MAVEN

CASE REPORT

MESENCHYMAL CHONDROSARCOMA OF THE MAXILLA-A RARE MALIGNANT CARTILAGENOUS TUMOR: REPORT OF TWO CASES

Dr. Debarati Mallick[1], Dr. Jay Gopal Ray[2]

[1]Assistant Professor, Dept. of Oral &Maxillofacial Pathology. North Bengal Dental College, [2]Professor and Head; Dept. Of Oral & Maxillofacial Pathol- ogy. Burdwan Dental College.

Corresponding Author: Dr. Jay Gopal Ray, Professor and Head; Dept. Of Oral & Maxillofacial Pathology. Burdwan Dental College.
E mail: jaygopalray@yahoo.co.in.

ABSTRACT:

Mesenchymal chondrosarcomas (MC) are rare malignant neoplasms that can arise from both soft and hard tissues. They are distinct tumors arising in unicentric or multicentric locations. They reveal unusual clinical behavior, characteristic histopathological features and poor prognosis with late recur- rences. Here is a report of two cases of 25 and 17 year old males with MCs affecting the maxilla. The importance of thorough evaluation and follow-up of the patient is emphasized. (2017, Vol. 01; Issue 01: Page 23 - 31)

Keywords: Mesenchymal chondrosarcoma, Chondrocytes, S-100 protein.

INTRODUCTION

Chondrosarcoma is a malignant tu- mor that arises from mesenchymal stem cells and undergoes a partial differentiation to form chondro- blastic differentiation and even de- finable cartilage. Chondrosarcomas of the jaws and facial skeleton are much rarer than in other bones presumably because of the scarcity of cartilage in development and in the joint areas. They are second to Osteosarcomas in their frequency as primary sarcoma of bone. Most chondrosarcomas will be seen ei- ther in the anterior part of the max- illa or in the posterior body region of the mandible. These areas of oc- currence have been postulated to arise from remnants of embryonic cartilage precursors from nasal septal development in the anterior part of maxilla and from Meckel's

cartilage precursors in the posterior aspect of mandible (1).

Mesenchymal Chondrosarcoma (MC) first described as a distinct en- tity by Lichtenstein and Bernstein in 1959, is a cartilaginous tumor of characteristic bimorphic appear- ance composed of sheets of primi- tive mesenchymal or precartilage cells and interspersed islands of well differentiated cartilaginous tis- sue (2). MCs develop from pluripo- tent mesenchymal stem cells and can differentiate into angioblastic, fibroblastic or cartilaginous struc- tures. Because of its prominent vascular pattern several cases re- ported in the earlier literature were initially interpreted as hemangi- opericytoma with cartilaginous dif- ferentiation. They arises from soft tissues or bone in the ratio of 1:2 to 1:6. These lesions affect females more commonly than males (4:1)

(3). It usually appears in the second and third decades of life. Any por- tion of the skeleton may be in- volved, but there is a definite ten- dency for involvement of the jaw bones and the ribs (4). When MCs occur in the jaws, maxilla was the more common site in one report; and in another, equal occurrence in both the jaws was reported. In the mandible, the most common site is preomolar and molar area, but the symphysis, coronoid and condylar process may also be involved. The prognosis of MC is unpredictable. The rare malignant cartilagenous neoplasm reveals aggressive local behavior and high metastatic po- tential (3).

In this report we present two cases of MC, primarily involving the max- illa & review its clinical presenta- tion, histopathology and treatment.

CASE REPORT

Case 1: A 25 year old male patient was referred to our Dept. for evalu- ation of a painless swelling in the right upper posterior region of the jaw for the past one and half months (Fig 1A). He gave a history of 17 extraction, 6 months ago due to caries and occasional bleeding from the swelling. Oral examination revealed the presence of a broad based, round, 4×3×2.5 cm sized swelling with smooth surface in the right upper posterior region of the jaw over the alveolar ridge. It was firm on palpation and 18 was mo- bile. Radiological examination (IOPA X-ray) revealed bone loss in 16 to 18 region (Fig 1B).

