Case Report

Clinicopathological and histological behavior of mesenchymal chondrosarcoma involving maxilla

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Abstract

Mesenchymal chondrosarcoma (MC) is a rare variety of chondrosarcoma (CS), which is both clinically unique and histologically distinct compared with conventional CS. Maxillofacial MCs are aggressive, have a tendency for recurrence and significant distant metastasis to lung and bone, and are associated with overall poor prognosis. Histopathologically, it is a biphasic tumor comprising of islands of hyaline cartilage with undifferentiated small round cells. Here, we present an interesting case of MC involving the left half of maxilla along with a brief review of the relevant literature.

Keywords: Biphasic, chondrosarcoma, hyaline cartilage, mesenchymal

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INTRODUCTION

Chondrosarcoma (CS) is an uncommon, slow enlarging malignant neoplasm that originates from primitive cartilage forming mesenchyme, and produces a pure hyaline cartilage that results in abnormal bone and/or cartilage growth.[1] It is the second most common primary bone cancer after osteosarcoma.[2] The involvement of the craniofacial region is very rare and accounts for <10% of all CSs.[2] Different types of CS have been described as follows:[3]

- Conventional CS-which accounts for nearly 90% of all CSs
- Dedifferentiated CS
- Clear cell CS
- Mesenchymal chondrosarcoma (MC)
- Juxtacortical CS
- Secondary CS.

MCs are relatively rare malignancies of bone and soft tissue with cartilaginous differentiation.[4] Fewer than 2% of all CSs are MCs. The maxilla and mandible are the most common sites of involvement, followed by the vertebrae, the ribs, the pelvis and the humerus.[3] It was first described by Lightenstein and Bernstein in 1959 as a biphasic tumor, comprising spindle cells interspersed with areas of chondroid differentiation.[5] Usually, MCs of the head and neck region arise from the maxilla, with relatively few arising from the mandible.[6] Females are more frequently affected than males. It is commonly seen in younger age group compared with conventional CS.[7] The majority of MCs are asymptomatic, although some patients present with rapidly invasive asymptomatic swellings, resulting in loosening of the teeth and expansion of the cortical plates.[8] Radiographically, it is difficult to differentiate between MC from other cartilaginous neoplasms or

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osteogenic sarcoma. Thus, there is no characteristic radiographic appearance as such. Maxillary MC presents as a radio-opaque or mixed mass that commonly invades the maxillary sinus.[8] Histopathologically, MC is described as a biphasic tumor with areas comprising sheet like proliferation of spindle or round cells interspersed with areas of chondroid differentiation.[9] Surgical excision of all the involved structure with negative margins is the primary treatment of choice for MC. The role of chemotherapy and radiotherapy is unclear. Due to the chances of late recurrence, the overall prognosis for maxillary MC is poor. Metastasis may occur at a relatively high frequency.[7] Hence, periodic follow-up is essential. Based on the above clinicopathological, radiological and histopathological findings, a case of MC involving the left side of maxilla was diagnosed and is discussed herewith.

CASE REPORT

A 28-year-old female patient reported to the Oral and Maxillofacial Pathology Department of Guru Nanak Institute of Dental Sciences and Research, Kolkata, with a chief complaint of a swelling involving left half of her face for the past 7 months.

Extra orally, there was presence of a diffuse swelling involving left maxillary middle third region, extending from the left half of upper lip to lower border of left eye with obliteration of nasolabial fold, causing obvious facial asymmetry. On palpation, swelling was bony hard, nontender and associated with the marked expansion of the mid-facial region. Overlying skin was free from underlying structure. There were neither neurological abnormalities nor any regional lymphadenopathy. There was no sign of epistaxis and nasal obstruction.

Intraoral examination revealed a large, diffuse, erythematous, exophytic, multilobular swelling involving left maxillary region extending from 21 to 27 region with obliteration of left mucobuccal fold. Prominent vascular markings were also noted on the surface of the lesion. On palpation, the lesion was bony hard, nontender along with expansion of left buccal cortical plate. The regional teeth were mobile, and 23 was displaced [Figure 1].

