Case Report - Oncology

Central chondrosarcoma of a pediatric mandibular condyle: A case report and review

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ABSTRACT

Chondrosarcoma of mandibular condyle is an extremely rare clinical entity with only 18 cases reported till date. We report a rare case of central myxoid chondrosarcoma in a 7 year old male child with a complaint of slow growing bony hard swelling of left mandibular condyle. Panoramic radiography and CT scan revealed a 5x3cm osteosclerotic and osteolytic lesion with cortical perforation on the medial side of the left mandibular condyle with a cortical expansion of lateral side. Segmental mandibulectomy with disarticulation of the left condyle was done. To the best of our knowledge it is the first case of chondrosarcoma of the paediatric mandibular condyle to be reported. The etiopathogenesis, clinical, radiological diagnosis and various treatment modalities of chondrosarcoma are discussed

Keywords: Chondrosarcoma, disarticulation of condyle, pediatric mandibular condyle, temporomandibular joint

INTRODUCTION

Chondrosarcomas (CH's) are rare malignant mesenchymal tumors arising from cartilage. They comprise 10-12% of all malignant mesenchymal tumors and are most commonly seen in femur, humerus, sacrum and pelvis.[1] Extragnathic CH's are more commonly seen in 5th and 6th decades. CH's occurring in the oromaxillofacial region are very rare. Of the published data, only 5-12% of CH's occur in maxillofacial region with the most common sites being maxilla (44.6%), nasal septum and ethmoids (41.1%), nose tip (3.6%).[2] CH's of temporomandibular joint (TMJ) commonly occur in 2nd and 3rd decades. CH's rarely occur in mandible (10.7%) if they do, they occur in areas of endochondral ossification of which symphysis is the primary site followed by condyle and coronoid process. CH's occurring in TMJ[3] is usually extremely rare with only 18 reported cases in the literature with all of them occurring in adult population with youngest age being 17 years. Extragnathic CH's are more aggressive then those occurring in maxillofacial region. We present here a rare case of 7-year-old male patient who was diagnosed with low grade central myxoid CH of left mandibular condyle.

CASE REPORT

This was a case report of a 7-year-old male patient who reported with a complaint of asymptomatic; slow growing, bony hard swelling on left preauricular region since 1 year. He developed non-specific progressive dull pain from past 2 months in the same region. His medical history was non-contributory. Clinical examination revealed 2 cm × 2 cm non-tender, bony hard swelling on left preauricular region with the overlying skin being pinchable, normal in color and consistency. Maximal inter-incisal opening was limited to 18 mm with posterior open bite on left side and contralateral posterior crossbite. There was midline shift of the mandible to the contralateral side with normal lateral movements. No evidence of dental foci of infection. Intraoral palpation revealed a 1 cm × 2 cm soft-tissue swelling with cortical expansion along the medial aspect of left ramus and coronoid of mandible. No cervical lymphadenopathy was present.

Panoramic radiograph revealed a mixed lesion measuring about 4 cm × 3 cm with osteolytic and osteosclerotic changes on the medial aspect of left mandibular condyle extending until the

sigmoid notch [Figure 1a]. Computed tomography (CT) scan confirmed a 5 cm × 3 cm osteolytic and osteosclerotic lesion with three areas of cortical perforation along the medial aspect of the lesion with soft-tissue mass extending in to the infratemporal fossa and cortical expansion on the lateral aspect of mandibular condyle along with intra lesional calcifications [Figure 1b]. CT revealed no evidence of cervical lymphadenopathy. Based on the clinical and radiological findings a provisional diagnosis of osteochondroma was made. Fine-needle aspiration biopsy was done, which revealed osteochondroma of left mandibular condyle.

