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Case Report

Mesenchymal chondrosarcoma of mandible: a case report

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Abstract: Chondrosarcomas are malignant mesenchymal neoplasm with cartilaginous differentiation of bone and soft tissue. It is an uncommon primary malignant neoplasm of maxillofacial regions. Majority of chondrosarcomas of head and neck arise from maxilla, with very few cases arise from the mandible. The clinicopathological and radiographic features are usually characteristic; however, not decisive. CS is usually treated by wide surgical resection and is relatively chemoradioresistant. Radiotherapy is usually advised for high-grade lesions, and chemotherapy has a palliative role. Here we present a case high grade chondrosarcoma treated by surgery and adjuvant chemoradiotherapy.
Keywords: Chondrosarcoma, mandible, mesenchymal, cartilage.

INTRODUCTION

Mesenchymal chondrosarcoma [MCS] is an uncommon, slow enlarging malignant neoplasm representing 1 % of all CS [1]. It is the second most common primary bone malignancy after osteosarcoma [2]. Chondrosarcoma affecting Head and neck is very rare, and accounts for less than 10% of all cases [3]. MC is characterized by a biphasic pattern consisting of areas of hyaline cartilage mixed with epitheloid or small cell morphology. CS usually grows within a bone or on its surface [4]. It may occur at any age; however, majority of cases occur between the 3rd and 6th decades of life with male predominance. Classical clinical feature is a painless swelling leading to the expansion of the buccal and lingual cortical plates with occasional premature loosening of teeth. Pain may be a late stage feature, and regional lymphadenopathy is very rare [3]. CS has aggressive course and a high malignant potential. Head and neck chondrosarcomas account for about 5 to 10 %, with larynx and nasal cavity being the most common site. Most common site affected in the head and neck is anterior maxilla. Chondrosarcomas arising from mandible is extremely rare and occurs mostly in the premolar and mandibular symphysis region. To the best of our knowledge, very few cases of mandibular mesenchymal chondrosarcoma are noted in the literature. Here we describe a patient with chondrosarcoma of the left mandibular region with brief review of the literature.

CASE REPORT

A 32 year old male presented with a chief complaint of swelling involving the left side of face for about 5-6 months. External examination of face showed presence of a medium sized, firm to hard, slightly tender swelling measuring 8 x 8cm with ill-defined margins involving the left mandibular region. Overlying skin appeared normal [fig 1a]. Intraoral examination revealed the presence of firm, lobular swelling in left alveolar region extending from left lateral incisor to molar region, associated with expansion of both the cortical plates. Egg-shell crackling was evident over some areas of the buccal cortical plates. Regional teeth were fallen with the overlying mucosa had a normal appearance [fig 1b].

Orthopantomogram [OPG] revealed a large, ill-defined radiolucent lesion, characterized by marked alveolar bone destruction, resorption and thinning of the lower border of the mandible [fig 2]. Computed tomography scan [CT scan] revealed the presence of a large defined osseous lesion involving body and ramus of mandible measuring 5 x 7 cm. The lesion is involving left medial pterygoid and muscles of floor of mouth.[fig 3a-d]

Based on the clinical and radiological findings, an incisional biopsy was performed from the representative site of the lesion. Microscopic examination revealed the presence of a proliferated mass of cartilaginous tissue showing marked cellular and nuclear pleomorphism with nuclear hyperchromatis

m and confirmed the diagnosis of high grade mesenchymal chondrosarcoma [fig 4]. Patient received 3 cycles of neoadjuvant chemotherapy [Cisplatin and doxorubicin] with partial response [fig 5a & 5b] and patient was planned for definitive surgery. Wide local excision (extended hemi mandibulectomy with supra-omohyoid block dissection of neck) was performed.[fig no 6a-c]. Post-operative period was uneventful. Histopathology of the resected specimen revealed well circumscribed lobulated tumor with myxoid areas and with the characteristic confirmative features of "High Grade Chondrosarcoma." Patient received adjuvant radiotherapy and was disease free for 4 months. Patient developed recurrence involving the left mandible [fig 7] and was treated by palliative chemotherapy for 3 months and expired later due to extensive lung metastasis.

