

Mesenchymal Chondrosarcoma of Maxilla: A Rare Case Report.

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ABSTRACT

Chondrosarcoma (CS) is a malignant tumor of long and flat bone characterized by the formation of cartilage. Mesenchymal chondrosarcoma is a rare variety of chondrosarcoma. It is a biphasic tumor with areas of spindle cell mesenchyme along with chondroid differentiation in the connective tissue stroma. A 17 year old male presented to us as a painless mass in maxilla. Contrast enhanced computed tomography (CECT) showed a lytic expansile lesion in the right maxillary bone with foci of calcification within soft tissue lesion. Fine needle aspiration cytology (FNAC) and incisional biopsy was performed which confirmed the diagnosis of maxillary Mesenchymal chondrosarcoma. The patient underwent right subtotal maxillectomy with 2 cm margins. The review of literature shows that very lesser number of maxillary Mesenchymal chondrosarcoma cases were reported so far. Therefore an attempt is made to add this rare case of MC of maxillary alveolus in the Existing literature.

Keywords: Maxillary tumor, Fine needle aspiration cytology, Mesenchymal chondrosarcoma.

INTRODUCTION

Chondrosarcoma (CS) is a malignant tumor mostly identified by the formation of cartilage and generally found in flat and peripheral long bones.^[1,2] Its presence holds true for almost 10-20% of bone tumors.^[2] Head and neck CS is a rare type accounting almost 0.1% of head and neck neoplasia.^[3] Many variety of chondrosarcoma are documented including clear cell, dedifferentiated, myxoid and mesenchymal types.^[3-5] Mesenchymal chondrosarcoma (MC) is a rare variety that amounts to 3-9% of all CS and has high propensity for head and neck region. These are mostly seen in young adults in comparison to conventional Chondrosarcoma which is seen in old age groups.^[6] The tumor is rare and has been known for its aggressive nature with an inclination for late reappearance and metastasis.^[3,6] The review includes very few cases of maxillary MC which are reported so far.

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CASE REPORT

A 17 year old male [Figure 1] patient presented to Department of Oral pathology/oral medicine, Dr Z A Dental College Aligarh Muslim University with a chief complaint of a swelling in palate since 8 months. The patient was apparently well 8 months back when he noticed a small swelling appeared in palate one year ago which was excised with a report of osteoma but the swelling recurred 2 months later. As the lesion was asymptomatic initially the patient neglected it but as it continued to grow in size he became concerned and visited OPD for treatment. His medical history was non-contributory.

Extra Oral findings [Figure 1] were Gross facial asymmetry with right facial swelling, the lesion was soft on palpation. Intraoral examination [Figure 2] revealed A huge smooth surfaced erythematous swelling and involving whole palate and extending to right maxillary ridge with surface ulceration. Regional teeth are mobile. The skin over the swelling was smooth and appeared to be stretched with no surface ulcerations or raised temperature [Figure 1]. The swelling measured approximately about 4 x3 x2 cm in dimensions and caused gross buccal displacement of 13 and 14. The buccal swelling was soft to firm in consistency with mucosa over swelling smooth and stretched.

There was no significant lymphadenopathy associated with the lesion. FNAC from the lesion revealed chondroid matrix advocating diagnosis of chondroid tumor. CECT [Figure 3] and OPG [Figure 4] revealed a large osteolytic mass in the right maxilla with extensive new bone formation in

the right maxillary bone extending medially up to midline and laterally extending into gingivobuccal sulcus. There were foci of calcification within soft tissue lesion with no evidence of significant lymphadenopathy.



Figure 1: Obvious facial Asymetry.



Figure 2: A huge smooth surfaced erythematous swelling and involving whole palate and extending to right maxillary ridge.



Figure 4: (X RAY OPG) Radiolucency in the palatal region revealing a large osteolytic mass in the palate.

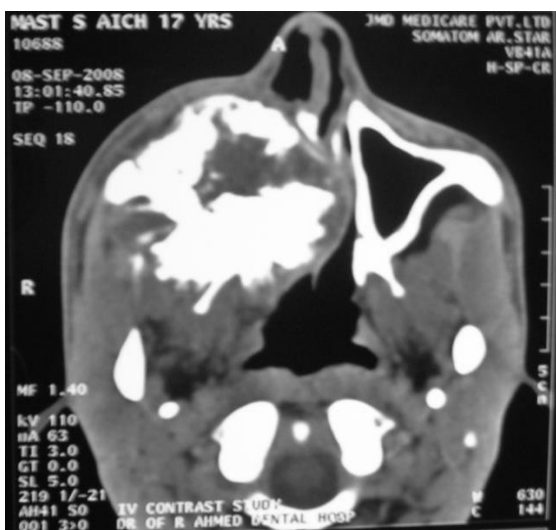


Figure 3: A large osteolytic mass in the right maxilla with extensive new bone formation in the right maxillary bone extending medially upto midline and laterally extending into gingivobuccal sulcus.

