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Case Report

Primary Chondromyxosarcoma of Maxilla- An Enigmatic Rare Case Report

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ABSTRACT:
Triple D, dreadful, drastic and dangerous, a clinical disorder which takes the mankind in panic, cancer and malignancy is a threat to psychological relief to patients and their peers. When hearing of cancer, often people turn around to sympathize and plead for quick recovery of the patient. Cancer is defined as uncontrolled cell division, often turns out to be malignant if metastasizes in distant tissues. Too many people seek evaluation for cancer, but a proper and prompt diagnosis is the question to be answered. Malignant tumours of connective tissue, being a rarity pose unnecessary situation of severe accompanied threats for the patient. One such being described in the presented case can prove how uncommon threats pose uninvited problems for the people. Often misdiagnosed, myxoid variant of chondrosarcoma can be the one to cause severe complications in the treatment planning, proving itself tobe an enigma. **Case analysis**: The case presented is of a rare tumour, chondromyxosarcoma in a 22 year old male in the middle third of the face. The lesion presented as a diffuse swelling on the right maxillary region which progressed aggressively. The histopathological investigations revealed bi and tri nucleation with a moderate degree of nuclear pleomorphism. The diagnosis of chondromyxosarcoma was confirmed and the clinicopathological findings of this rare case has been presented thereafter.

Keywords: Chondromyxosarcoma, rare cancer, invasive, myxoid, chondroid, malignant tumor.

Key Messages : chondromyxosarcoma is a rare case in India, and thus poses serious challenging threats to the clinician for prompt diagnosis and treatment planning.

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INTRODUCTION

Rare cases often pose challenge for clinicians as they need to have an eagle's eye to understand the clinical features of the lesion. Mesenchymal chondrosarcoma is rare, aggressive and uncommon type of chondrosarcoma as it arises from cartilage cells. It was first described in 1959 and less than 700 cases are recorded till date in the medical literature. The incidence of this variant if chondrosarcoma is 1 in 100,000 individuals worldwide. Any solitary lesion lasting for longer than 3weeks should have a high index of suspicion in the clinicians mind, particularly if it is indurated, accompanied by cervical lymphadenopathy, or found in a patient who is in a high-risk group. One such is, chondromyxosarcoma, a malignant tumor of mesenchymal origin. Chondrosarcomas most commonly occur on axial skeleton and are rarely found in the head and neck.[1]

The clinicopathological findings of this case and a review of chondromyxosarcoma is presented and discussed. The case described is interesting because of its unusual presentation in youth. Myxoid differentiation of this type of tumour is very common and its revelation by a fracture led to a very erroneous pathological diagnosis.[1,2]

CASE DISCUSSION

A male patient of age 22 reported to the Department of our institute with a chief complaint of swelling on the right cheek since the last one week. On further interrogation with the patient, he revealed that developed a swelling on the gingival in relation to 14, 15 and 16, one month back which was small in size. He had scratched the gingival swelling with a twig and since then the swelling had been growing and then was visible as an extraoral swelling on the right cheek. The patient had no relevant medical history and had visited a dentist three years back for oral prophylaxis.

On extraoral examination the patient presented with a solitary, sessile swelling in the right zygomatic region extending superiorly from the infraorbital notch to the corner of the mouth, anteriorly from the ala of the nose to the preauricular area. The swelling was non tender, hard in consistency, pink in color, afebrile and non-fluctuant. No sinus opening and no extraoral deviation was present. There was facial asymmetry seen. On examination of the submental, submandibular, cervical group of lymph nodes, occipital, preauricular, supraclavicular, none of the lymph nodes were found palpable. On intraoral examination a solitary, lobulated, erythematous, exophytic growth measuring approximately 3.5 x 4.5 cm was present occupying the right side maxillary attached gingival in relation to tooth 14, 15, 16 on the buccal aspect, on palpation the growth was tender, firm in consistency, and pedunculated, the growth bled profusely on slightest digital manipulation. The associated teeth was periodontally compromised.

The patient was advised radiographic investigation of computed tomographic scan of face, intraoral periapical radiograph in relation to 11, 12, 13, 14 and 15 and orthopantamograph.

Radiographic investigations were done. The orthopantamograph revealed slight periapical radioluscency in relation to 12 which was later confirmed after the intraoral periapical radiographs in relation to 11 and 12. It showed wide periradicular radioluscency in relation to 12 of thickness 2- 3 mm approximately around the root with widening of periodontal ligament space and loss of lamina dura. The computed tomography of the face contributed much to the diagnosis showing subtle bony erosion with speculated periosteal reaction involving right maxilla and adjacent bony wall of right maxillary sinus with associated lobulated, periosseous, hypodense soft tissue. It also showed mucosal thickening in right maxillary sinus, and few cervical lymph nodes. The differential diagnosis of pyogenic granulaoma, peripheral giant cell granuloma, peripheral ossifying fibroma, capillary lobulated haemangioma was considered.