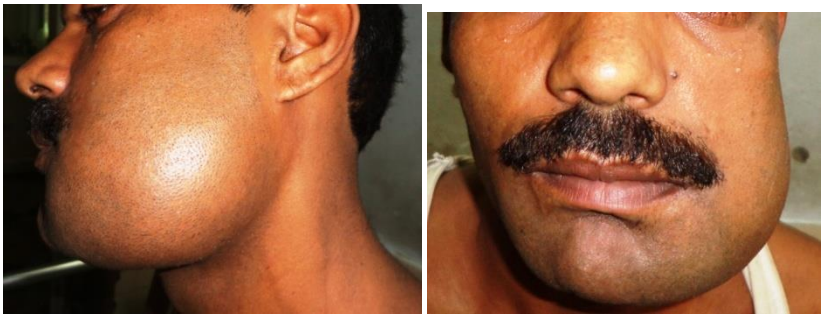


Fig 1a & 1b: Extraoral clinical photograph showing diffuse swelling in left mandibular region .

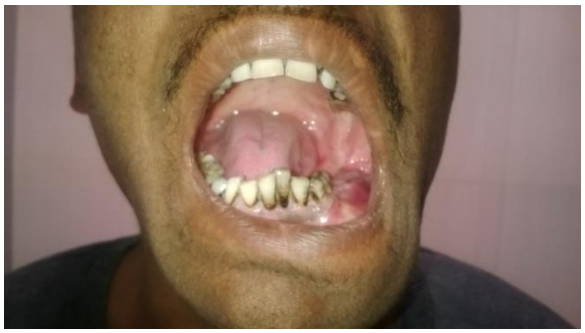


Fig 1c: Intraoral examination showed presence of firm, lobular swelling in left alveolar region.

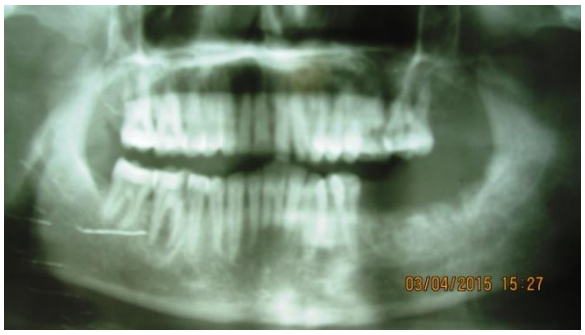
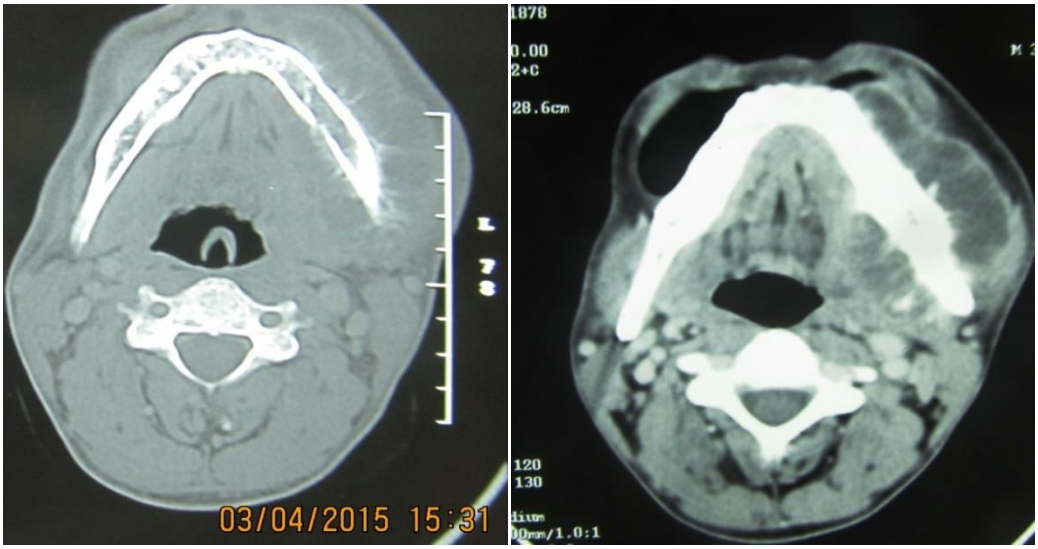
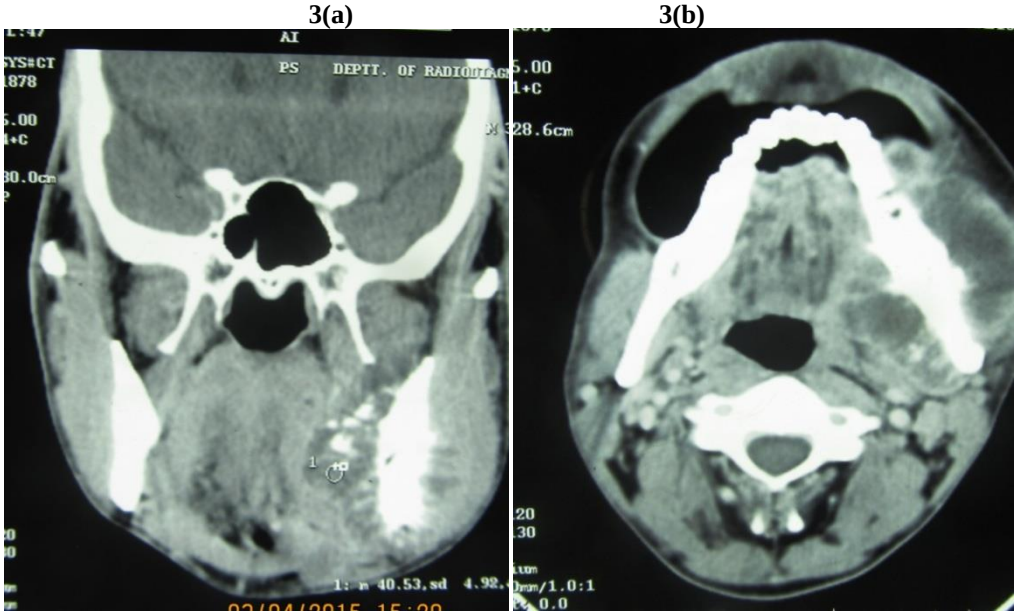