Upper occlusal X-ray revealed the presence of a diffuse mixed radiographic lesion involving 21–26 region with characteristic sunray appearance. Computed tomographic view (CT scan) showed the presence of a large (3.5 cm × 4.5 cm × 4.8 cm) intraosseous lesion with "sun burst" appearance, involving left maxillary region along with intrusion of the lesion within left maxillary antrum

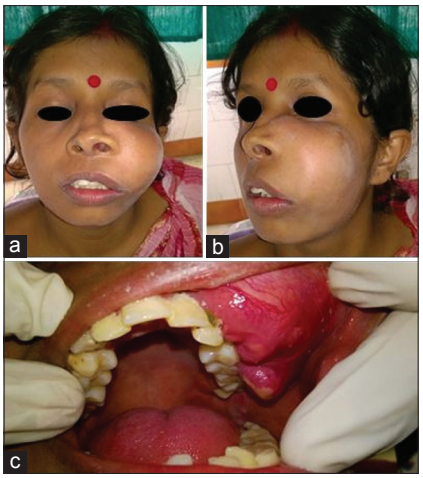

Figure 1: (a and b) Extra-oral photograph showing a diffuse swelling involving left maxillary region with obliteration of naso-labial fold, (c) intraoral photograph showing a large, diffuse, erythematous, exophytic, multilobular swelling involving left maxillary region with prominent vascular markings on the surface of the lesion

[Figure 2]. Routine blood investigations were within normal limits except serum alkaline phosphatase level which was increased to 195.7 U/L.

Based on the above clinical and radiological findings, a provisional diagnosis of osteosarcoma was made, and an incisional biopsy was performed under local anesthesia from the representative site. The light microscopic features revealed the presence of basophilic chondroid like material along with sheets of numerous small round cells on the superficial zone of the lesion. Cells of chondroid differentiation exhibited both nuclear and cellular pleomorphism along with nuclear hyperchromatism. Areas of partial calcifications and numerous slit-like vascular spaces were also observed [Figures 3 and 4]. The overall histopathological findings were suggestive of "Mesenchymal chondrosarcoma." The patient was advised for surgical treatment.

DISCUSSION

The present school of thought says that CS may be derived either from cartilaginous tissue reserve cells (primary CS) or from primitive mesenchymal cells displaying chondromatous differentiation. As maxilla is a bone of exclusive membranous ossification, so CS involving maxilla is derived from cartilaginous differentiation of primitive mesenchymal cells rather than from embryonal cartilaginous nests.[10]

Most CSs of the head and neck region occur in the maxilla. The other sites involved in descending order of frequency are the body of the mandible, the ramus, the nasal septum and the para nasal sinuses. The most common site in maxilla

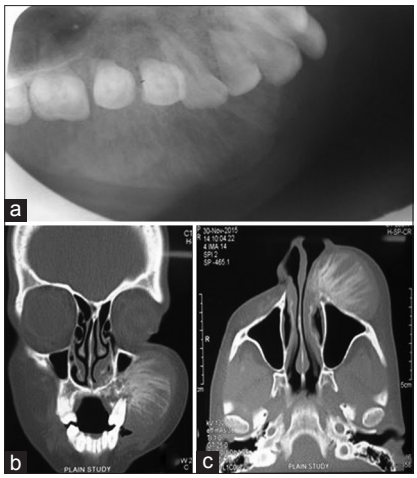

Figure 2: (a) Upper occlusal X-ray showing the presence of a mixed radiographic lesion involving 21–26 region with sun ray appearance, (b and c) computed tomography scan showing the presence of a large intraosseous lesion with "sun burst" appearance involving left maxillary region