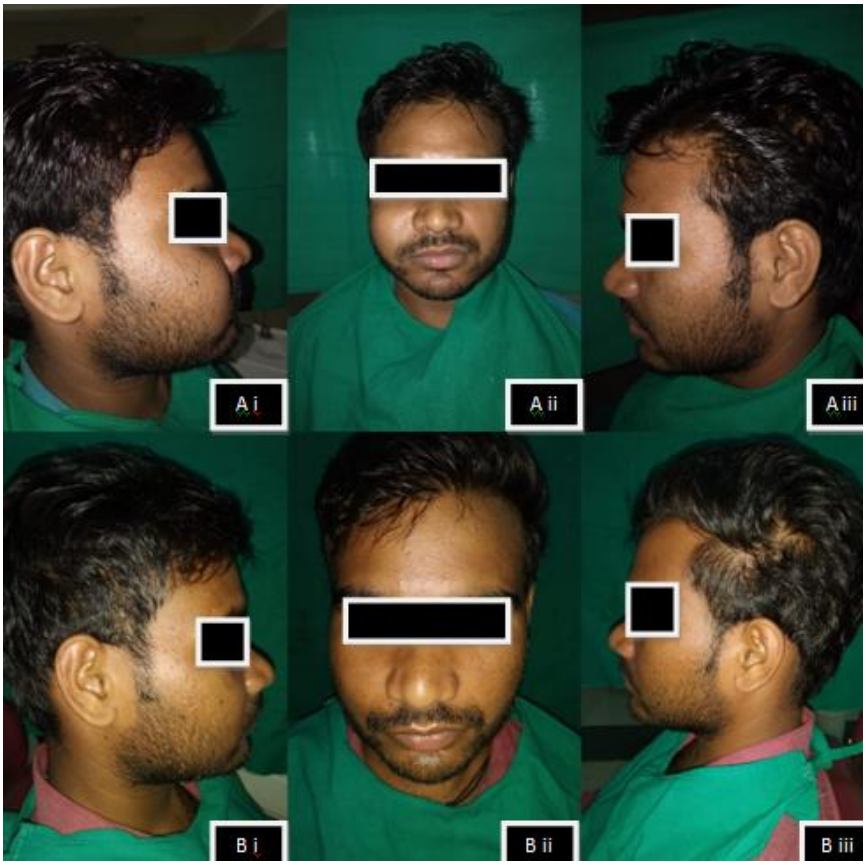
The incisional biopsy of the lesion was done. The gross specimen showed yellowish, friable tumor with lobulated contours, and peripheral sclerosis which may be attributed in our case to a healing fracture. Many spicules extend into soft tissues. There were myxoid nodules separated by fibrous septa and minute haemorrhagic sites were also prevalent.

On histopathological examination of the excised tissue, there was neoplastic changes comprising of lobules of cellular cartilage with nuclear atypia surrounded by spindled stroma. There was fibrous septa dividing the tumor in pools of abundant myxoid and chondroid matrix containing tumor cells.

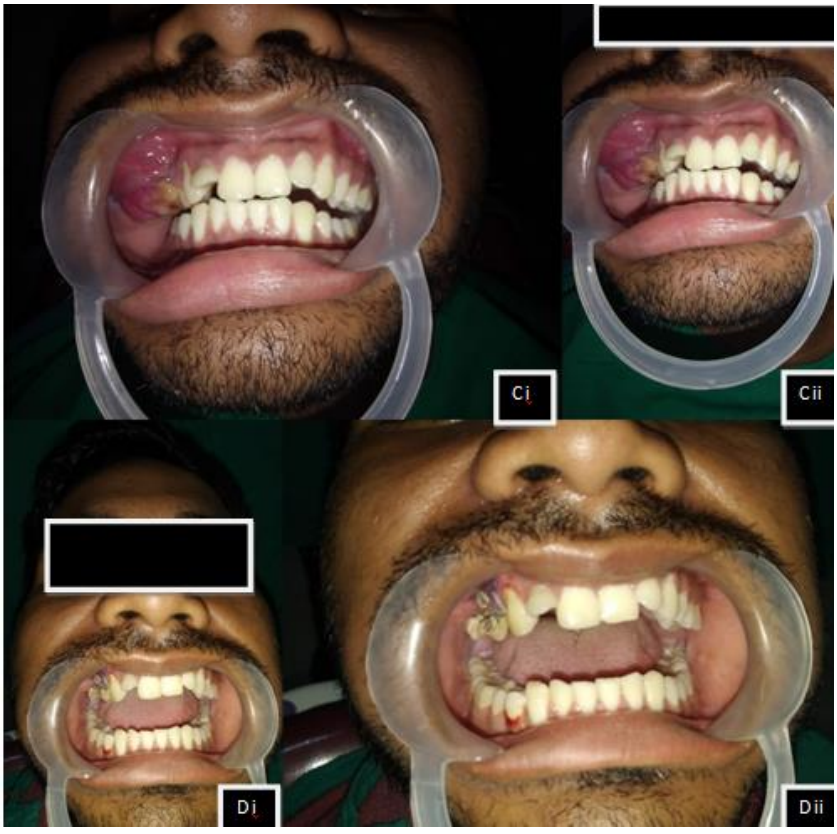
Tumor cells producing cartilaginous matrix, minor or focal atypia were present. Sheaths of chondrocytes present with a lobulated growth pattern under low power. Nucleus spindle shaped with widely interspersed network of fibres and focal mucinous material giving characteristic myxoid appearance with cartilaginous matrix. Abundant myxoid stroma was present which was hypovascular with discernible cartilaginous histology.