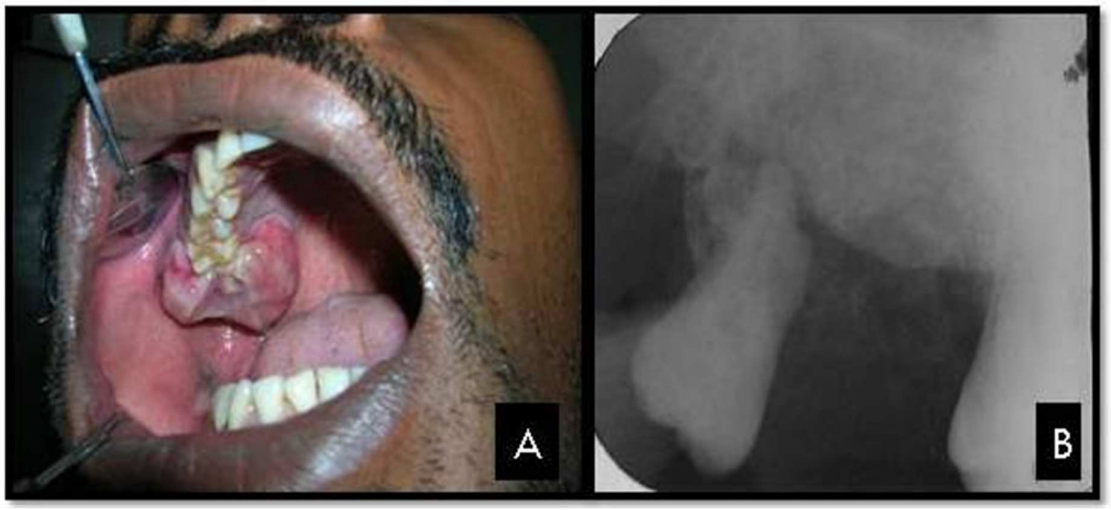

Fig 1: A- Intraoral photograph showing nonulcerated broad based swelling over the right maxillary posterior alveolar ridge, B- IOPA X-Ray showing bone loss.