Patient was planned for segmental mandibulectomy with distarticulation of the left condyle under general anesthesia. Through high cervical submandibular incision, subplatysmal flap was raised [Figure 2a]. Marginal mandibular nerve was preserved. Facial artery and vein were ligated. The lower border of the mandible was identified and subperiosteal dissection was done until the lesion was exposed in glenoid fossa [Figure 2b]. Osteotomy cut was placed in the angle region with tumor clearance margins of 1.5 cm [Figure 2c]. Hemostasis of inferior alveolar vessels was achieved [Figure 2d]. The lingual attachments to the mandible were stripped along with the lateral pterygoid muscle and disarticulation of the condyle [Figure 2e and f] was done along with discectomy. Intraoperative frozen section analysis revealed tumor free margins. A 2.5 mm titanium reconstruction plate was fixed with 2.5 mm × 8 mm titanium screws to maintain the anatomical contour of the mandible [Figure 3]. Hemostasis was achieved, suction drains were placed and closure was done in layers. Post-operative period was uneventful with no evidence of facial nerve palsy. Physical rehabilitation was started from the 3rd day. Mandibular movements were normal. Patient was discharged on the 5th day of the surgery.

Histopathological examination of excised specimen revealed an infiltrating cellular lesion composed of lobules of hyaline cartilage invading surrounding fibrous tissue, marrow spaces and bony trabaculae. The chondrocytes show enlarged nuclei with nuclear hyperplasia hyperchromatism [Figure 4a and b]. Myxoid matrix was strikingly prominent with foci of intra lesional calcifications [Figure 4c]. It was diagnosed as well-differentiated or low grade CH of left mandibular condyle.

The patient was sent for whole body three phase bone scintigraphy scan to rule out occult metastasis and postoperative residual loco-regional lesions. There was no evidence of any metastatic bone

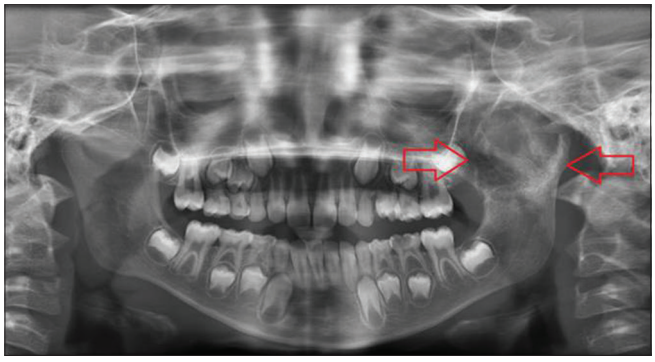

Figure 1a: Panoramic radiography showing a lesion on left temporomandibular joint involving ramus and coronoid process extending till sigmoid notch

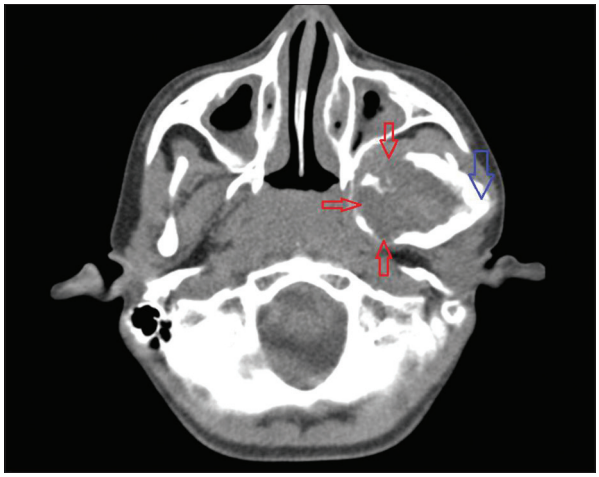

Figure 1b: Computed tomography scan showing a osteolytic and osteosclerotic lesion of 5 × 3 cm with cortical expansion on lateral side with three areas of cortical perforation on medial side with intra lesional calcifications

