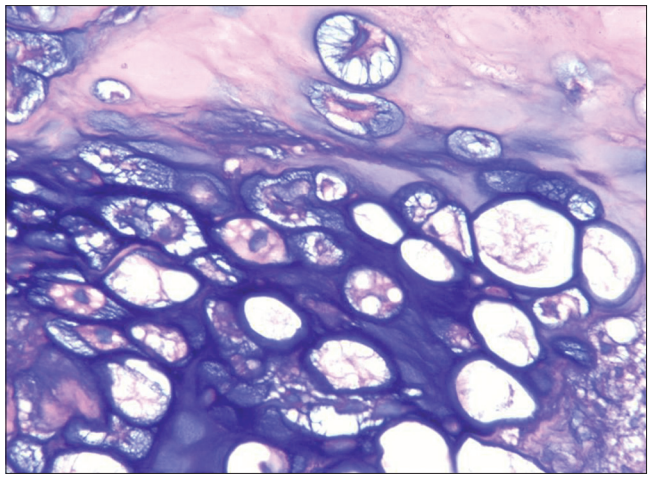

Figure 8: Photomicrograph showing malignant chondrocytes (H&E stain, x400)

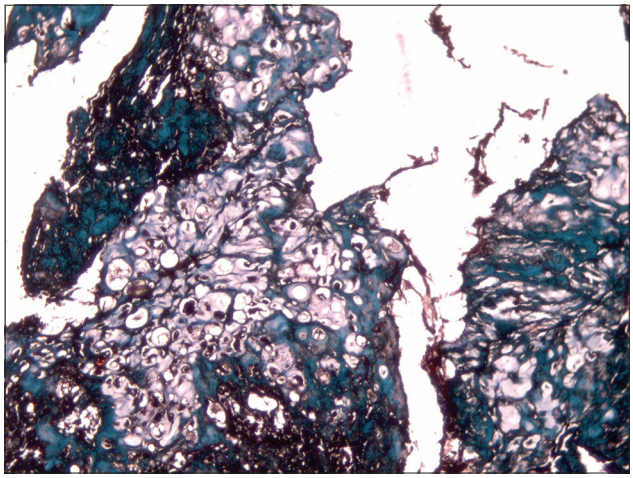

Figure 10: Photomicrograph showing areas of mineralization (Goldner stain, x100)

From the above histopathological features, the diagnosis of MCHS of maxilla was given.

The lesion was treated by radical surgery and follow-up of the patient was done periodically every 6 months.

DISCUSSION

MCHS is a rare highly malignant tumor that arises in bone but can occur in extraskeletal sites and is characterized by highly cellular areas composed of undifferentiated small round or spindle cells admixed with lobules of mature hyaline cartilage.[1]

CHS can be classified based on the topographical location of the tumor, the histological characteristics of the cancerous cartilage cells and the make-up of the surrounding matrix material associated with the tumor as:[1]
Juxtacortical CHS
Myxoid CHS

Mesenchymal CHS
Clear cell CHS
Dedifferentiated CHS.

The etiology of MCHS is unknown. Patients may have a history of enchondroma or osteochondroma. A small minority of secondary CHSs occur in patients with Maffucci syndrome and Ollier disease.

The pathogenesis and biologic behavior of the chondrogenic tumors is not fully understood, but it is evident that these lesions represent a spectrum from benign chondroma to the malignant CHS, through all degrees of intermediate type.[4] The exact origin of chondrogenic sarcoma in head and neck is controversial. It may be induced by irradiation, arise from pre-existing Paget's disease of bone or in association with fibrous dysplasia and the solitary bone cyst or it may arise from the vestigial cartilaginous rests. In premaxilla and maxilla, these cell rests are quite possible because of the proximity of chondrocranium throughout the fetal development. In mandible, the lesion may arise from the coronoid or condyloid process, from mental symphysis or from remnants of Meckel's cartilage.[5,6] Whereas, some believe that CHS can arise *de novo* from osseous tissues without the presence of cartilaginous rests.[7] Some authors believe that mandible is more common a site for CHS than maxilla.[6] While Terezhelmy[5] suggested that the tumor is found in equal frequency in both arches and Huvos has reported maxillary predilection.[8]

The most common clinical finding of mesenchymal CHS is painless swelling, expansion of buccal and lingual plates, premature eruption or exfoliation of teeth.[4] The mass is usually rapidly growing and covered with mucosa which can ulcerate and there can be pain at later stages. Rarely, there can be lymph node involvement. It can also cause nasal obstruction, nasal discharge or epistaxis, facial paralysis and bleeding from the lesion.

Histologically, CHS continues to be defined as a malignant tumor composed of fully developed cartilage without tumor osteoid, being directly formed from a sarcomatous stroma.[4] Myxoid changes, calcification and ossification may be seen. Evan's and co-workers have attempted to associate the histologic grade (grade I to III) of CHS with the ultimate biologic behavior of the tumor, depending on the cellularity, nuclei size, presence of mitotic figures, multinucleation, spindle cell formation and mineralization in the form of osseous development at the edge of the cartilagenous lobules.[9] In head and neck, the largest percentage of CHSs has been reported as grade I.[10] MCHS has a characteristic histological feature showing presence of highly cellular, undifferentiated zones with islands of chondroid differentiation as seen in this case.

The radiological pattern of CHSs is variable.[6] It includes single or multiple radiolucent areas. These lytic changes are prominent in more advanced cases. Other findings are opacification of air spaces, a densely calcified bone mass and root resorption.[5] Also, it may reveal ground glass appearance or a sunburst appearance. Cortical destruction occurs late in the course of disease and periosteal bone formation is often limited.[6] Some authors have reported a uniform widening of periodontal membrane space. In late stage disease, the primary lesion may penetrate the cortical plate and extend into adjacent soft tissues, resulting in a fuzzy soft tissue, peripheral shadow radiologically.[7]

The immunohistochemical markers that can aid in the diagnosis of chondroid tumors are Epithelial membrane antigen EMA, panCK, D2-40, S-100 and glial fibrillary acidic protein (GFAP). Chondroma typically shows positivity for EMA and panCK and negative for D2-40 and GFAP, whereas CHS revealed positivity for D2-40, S-100 and is negative for EMA, panCK and GFAP.

The MCHSs of maxilla are classically treated by radical surgery with radiotherapy being used as an adjunct or a form of palliative treatment for recurrent lesions. These lesions are radio-resistant and therefore radiotherapy is not generally recommended as a primary modality.[11] Chemotherapy is sometimes used for palliation and so is the role of radioactive sulfur.

The prognosis of MCHS of jaws is disappointingly poor as compared to that of long bones.[12] The cause of death is usually by direct extension into the base of skull and also through distant metastasis, chiefly to lungs and bones.

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How to cite this article: Gawande M, Swastika N, Chaudhary M, Patil S. Chondrosarcoma of maxilla. J Oral Maxillofac Pathol 2014;18:423-7.

Source of Support: Nil. **Conflict of Interest:** None declared.