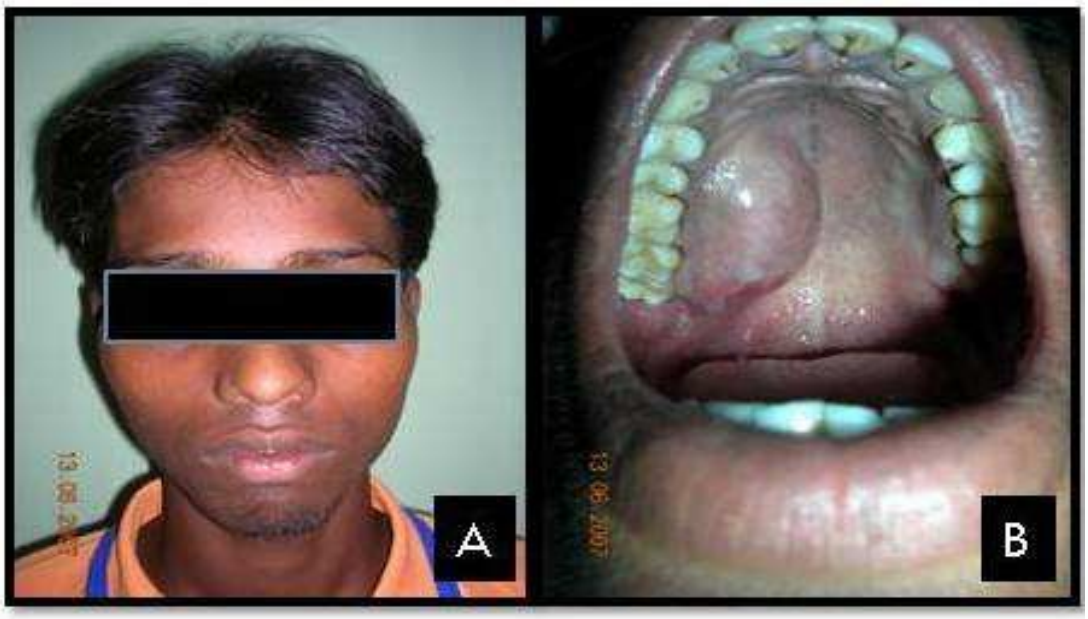

Fig 2: A- Extraoral photograph ten months ago, B- Intraoral photograph showing bony hard palatal swelling ten months ago.

Case 2: A 17 year old male patient presented with a gradually enlarg- ing large swelling inside the mouth for which he could not chew food or talk properly. His past medical his- tory was quite remarkable and in- teresting. Ten months ago he no- ticed a swelling in the palate for the past 15 days. The swelling was bony hard on palpation, present palatally opposite to 17,18 , approx- imately 3×3 cm in size, with normal mucosal covering. The lesion was excised and diagnosed as osteoma. The patient was relieved for 2 months, following which again the lesion appeared, increased gradu- ally and is now present for the past
8 months. Extraoral examination revealed obvious facial asymmetry with a massive swelling in the mid- third of the face. Intraorally a soft to firm swelling was present with ex- pansion of both buccal and palatal cortical plates in the right maxillary arch. It has involved whole of the palate crossing the midline. Two ar- eas of ulcerations are also noted, due to trauma from the opposing teeth. All the regional teeth were mobile & bilateral lymph nodes were palpable. OPG X-ray shows a dense irregular radiopacity in the right maxillary sinus region with periodontal space thickening of the regional teeth (Garrington Sign). CT scan revealed a large osteolytic mass with extensive new bone for- mation arising from the right max- illa, extended to the nasal cavity and ethmoidal sinuses causing de- viation of the nasal septum to the left side & proptosis of the right eye. Biochemical investigation reveals a high serum Alkaline Phosphatase level (1340 IU/L).