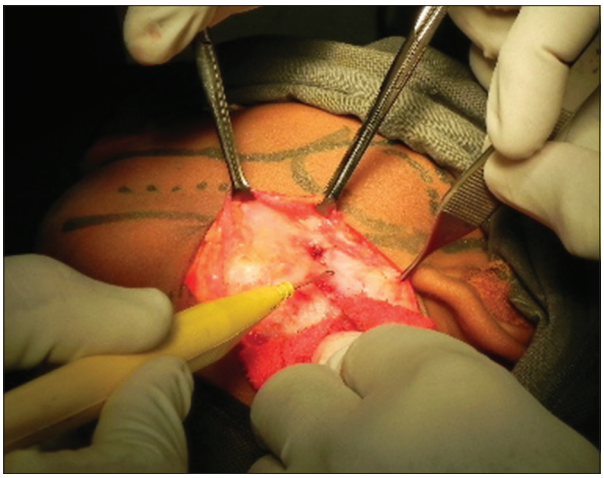

Figure 2a: Marking of high cervical submandibular incision and dissection in layers

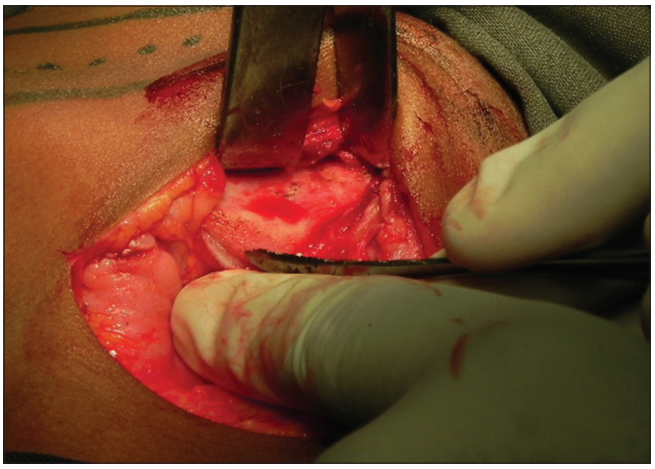

Figure 2b: Subperiosteal dissection of mandible to expose the lesion

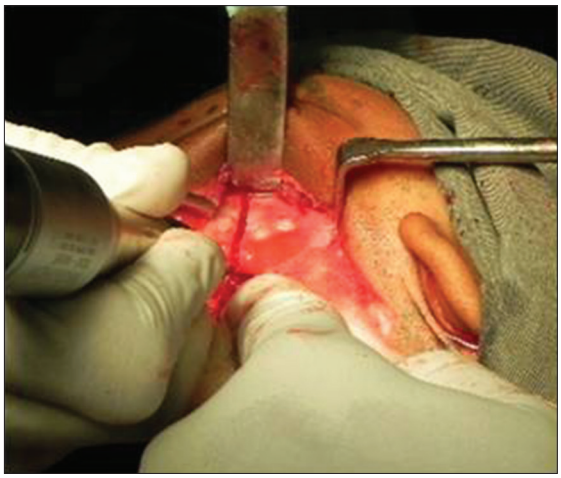

Figure 2c: Osteotomy cut placed at the angle region with a clearance margins of 1.5 cm

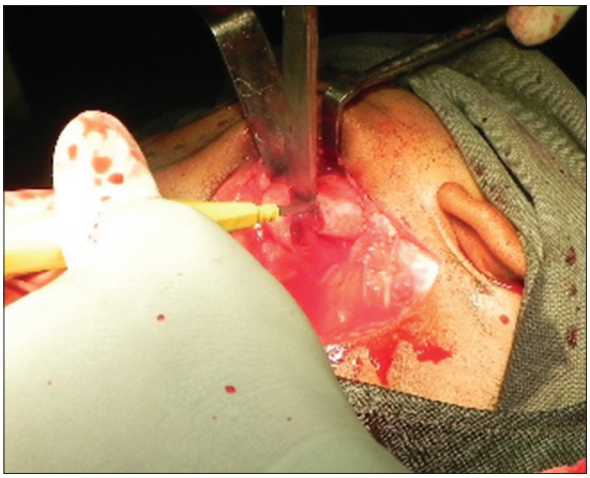

Figure 2d: Completion of osteotomy and hemostasis of inferior alveolar vascular bundle