Fig-2: Orthopantograph showing diffuse radiolucent osteolytic lesion in the body and ramus of left mandible



3(a) 3(b)



3(c) 3(d)

Fig 3a-3d: CECT scan showing the presence of a large defined osseous lesion involving body and ramus of mandible

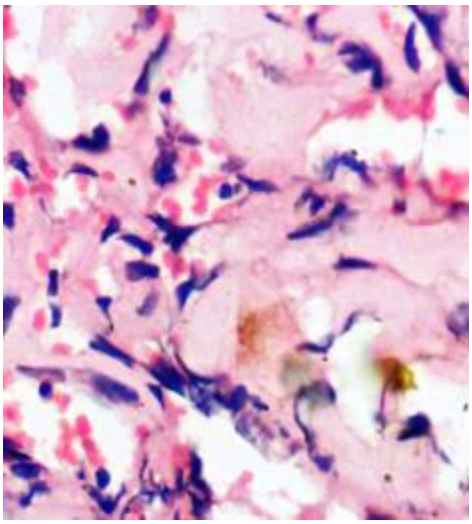
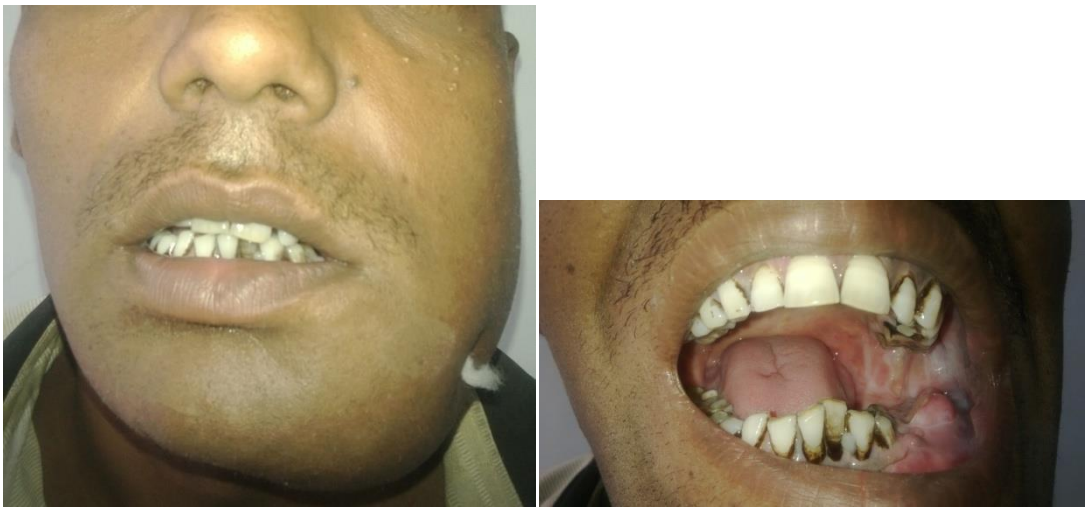


Fig-4: Photomicrograph showing presence of a proliferated mass of cartilaginous tissue showing marked cellular and nuclear pleomorphism with nuclear hyperchromatism.



