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CASE STUDY

CHONDROSARCOMA OF RAMUS AND CORONOID PROCESS: A RARE ENTITY

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ABSTRACT

Chondrosarcoma is a malignant neoplasm that is characterized by formation of cartilage, with progressive local invasion to the surrounding tissues. Currently, it is the second most common bone tumour next to osteosarcoma and comprises 10-12% of all malignant bone tumours. They usually involve the long bones, approximately 1% to 3% of chondrosarcomas occurs in the maxillofacial region, predominately in the anterior maxilla, while other facial bones are less affected. The favoured site of involvement in mandible is the molar region and symphysis region, they rarely occur in ramus, condyle and coronoid process. Even though, the clinical and radiological features are characteristic but not conclusive at early stages. It is reported that ultimate diagnosis of chondrosarcoma takes about 12 months in average, due to its varied clinical behaviour. Treatment of chondrosarcoma is usually surgical but prognosis of the jaw lesions is poor as compared to that of long bones due to their direct extension to the skull base or distant metastasis. Thus, clinical course of chondrosarcomas of jaws necessitates a prompt diagnosis and early intervention which could be crucial in saving patient's life. Hence, Oral physician plays a pivotal role in early diagnosis which could contribute towards better prognosis. This paper reports a rare case of chondrosarcoma of mandible in a 40 year old male who has presented to our department with a chief complaint of swelling in left parotid region with a follow up of 4 months.

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INTRODUCTION

Chondrosarcoma also known as chondrogenic sarcoma is a malignant neoplasm that is characterized by formation of cartilage, with progressive local invasion to the surrounding tissues.Primarychondrosarcomas develop de novo, whereas secondary chondrosarcomas arise from pre-existing chondroma or osteochondroma. (Deyhimi and Keshani, 2012) Chondrosarcomas comprise 10-12% of all malignancies of bone tumours. They usually arise in the femur, humerus, pelvis, and sacrum. Chondrosarcomas occur in patients of all ages and the majority of them are over 50 years old. Approximately 1% to 3% of chondrosarcomas occurs in the maxillofacial region, predominately in the anterior maxillary region, while mandibular body, ramus, nasal septum and paranasal sinuses are less affected. The favoured site of involvement in the mandible is the molar and symphysis region, and they rarely occur in the ramus, condyle, coronoid process. In the mandible, remnants of Meckel's cartilage could be the sources of cartilaginouscells which are able to produce secondary cartilage in the mental region and the coronoid and condyloid processes. These structures could be a rational for development of a tumour in these regions. (Deyhimi and Keshani, 2012) Chondrosarcomas of head and neck present without pain incontrast to excruciating painful presentation of chondrosarcoma of long bones. Some authors believe there is no sexpredilection, however others have found a male to female ratio of 2:1. (Patel *et al*., 2013) The radiological findings are not pathognomonic and usually include irregular intramedullary radiolucencies interspersed with punctate radiopacities, expansion and destruction of the cortical plates, widening of the PDL spaces or even sunburst appearance at the periphery. (Kundu *et al*., 2011) The ultimate diagnosis of this lesion depends on histological analysis.It has been depicted that many cases with initial diagnosis of benign chondroma or osteochondroma had a final diagnosis of chondrosarcoma. The management of chondrosarcoma has been primarily surgical and the extent of which is modified by site and size and grade of the tumour. (Kanlikama, 1991) Treatment of chondrosarcoma of the head and neck has posed a challenge because of varying biological behavior at different anatomic sites. (Kanlikama, 1991) Prognosis of the jaw lesions is poor as compared to that of long bones due to their direct extension to

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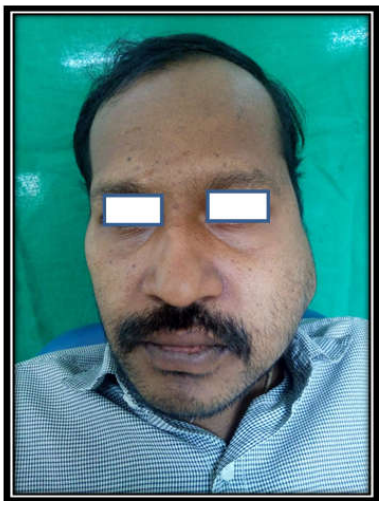
the skull base or distant metastasis. (Kundu *et al*., 2011) Here we present a rare case of chondrosarcoma primarily involving the coronoid process and ramus of the mandible with emphasis on the clinical and radiological features.