To confirm the diagnosis incisional biopsy was undertaken, a hard calcified tissue was seen and palpated which was later on included in the biopsy along with the soft tissue parts. Histopathologically [Figure 5] & [Figure 6] the picture reveals a cellular connective tissue stroma with spindle shaped cells arranged in lobular pattern. The cells showed dysplastic features with increased mitotic figures. Large areas of chondroid matrix with areas of calcification were also seen.

Hematological Findings were as follows:

Serum Alkaline phosphatase = 1340IU / lit (normal in adults = 100- 200 IU / lit)

Serum Ca = 8 mg/ dl(8.5-11)

Serum Phosphorus = 3.5 mg/ dl(2.5- 5 mg/dl)

Overall, histopathologic picture was suggestive of Mesenchymal chondrosarcoma of Maxilla. The patient was operated under general anesthesia and underwent right subtotal maxillectomy taking 2 cm margin. The palatal defect was closed with a medicated gauze beneath the palatal surgical splint. Post healing palatal obturator was placed to close

oro-nasal communication. The patient is under observation and after 5 year of follow up is doing well.

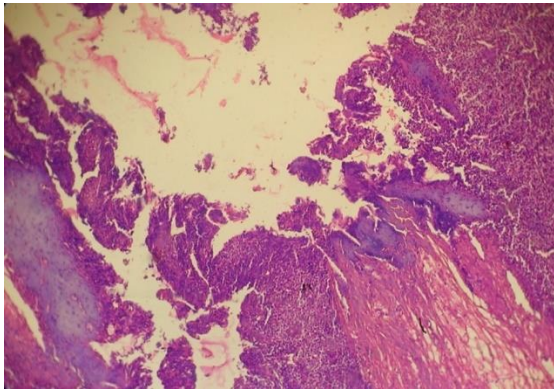


Figure 5: (10 X View) Connective tissue stroma with spindle shaped cells interspersed with chondroid areas.

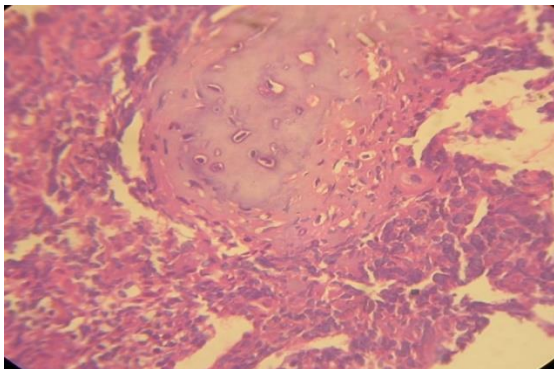


Figure 6: (40 X View) A cellular connective tissue stroma with spindle shaped cells arranged in lobular pattern. The cells showed dysplastic features with increased mitotic figures. Large areas of chondroid matrix with areas of calcification were also seen.

DISCUSSION

MC is an infrequent type of CS that was first documented in the year 1959 by Lichenstein and Bernstein as a biphasic tumor comprising of spindle cell mesenchyme interspread with areas of chondroid differentiation.^[3,5-8] Although it is included in rare tumours, the jaw bones are considered to be the most common site for MC.^[7-9] CS has been known to be a malignant tumor variety histogenetically originated from mature cartilaginous tissue.^[10] Although maxilla is a bone of mainly membranous ossification; the possibility of CS is mainly incidental and has been best attributed to the vestigial nests of cartilage with origin from.^[11]

1. Cartilage present in incisive papilla.
2. Foci of cartilage from cartilaginous nasal capsule
3. paraseptal cartilage.

The latest hypothesis proposes that Chondrosarcoma is originated either from cartilaginous tissue reserve cells or primitive mesenchymal cells

with subsequent chondromatous differentiation. Therefore it is assumed that CS maxilla is derived from cartilaginous differentiation of primitive mesenchymal cells and not from embryonal cartilaginous nests.^[10]

Most chondrosarcomas of head and neck area are found in the maxilla; followed by (in descending order) in the body of the mandible, the ramus, the nasal septum, and in the paranasal sinuses. In maxilla the most common site is front or anterior alveolus where already existing nasal cartilage is present.^[12-13] This case which we are reporting occurred initially in anterior part of maxilla but as the lesion grow it involved the posterior part of jaw as well.