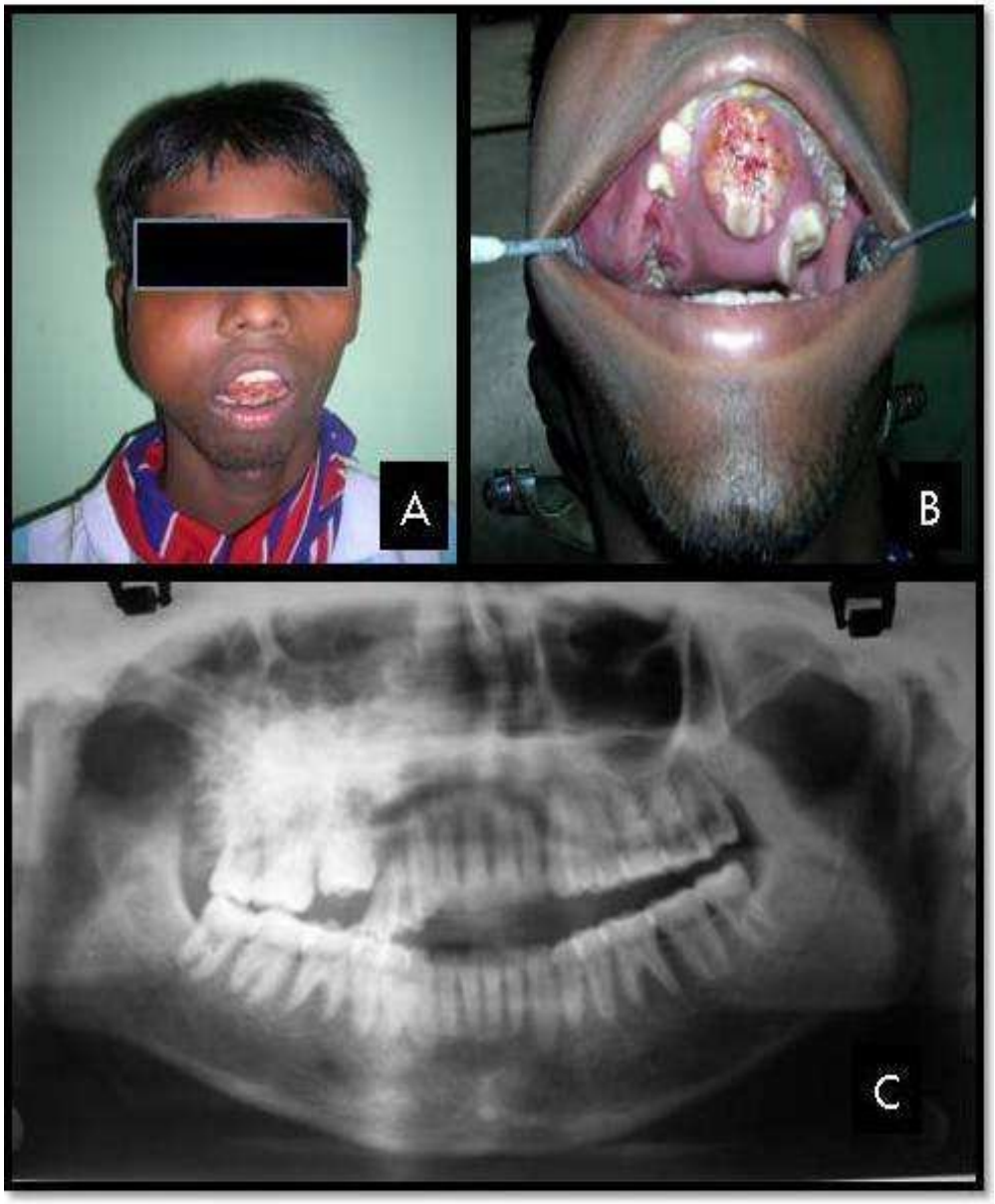

Fig 3: A- Present extraoral photograph with obvious mid-facial swelling, B- Present intraoral photograph showing buccal and palatal swelling with sur- face ulceration, C- OPG shows a dense irregular radiopacity in the right maxillary sinus region.

Incisional biopsy was advised in both the cases and histological evaluation revealed MC (Fig 4 and Fig 5).

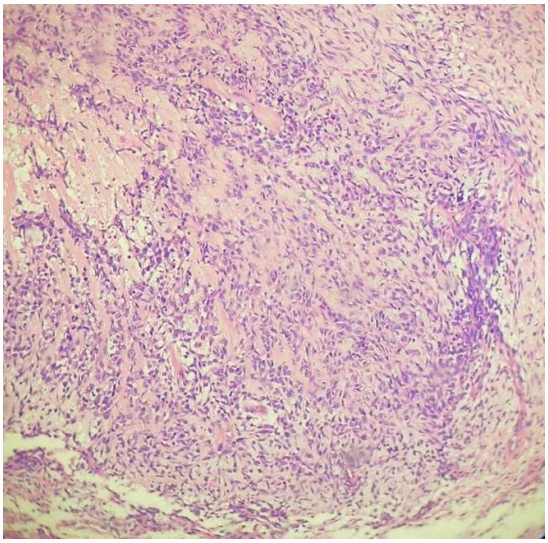

Fig 4: Photomicrograph showing primitive appearing mesenchymal cells with mildly pleomorphic nu- clei (H & E, 10X) (Case 1)