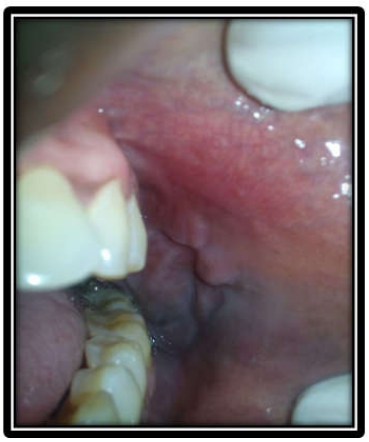
Case report

A male patient aged 40 years reported to our department with a chief complaint of swelling on the left side of the face with difficulty in opening mouth and tooth pain in left upper back teeth region since 4 months. History of presenting illness revealed that the patient had visited our department for tooth pain in the same region 4 months back for which analgesics were prescribed and recalled the patient was recalled for further evaluation. But the patient could not report back to us following which he noticed a swelling on the same side which was insidious in onset and was gradually increasing in size. There was no associated history of pain, paraesthesia or pus discharge. Patient had tooth ache in left upper back region since 4 months which was dull in nature. There was no associated history of trauma or fever in the past and any other associated symptoms. There was no history of similar swelling in the past. Patient had a deleterious habit of consumption of alcohol since last 15 years. Patient used to consume 180ml of alcohol per day. Patient had no other deleterious habits. Patient is a 40year old adult male well oriented to time, place and person with normal built and gait and all vital signs were under normal limits. Head and neck examination revealed, a diffuse swelling on the left middle and lower third of the face extending antero-posteriorly from the left corner of the mouth to left tragus of the ear. Supero-inferiorly from the left ala tragus line to inferior border of the mandible. Skin over the swelling appeared apparently normal. The swelling was pointed in front of the tragus of the ear in the left TMJ region. On palpation all the inspectory findings were confirmed. The swelling was non tender, firm to hard in consistency in the region adjacent to front of the tragus of the ear and firm in the rest of the region of swelling. There was no localised rise in temperature of the swelling. The plane of the swelling from which it was arising could not be ascertained.