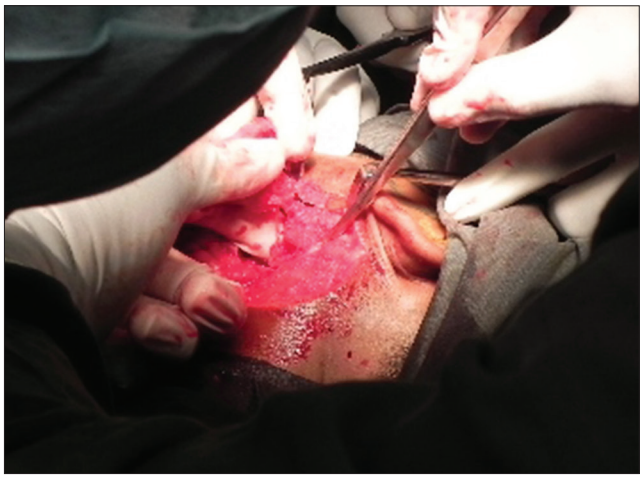

Figure 2e: All muscular attachments to the mandible were stripped off and disarticulation of condyle was done

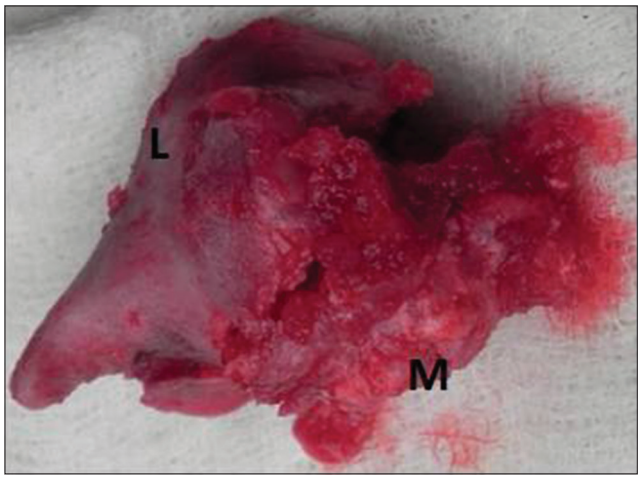

Figure 2f: Excised tumor mass (M-medial side, L-lateral side)

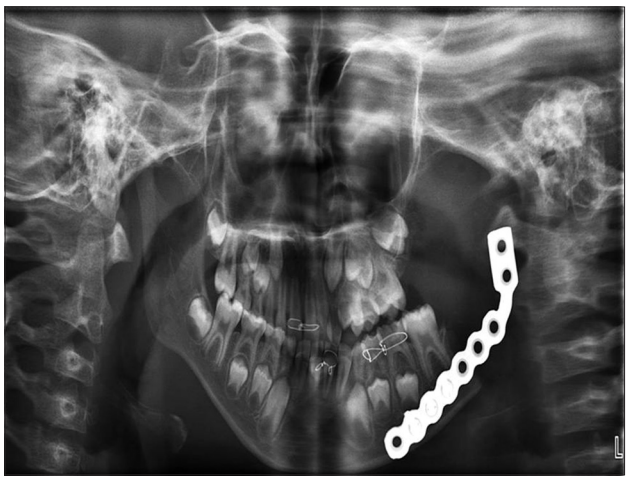

Figure 3: Fixation of reconstruction plate to maintain the anatomical contour

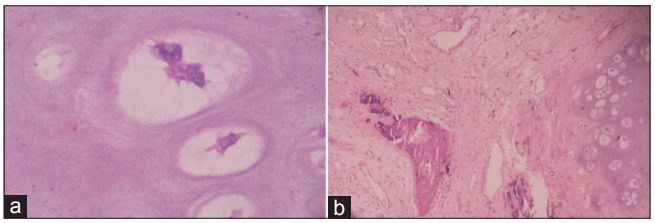

Figure 4 (a and b): Histopathological examination of the tumor revealed chondrocytes with oval, enlarged binucleate forms with nuclear hyperchromatism in myxoid matrix

disease. Patient was free from locoregional occurrence for a follow-up of 1 year [Figures 5 and 6].