The most common presentation of Mesenchymal chondrosarcoma of jaws is a painless mass or swelling on contrary to its occurrence in other parts of the body where pain and swelling are the most common presenting symptoms.^[6,12-15] In a study of 36 Mesenchymal chondrosarcoma of maxilla by Niven et al. the most common clinical presentation was a mass or growth in 68% of cases, followed by nasal obstruction (32%), epistaxis (32%) and tooth mobility (24%).^[7] Our reported case also have the chief complaint of a painless mass in the palate with grade I mobility of the involved teeth.

In the same study by Niven et al. they have shown that the period of signs and symptoms before diagnosis ranged from 2 weeks-4 years and in most of the cases did not go beyond a few months. Our case was presented and diagnosed after a period of 3 months.

The radiographic features of MC are not very classical. The tumor usually presents picture of a malignancy, characteristic of a radiolucent process with ill defined borders. The radiolucent part mostly contains scattered foci, which are formed by calcification or ossification of the cartilage matrix. It usually presents a typical picture of profound infiltration within the osseous trabeculae of the already existing bone without facilitating significant resorption. The lesion can cause symmetrical widening of the periodontal ligament of the teeth affected by the lesion.^[14] Our case presented with same radiographic picture of an ill defined lytic lesion with radio-opaque sclerotic foci in the maxillary alveolus without causing any significant root resorption.

The histologic appearance of MC is a characteristic biphasic pattern. Undifferentiated areas presented as sheets of primitive mesenchymal spindle/round cells as seen in small cell anaplastic sarcoma. These islands of well differentiated cartilaginous tumor generally assist in arriving at specific diagnosis (6). Calcification can be seen and can occur within the chondroid matrix. Neoplastic cartilage can be replaced by bone in a style similar to normal endochondral ossification. Microscopically speaking the lesion must be

distinguished from hemangiopericytoma, PNET, leukemia/lymphoma, Ewing's sarcoma, malignant melanoma, rhabdomyosarcoma and small cell osteosarcoma.^[7] Evans et al have classified CS into grades I, II and III on the basis of, cellularity, mitotic rate and nuclear size.^[12] Grade I lesions are similar to benign cartilage and they generally don't metastasize. Grade II lesions demonstrate more myxoid stroma. They recur more than grade I lesions and have 10% potential for distant spread. Grade III lesions present with cellular pleomorphic appearance. They have a spindle cell predominance with a significant increase in number of mitotic figures. The incidence of metastasis in these lesions is around 70%.^[1] The present case showed histologic picture of highly cellular connective tissue background of spindle cells with increased mitotic figures and interspersed with chondroid differentiation suggestive of grade III lesion.

Therapeutically many treatment modalities have been utilized. The best treatment is surgical resection with wider margins. The overall lesser incidence of metastasis may support the view that neck dissection is not mandatory.^[13,14] Radiotherapy generally plays a role when used in conjunction with surgery although MC comes under radio resistant tumor variety. Chemotherapy does not have much role in chondrosarcoma and should be used as an adjuvant therapy in those cases with aggressive behavior and having a propensity for metastasis, rapid local recurrence and high grade lesions.^[5,13]

Generally speaking the prognosis with MC is not very good because of the tumor's tendency for local recurrences and metastasis after a long disease free interval. The duration of around 20 years has also been reported before recurrence or metastasis occurred. The principal areas for metastasis are the lungs and bone. The reported 5 year and 10 year survival rate is 48% and 28% respectively.^[5]

The patient in present case was treated surgically with 2cm margins of resection and is doing well but since considering the aggressive nature of MC the patient is presently under a close long term follow up for the last 5 years.

CONCLUSION

MC is a unique and an infrequent mesenchymal tumor that arises frequently in both soft and hard tissues of the human body. These tumors display resident aggressive behaviour as well as great metastatic and recurrence potential. Owing to these, the prognosis is not very good. Bearing in mind the tendency of these tumors to metastasize and the poor outcome of patients with MC, early identification may permit earlier, more aggressive interventions and better results and quality of life.

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