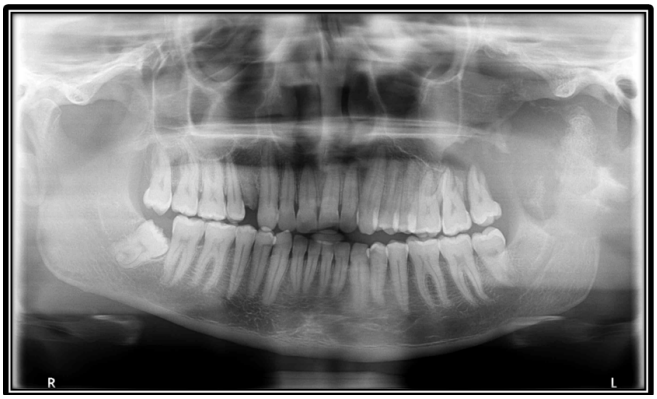
TMJ examination revealed limited mouth opening of 15mm with restricted protrusive and lateral movements. On palpation the left TMJ movements could not be appreciated. However, no tenderness or clicking was noted in the TMJ region bilaterally. Intra oral examination revealed a diffuse swelling noted in left posterior region of the oral cavity in the retromolar region obliterating the posterior buccal vestibule in the 28 region extending inferiorly to 38 region. Mucosa over the swelling appears irregular with presence of two prominent mucosa tags. Retromolar area over the swelling appears to have diffusegreyish discolouration of the mucosa.The stenson's duct appeared to be obliterated. The palatal gingiva of the 27,28 region also showed surface irregularities. On palpation, inspectory findings were confirmed but extensions of the swelling could not be well appreciated. The mucosa overlying the lesion was soft and non-tender on palpation.Patency of the stenson's duct could not be appreciated but salivary secretion was normal. Hard tissue examination revealed grade III mobility of 28 and grade II mobility of 27 with tenderness on percussion.Based on the history and clinical findings a provisional diagnosis of malignant jaw bone tumour of the left lower ramus region of the jaw was given. Differential diagnosis of malignant tumour of the left parotid gland andleft parapharyngeal space infection was considered.Patient was further subjected to orthopantamograph which revealed a poorly defined radiolytic lesion measuring about 6x5 cm in its greatest dimension in left ramus region with irregular borders andextending supero-inferiorly from posterior maxillary tuberosity to the mid ramal area.Extending anteriorly to the second molar region resulting in complete destruction of coronoid process and sigmoid notch and the maxillary alveolar segment in 27, 28 regionwith destruction of the floor and posterior wall of maxillary sinus. "Floating teeth appearance" in the 27,28 region was noted. The lesion has caused the erosion of condyle and condylar head altering its morphology leaving a thin border of anterior ramus.Radiographic diagnosis of malignant neoplasm in the left ramus region of the jaw was given. Differential diagnosis of chondrosarcoma and osteosarcoma was considered. Further ultrasound of left parotid gland region was done which revealed heterogenous predominantlyhypoechoic lesion in the superficial lobe of parotid gland. Minimal peripheral vascularity was noted within the lesion with few anechoic areas (necrosis) and tiny hyperechoic areas (calcifications) within. The lesion was extending upto the ramus of left hemimandible. Mild irregularity noted in the cortex of the ramus of mandible. Multiple enlarged bilateral cervical lymph nodes were also noted. The ultrasound findings gave the impression of a likely malignant heterogenous lesion in the left parotid gland with bilateral cervical lymphadenopathy. Further fine needle aspiration cytology depicted a hemorrhagicgelatinousaspirate which revealed tumour cells in clusters, sheets scattered in a myxoidstroma.



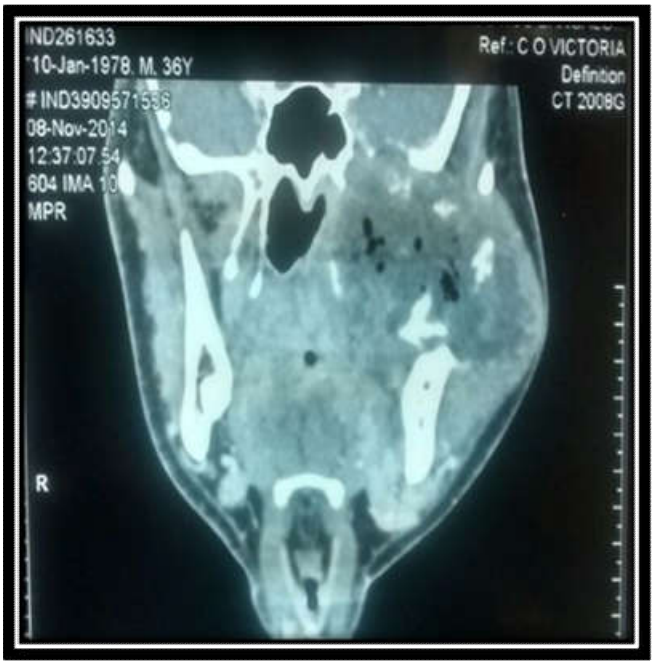
Photograph 1:-Front profile of patient showing swelling in the left middle and lower 1/3rd of the face