DISCUSSION

Chondrogenic tumor is an uncommon pathological condition arising in the head and neck region. Their occurrence in the mandible is rare. World Health Organization defined CH's as malignant tumors arising from a hyaline cartilage differentiation and is therefore characterized by cartilage not bone, origination from neoplastic cells. They account for 20% of all primary malignant bone tumors usually affecting axial skeleton, humerus, femur, vertebrae, sternum and ribs. CH's of TMJ are extremely rare. The affected age group being 2nd and 3rd decade and are

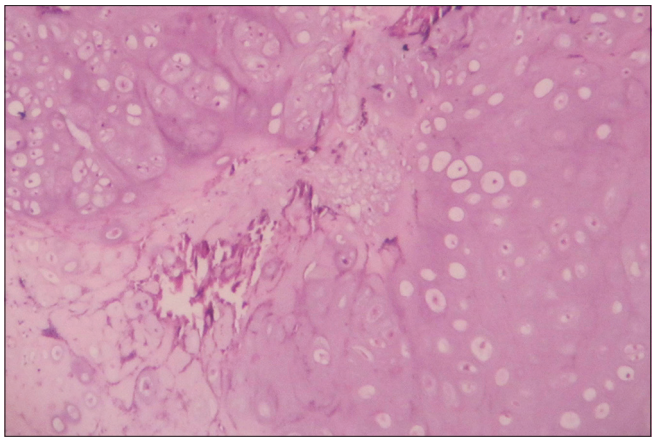

Figure 4c: Intra lesional calcifications are evident with myxoid matrix

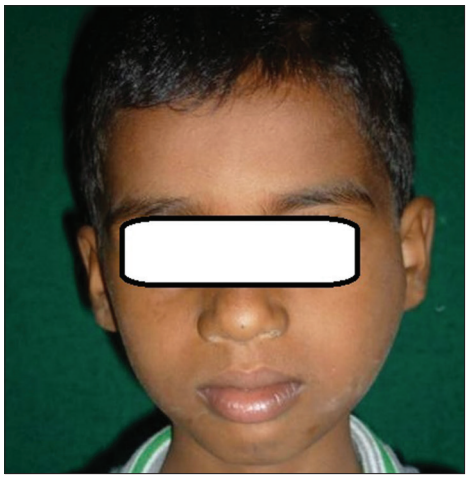

Figure 5: Preoperative photo showing facial asymmetry on left side auricular region

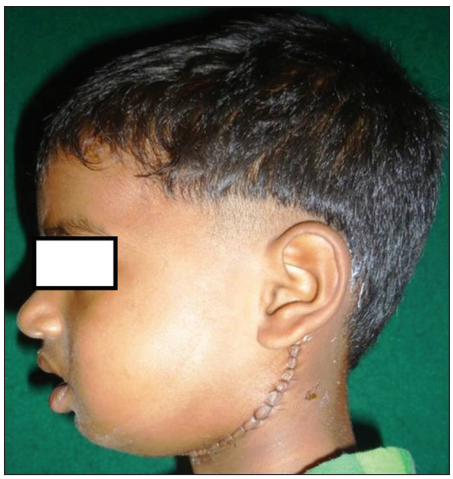

Figure 6: Postoperative photo

rarely found in children. The literature search revealed that 18 cases of CH's of TMJ have been reported with youngest being 17-year-old female with male to female ratio 2:1.[4] We report a case of CH of pediatric left mandibular condyle in a 7-year-old child which is the first case to be reported to the best of our knowledge.