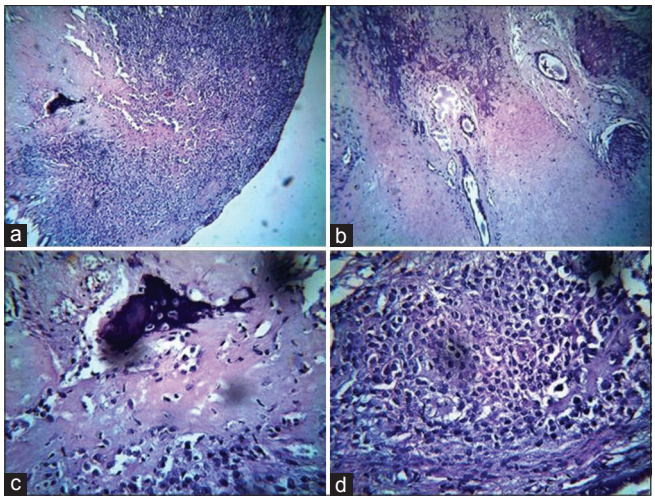

Figure 3: (a and b) Low power photomicrographs showing malignant chondroid matrix with partial calcification (×10), (c and d) high power photomicrographs showing malignant chondroid matrix with pleomorphic chondrocytes, some binucleate forms and calcifications (×40)

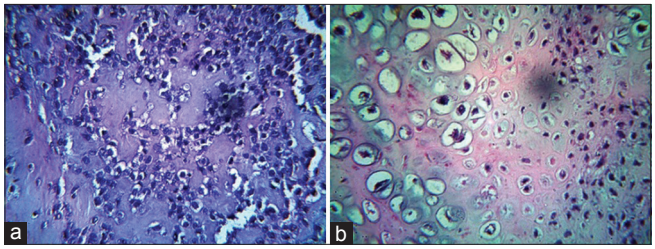

Figure 4: (a and b) High power photomicrographs showing chondroid areas with abnormal chondrocytes (×40)

is anterior portion of alveolus where preexisting nasal cartilage is present.[11] According to different literature, the MCs of head and neck reveal female predilection with 1.6:1 female to male ratio. It is frequently found in adults between second and third decade of life.[8,12] In a study of 36 MC of maxilla by Tien *et al*. the most common reported symptom was swelling/mass in 68% of cases, followed by nasal obstruction (32%), epistaxis (32%) and tooth mobility (24%).[8] The patient under discussion was a 28-year-old female having a diffuse, hard, lobulated swelling involving left maxillary region causing expansion of left buccal cortical plate and mobility of regional teeth. No regional lymphadenopathy, neurological abnormalities, or epistaxis were noted. Takahashi *et al*. indicated differences in the radiographic features between mandibular and maxillary lesions. Maxillary MC frequently presents as a radio-opaque or mixed mass that commonly invades the maxillary sinus. Mandibular MC appears as a radiolucent lesion with occasional calcifications.[13] Radiologically, the present case showed diffuse mixed radio-opaque and radiolucent lesion having "sun ray" appearance along with the involvement of the left maxillary antrum. The histologic appearance of MC characteristically exhibits the biphasic pattern. Undifferentiated areas appear as sheets of the primitive mesenchymal spindle or round cells. However, islands of relatively well differentiated cartilaginous tumor help in making a definitive diagnosis.[12,14] Calcification or ossification may occur within the chondroid matrix. Neoplastic cartilage may be replaced by bone in a manner similar to normal endochondral ossification.[8] Evans *et al*. have classified CS into Grade I, II, and III based on mitotic rate, cellularity and nuclear size.[11] Grade I lesions resembles benign cartilage and they do not metastasize. Grade II lesions demonstrate more myxoid stroma. They recur locally more often than Grade I lesions and have 10% incidence of metastasis. Grade III lesions have a more cellular pleomorphic appearance. They have spindle cell proliferation with a marked increase in the number of mitotic figures. The incidence of metastasis in these lesions is more than 70%.[15] In our case, histological features were similar to that of Grade III or high-grade CS. Radical surgical resection of the involved area is the most effective primary modality for the treatment of CS, as it is considered to be radioresistant. The reported 5 years and 10 years survival rate is 48% and 28%, respectively.[1] Death occurs usually due to direct extension of the tumor cells into the base of the skull and also through distant metastasis to lungs and bones. Hence, long-term follow ups are required for patients with MC involving maxilla.[7]

CONCLUSION

MC is a rare, aggressive jaw neoplasm with increased rate of recurrence and delayed metastasis. The clinical, radiological and histological features of MC is varied

leading to diagnostic dilemma and delay in treatment. Hence, oral pathologists play a pivotal role in recognizing this rare neoplasm.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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