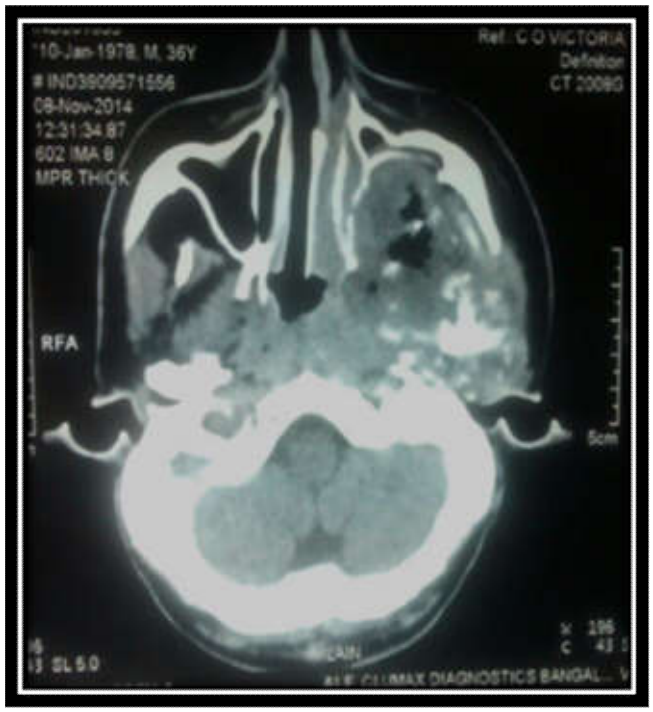


Photograph 2 :- Showing diffuse lobulated swelling with lobules in left posterior buccal mucosa



Photograph 3 :- OPG showing a large ill-defined radiolucency in left ramus region





Photograph 4:- Cross sectional CT image showing lytic destruction of left hemimandible and ramus with multiple coarse calcific foci described as "ring and arc pattern''

Chondroid cells with mild pleomorphism was also noted some of which were binucleated. These cytological features gave an impression of suspected low grade chondrosarcoma and advised excisional biopsy. CT scan revealed a well-defined heterogeneously enhancing lesion with multiple coarse cloud like calcific foci described as "ring-arc pattern", within causing lytic destruction of the ramus of left hemi-mandible. The lesion is also showing few nonenhancing fluid attenuation areas and air foci within (suggestive of Necrotic areas). Superiorly, it is filling the left maxillary sinus, causing lytic destruction of the posterior wall of left maxillary sinus and also extending up to the inferior wall of left orbit with resultant destruction of inferior wall of orbit. Medially, it is involving the left lateral pterygoid muscle and extending deep into the left parapharyngeal space. Laterally, it is involving the left masseter muscle and superficial portion of the left parotid gland. These CT scan findings gave an impression of a well-defined lytic lesion with chondroid matrix involving ramus of left hemi-mandible with invasions as described above suggestive of Cartilage forming Malignant bone tumour

DISCUSSION

Chondrosarcomas are malignant cartilaginous tumours that arise in bone or superimpose on benign cartilaginous tumours and arise in bone or superimpose on benign cartilaginous tumours. These tumours are slow growing and locally aggressive malignancies, rarely encountered in head and neck region and they display a propensity for progressive spread and multiple recurrences that eventually lead to death, if inadequately treated. (Kanlikama, 1991)

Pathogenesis

There is considerable doubt about the origin of all cartilaginous tumours. The predisposition for chondrosarcoma of the anterior mandible and posterior maxilla has led some authors to postulate that the tumour arises in association with cartilaginous remnants of the nasal capsule and Meckel's cartilage respectively. On contrary, some other authors suggested that chondrosarcoma can arise de novo from osseous tissues without presence of cartilaginous rests. (Kanlikama, 1991)

Clinical Features

Only 5% to 10% of chondrosarcomas occur in the head and neck, with the larynx and the nasal cavity being the most common sites. Chondrosarcoma of the jaw occurs primarily in the anterior maxilla, where pre-existing nasal cartilage is present. Chondrosarcoma of the mandible is rare and occurs mostly in the mandibular symphyseal and molar region. Clinically, the tumour presents as a swelling, which may or may not be painful and cause loosening of the involved teeth, with widening of the periodontal ligament space. Chondrosarcomas of the jawmay mimic periodontal lesions, with associated bone loss. (Saini *et al*., 2007) The importance of this case is ascribed in its rarity, which was later diagnosed as well differentiated chondrosarcoma. The clinical presentation of this case is unique as it presented with a gross facial asymmetry on the left side of the face withnontenderswelling in the parotid region with trismus. Very few studies have reported that chondrosarcoma of mandibular condyle may provoke preauricular swelling. Based on the aggressive nature of the painless swelling associated with

loosening of teeth a provisional diagnosis of malignant jaw bone tumour was given.