CH's may arise primitively or secondary to any lesion with most common being enchondroma and osteochondroma. CH's may arise from bone-central CH's, from periosteum-juxtacortical CH's or from soft tissues – peripheral CH's. CH's rarely occur in mandible (10.7%) if they do, they occur in areas of endochondral ossification of which symphysis is the primary site followed by condyle and coronoid process. These tumors may often mimic disorders of TMJ and go unnoticed until they attain a considerable size. Clinical features may include increased condyle ramus height on affected side causing posterior open bite, latero deviation of the condyle, painful and limited mouth opening, swelling in the preauricular region with or without pain. Mostafur and futran pointed out that CH's are more painful than endochondroma and are unnoticed with an average diagnostic delay ranging from 13 months to 8 years.[5] In our case the period of delay was 10 months. Radiological investigations like CT and magnetic resonance imaging scan reveal irregular erosion of the condyle with calcifications localized within the intra lesional space. The diagnosis of CH is purely based on the histological findings. Fine needle aspiration biopsy or incisonal biopsy can be done to have a definitive preoperative diagnosis for a proper surgical treatment protocol. Some of the authors believe invasive investigations may induce anaplastic transformation and diffusion during the manipulation of the tumor.

Chaudhry *et al*.[6] studied 36 cases of chondrogenic tumors of the head and neck region and found under diagnosis or misdiagnosis in 20% of these lesions. This underlines the importance of proper clinical and histological diagnosis to achieve optimal management of these hidden tumors. Evans *et al*.[7] gave the histological grading of the tumor based on the mitosis, cellularity and tumor size. Grade I or low grade CH's have an ample chondroid matrix with hypercellularity, chondrocytes with an increased volume and polymorph hyperchromatic binucleate forms. Grade II or intermediate grade CH's display a higher degree of cellularity with less matrix increased mitotic figures. Grade III or high grade CH's exhibits high degree of pleomorphism and dysplastic figures. Based on the tissue involved they are classified as myxoid, clear cell, mesenchymal and dedifferentiated CH's. Morris *et al*. said that tumor grade and complete resection are the most important prognostic factors for head and neck CH's.

Three phase bone scintigraphy can be done to find metastatic secondary tumors. Lymphatic spread in CH's are very rare, whereas hemotogenous spread occurs in 10% in Grade II and 71% in Grade III CH's.[8] Distant metastasis is very rare, occurring most commonly to the lungs, although metastases to the vertebrae have also been reported. Local recurrence of the tumor is dependent on tumor grading.[9] Due to the rarity of the CH's in maxillofacial region, no proper treatment protocols are available in literature. They are generally treated by multimodal approach such as en bloc surgical resection, radiotherapy, chemotherapy, immunotherapy based on the histopathological grading of the tumor. Surgical resection with wide local tumor free margins remains the gold standard primary treatment for this

neoplasm. Grade I CH's have a lesser aggressive behavior when compared to other grades. Hence elective neck dissection is not always necessary.[10]

In our case, patient was diagnosed with low grade CH with negative metastatic bone disease and clear surgical resection margins; postoperative adjuvant therapy was not given. The use of postoperative chemotherapy is also controversial in the cases of clear pathological margins due to paucity of cases. Arlen *et al*. reported 40% (8 of 18 patients) with disease-free survival at 5 years in patients with CH of the mandible. He noted that locoregional recurrence at or near the region of the primary lesion and eventually resulting in uncontrollable disease was the primary cause for death in the remaining patients. In our case, locoregional control was achieved with no recurrence in 1 year of follow-up. Other treatment modalities such as radiotherapy, chemotherapy and cryotherapy can be used as an adjuvant. Crawford *et al*. used chemotherapy as neoadjuvant treatment and suggested that although combined chemotherapy did not decrease the size of the tumor, it inhibited tumor growth and spread.

CONCLUSION

Malignant tumors of the condyle in pediatric patients often possess a challenge for diagnosis and surgical treatment. Thorough clinical, radiological and histopathological investigations have to be carried out by the maxillofacial surgeons to recognize these hidden tumors at the earliest. This case report reveals that CH though rare, can occur in pediatric patients.

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