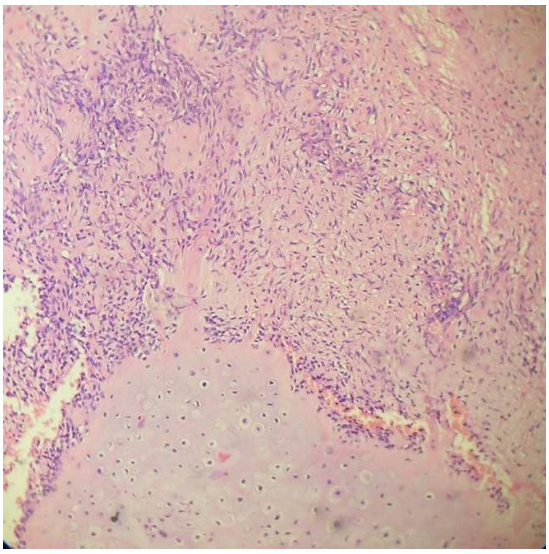

Fig 5: Photomicrograph showing mature chondroid areas with few atypical chondrocytes (H & E, 10X) (Case 2).

DISCUSSION

MC is a rare malignant cartilagi- nous tumor with a distinct histo- logic appearance and biologic be- havior (5). The head and neck re- gion is a relatively common location for MC, accounting for 20% to 30% of cases (6).

Clinical features: Although congen- ital examples of MC are reported as well as cases in patients older than 80 years of age, most patients are in the second or third decades of life with mean and median ages of 20 to 30 years. In the head and neck re- gion, the mandible and maxilla are the most frequent sites. Other sites include the orbit, nasopharynx, ethmoid sinus, maxillary sinus, parapharyngeal-tonsillar area, soft tissue of the face and neck, cerebel- lum, bones of the skull and the cer- vical vertebrae. The predominant symptom was usually a painless mass or swelling (53%) as in the present cases. However, painful mass (16%) has also been reported
(3). The duration of the symptoms is quite variable, with patients hav- ing symptoms for as long as 10 years or for only a few days or weeks (6).

In the first case, the perplexing his- tory of occasional bleeding, clini- cally benign appearance of the soft tissue mass and little bone loss misleaded us as to be a reactive le- sion. An attempt of excisional bi- opsy was thus made, which re- vealed unhealed areas even after two weeks.

In the second case with the history of previously diagnosed osteoma, massive bone enlargement follow- ing surgery, the biochemical re- ports of serum alkaline phospha- tase, a provisional diagnosis of os- teosarcoma was made.

Radiological features: The most common radiographic appearance of MC of jaw is a radiolucent osteo- lytic shadow and it is difficult to distinguish MC from other cartilag- inous or osteogenic sarcomas. The area of radiolucency is usually speckled with calcifications (3). In the second case presented here, ex- treme destruction of the jaw bone with the large radiopaque mass having sun-ray periosteal reaction and widened periodontal spaces aroused the suspicion of an osteo- genic sarcoma.

Histological features: In both the cases multiple H & E stained sec- tions revealed biphasic pattern of tumor consisting of islands of cyto- logically benign hyaline cartilage with surrounding hypercellular ar- eas containing small primitive-ap- pearing round and spindle shaped mesenchymal cells. Mitotic activity was variable. It is important to note that in contrast to some other forms of chondrosarcomas, cellular pleo- morphism and high mitotic activi- ties are usually not seen in MCs. In some areas, the cells had a pattern mimicking Ewing's Sarcoma. The transitional zone seen between the chondroid foci and the mesenchy- mal component was more gradual and not sharp (3).

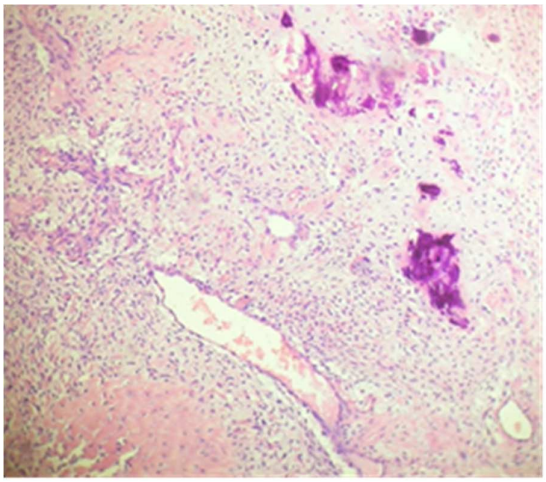

Fig 6: Photomicrograph showing osteoid areas with flecks of calcific deposits in the highly primitive mesenchymal stroma (Case 2)