Radiological Features

There are no radiographic findings that are pathognomonic for chondrosarcoma, although single or multiple radiolucent areas with poorly defined borders can be seen on plain films. Evidence of bone destruction is often present, and mottled densities caused by calcification are occasionally seen. (Saini *et al*., 2007) In the case described here orthopantamograph revealed anexpansileradiolytic lesion causing complete destruction of the coronoid process and sigmoid notch, condyle and adjacent maxillary alveolar segment. Radiographic diagnosis of malignant intraosseous neoplasm was given. Differential diagnosis of chondrosarcoma and osteosarcoma was considered. However calcifications and "sunburst" periosteal reactions which are highly suggestive of chondrosarcomas or osteosarcomas were not observed. It should be mentioned here that the differential diagnosis between chondrosarcoma and osteosarcoma may be troublesome and sometimes impossible on plain radiographs. Further Ultrasound of the lesion was done immediately taking into account the anatomic extensions of the lesion. Diagnostic ultrasound imaging of the lesion showed tiny hypoechoic areas suggestive of calcific foci which lead to the suspicion of chondroid nature of the lesion. The findings of the ultrasound was further supported by CT scan which showed well defined heterogeneously enhancing lesion with multiple coarse cloud like calcific foci within causing lytic destruction of the ramus of left hemi-mandible. These calcific foci are typically described as "ring in arc" pattern of chondroid matrix mineralisation and are highly suggestive of chondrosarcoma coupled with aggressive growth features.

Histopathology

Histologic spectrum of chondrosarcoma is ranged from well differentiated growth which may be difficult to differentiate from a benign cartilage tumour, to that of high grade malignancy which has aggressive local behaviour and potential for metastasis. Evans *et al.* divided chondrosarcomainto three grades base on their cellularity, mitotic rate and nuclear size. (Kanlikama, 1991) In the case reported here the patient was subjected to Fine needle aspiration cytology which depicted chondroid cells with mild pleomorphism with cluster of tumour cells which were suggestive of "low-level" chondrosarcoma. Finally incisional biopsy was done which revealed the lesion to be grade I chondrosarcoma. It is vital to mention here that Chondrosarcoma may be misdiagnosed withchondroblastic osteosarcoma or even Ewing's sarcoma, because of their similarity in histological features.Differential diagnosis of chondrosarcoma from chondroma and chondroblasticosteosarcoma is difficultbut important. Chondroma usually arises in smallbones, and is extremely rare in the jaws and facialbones. It is wise to consider any cartilaginous tumours of the jaws, malignant or potentially malignant ratherthan benign. It is also important to differentiate chondrosarcomafrom chondroblastic osteosarcoma in thejaw; since prognosis of chondrosarcoma is more favourablethan chondroblastic osteosarcoma. Chondrosarcomagrows more slowly than chondroblasticosteosarcoma and metastasizes late in the course oftumour growth. (Deyhimi and Keshani, 2012) The 5-year survival for chondrosarcoma is approximately 90% for grade I, 81% for grade II and 4.3% for grade III. (Deyhimi and Keshani, 2012) Fortunately our patient has been diagnosed with grade I chondrosarcoma and has been planned surgical resection of the tumour. However, long-term follow-up is essential, because chondrosarcomas of head and neck region show a wide disparity in theinterval between recurrences and also in their possibilityof metastasis. (Deyhimi and Keshani, 2012)

Conclusion

Chondrosarcomas of head and neck region present with varied biological and clinical behaviour and have a poor prognosis when compared to the lesions involving the long bones due to direct extension to the skull base and distant metastasis. Oral physicians play a key role in early recognition of clinical and radiological features of these lesions which could prove crucial in saving patient's life and decrease morbidity.

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