5(a) **5(b)**

Fig 5a & 5b: Clinical photograph showing decrease in the size of the swelling (partial response to chemotherapy).

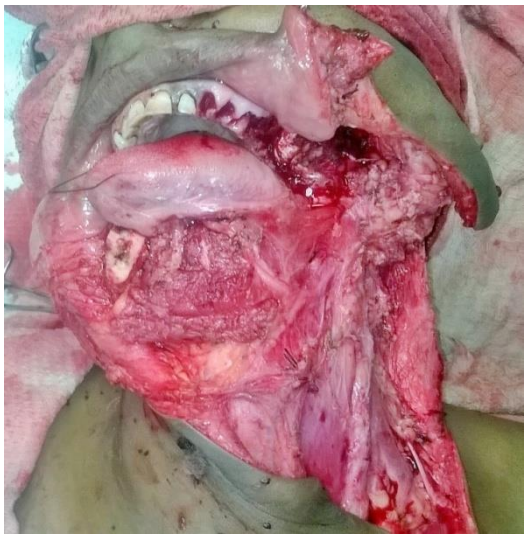
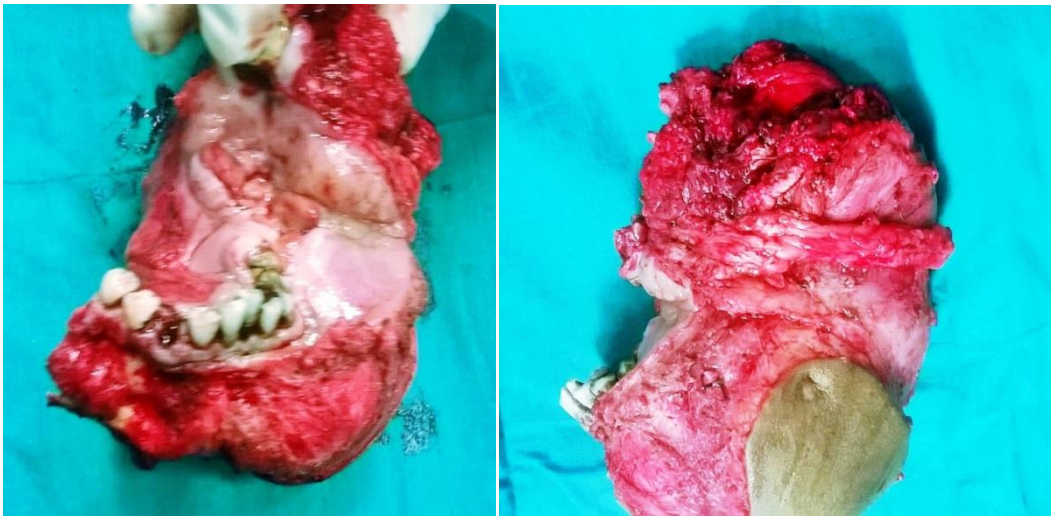
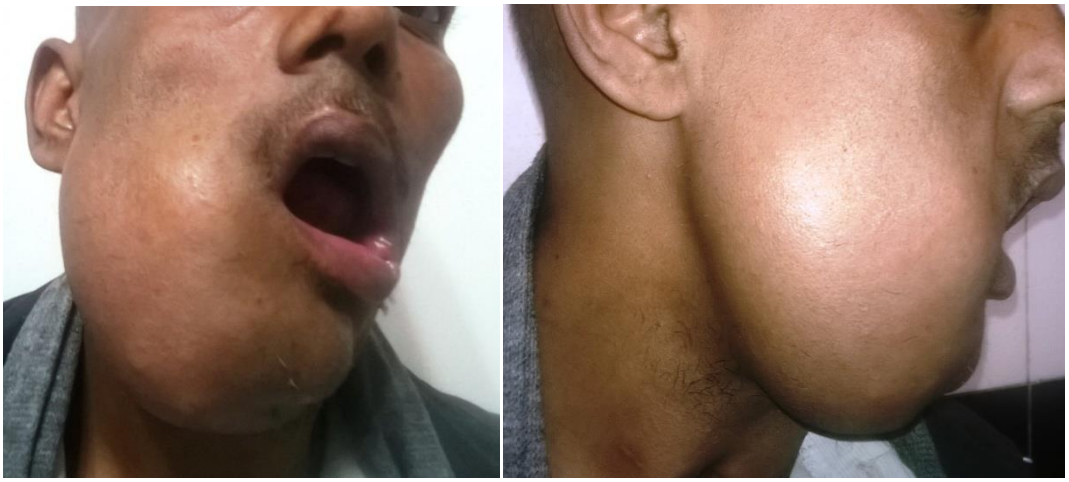