At the microscopic level, the provi- sional diagnosis of 'poorly differen- tiated osteosarcoma' in case 2 arose from the primitive appearance of the malignant mesenchyme, which in places, appeared to be associated with osteoid seams as well as tumor cartilage. It is sometimes difficult to distinguish these two lesions. The sparse osteoid was essentially pe- ripheral and reactive or metaplastic and noted in other cases. Immuno- histochemistry is advised for diffi- cult cases. Chondroid areas are positive for S-100 protien, & neu- ron-specific enolase is focally posi- tive for primitive mesenchymal cells.

Certain malignant jaw tumors in the osteosarcoma-chondrosarcoma spectrum may have microscopic appearances so variegated as to suggest hybridization, a feature that is more familiar in certain dys- plastic lesions of the jaw bones. We suspect that zones reminiscent of mesenchymal chondrosarcoma may occur in tumors that are pre- dominantly of more mature or dif- ferentiated type but detailed study

is required to confirm this. Such tu- mors may be allied to the 'primitive multipotential primary sarcoma of bone', which combines undifferen- tiated cells with multiple lines of differentiation into cartilage, bone, vessels, squamous epithelium etc (3).

Cytogenic studies of MC have shown a chromosomal transloca- tion, t(11;22) (q24;q12) similar to that which occurs in Ewing's sar- coma (6).

Differential diagnosis: The MC has to be differentiated histologically from Ewing's sarcoma, malignant peripheral neuroectodermal tumor (MPNET), hemangiopericytoma, small cell osteosarcoma, and dedif- ferentiated chondrosarcoma.

Ewing's sarcoma lacks vascular pattern and cartilaginous pattern in MC. MPNET is immunohistochemi- cally reactive for CD99 and a variety of other neural markers, lacks car- tilage formation, the nuclei are more pleomorphic and may show evidence of neural differentiation by the presence of rosettes. Hemangi- opericytoma is distinguished from MC by lack of cartilage and positive immunohistochemical staining for CD34. Dedifferentiated chondro- sarcoma may simulate MC with lob- ules of normal or low grade chon- drosarcoma juxtaposed to spindle shaped stromal cells; however, these spindle cells are large and pleomorphic with highly atypical nuclei, in contrast to the nuclei of the small cells of MC. These tumors also lack hemangiopericytoma like pattern (6,7).

Treatment and prognosis: MC is highly malignant and the frequency of metastasis is high. Metastasis is primarily by hematogenous spread, with the lungs being the most com- mon site but regional lymph nodes and bones may also be affected. The most effective therapeutic mo- dality is early radical surgery with wide surgical excision followed by chemotherapy to eradicate micro metastases that have not been pre- viously noted. Overall, there is a 10- year survival rate of 28%. Survival time is longer with maxillary MCs than with mandibular lesions (8, 9). In our reported two cases, the first patient has undergone a surgical excision followed by radiotherapy and is leading a normal life for the past 9 months. He has been advised for checkups at regular intervals. The second patient was refused of any treatment due to the large tu- mor and poor prognosis by two leading medical institutions and is now under palliative medicines.

CONCLUSION

The present cases emphasize that any history of a painless mass fast or gradually growing, or recurrence of a lesion following surgical inter- vention of a benign bone tumor should be cautiously evaluated. Ad- equate biopsy and multiple sections should be examined properly for di- agnosis of MC. As recurrence and metastasis is very frequent with MC, long term follow-up should be advised. MC should be considered rationally as a completely separate entity from other chondrosarcomas due to their unusual behavior, dis- tinct histopathology, late recur- rence and poor prognosis.

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