Fig 6a: Intraoperative photograph showing defect on the left side of face following resection.



6(b) **6(c)**

Fig 6b-6c: Specimen photograph show tumor resected by left extended hemimandibulectomy .



7(a) **7(b)**

Fig 7a-b: clinical photograph showing recurrence on the right side of mandible

DISCUSSION

Chondrosarcomas are slow growing, malignant mesenchymal neoplasm. CS arises from normal chondroid tissues or from embryonic cartilagenous rest. Exact origin of this malignant neoplasm in the head and neck region is controversial. CS may be primary (de novo) or secondary which arise from preexisting enchondroma or osteochondroma. It may be induced by irradiation, from pre-existing Paget's disease of bone or in association with Fibrous Dysplasia [2]. Approximately 5 to 10% of CS occur in head and neck, with larynx and nasal cavity being the most common site. Anterior maxilla is the most common site affected in the head and neck, where preexisting nasal cartilage is present. Chondrosarcomas arising from mandible is very rare and occurs mostly in the mandibular symphysis region. Majority of CS present as painless swelling and pain may be a late stage feature, and regional lymphadenopathy is very rare.

Radiological findings are usually non specific, includes irregular intramedullary radiolucenct osteolytic shadow, interspersed with punctuate radiopacities, expansion and destruction of the cortical plates, widening of the Peri odontal ligament spaces or even sun-burst appearance at the periphery [2,3,5]. CT scan is superior in defining the peripheral extent of the neoplasm compared to panoramic or flat-plate radiographs [10]. Microscopic features of high-grade chondrosarcoma usually reveal pronounced cellular and nuclear pleomorphism including the occasional presence of giant cells and abundant necrosis [2,5]. Conventional chondrosarcomas are characterized by varied light microscopic features and are divided into following four histologic grades primarily depending on cellularity, nuclear staining (hyperchromasia) of the tumor cells and size of the nuclei:- [2,5,6,9].

Grade I (or Low grade) – These tumors are characterized by the presence of benign cartilage, have a relatively uniform and lobular histologic appearance. Presence of atypical cells including binucleate forms may also be recorded.

Grade II (or Intermediate grade) – These tumors are characterized by a higher cellularity with a greater degree of nuclear atypia, hyperchromasia with often having myxoid stroma and enlarged chondrocyte nuclei.

Grade III (or High grade) - These tumors are characterized by a higher cellularity, marked cellular and nuclear pleomorphism, nuclear hyperchromasia and increased mitosis with occasional presence of giant cells.

Grade I and Grade II chondrosarcomas of the jaws and facial skeleton are best treated with local resection using 1-1.5 cm margins of bone and soft tissue. Grade III chondrosarcomas are treated with an initial aggressive resection margin of 3 cm in bone and 2 cm in soft tissue followed by chemotherapy [8]. Distant metastasis is usually uncommon; however, frequent in high grade, advanced or recurrent cases to the lungs, sternum and vertebrae [7]. Prognosis of chondrosarcoma of the jaws is poor as compared to that of long bones. The cause of death is usually by direct extension in to cranium.

CONCLUSION

Mesenchymal chondrosarcoma is an uncommon neoplasm of bone and soft tissue. CS usually present as painless swelling. tumor grade and respectability are the most important prognostic determinants of head and neck chondrosarcomas. Chondrosarcomas of the jaws have poor survival rate than that of chondrosarcomas arising in other parts of body. Role of chemotherapy is not well established and these are usually radio resistant.

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