The presented lesion was finally diagnosed as Chondromyxosarcoma. It was a moderately differentiated type of chondrosarcoma with myxoid variant suggesting it to be grade2 chondrosarcoma. Histological differential diagnosis comprised of ossifying fibromyxoid tumor, soft tissue chondroma, myxofibrosarcoma, myxoid leiomyosarcoma and soft tissue myoepithelioma.

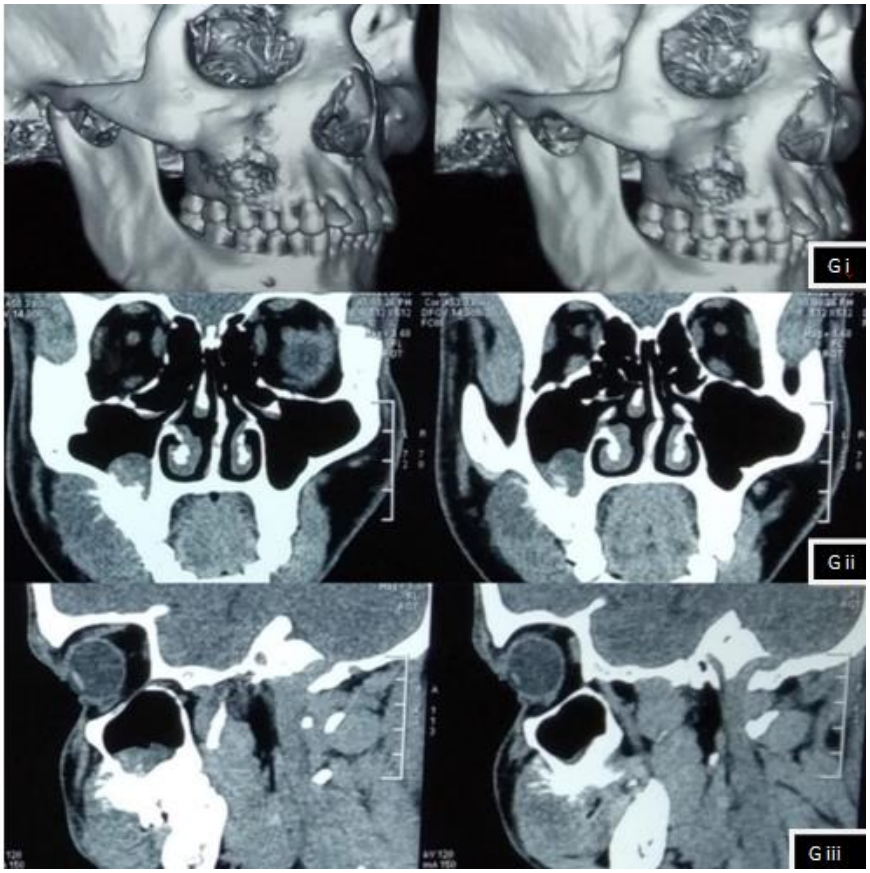
The treatment to the patient was surgical intervention of the soft tissue mass under general anaesthesia along with corticotomy. Buccal advancement flap retracted and closure was done. Patient was put on antibiotic prophylaxis. The excised tissue showed almost same histopathological features as that of the biopsy examination with low mitotic activity of the cells. It was hypocellular and showed finely vacuolated cytoplasm and variable sized nucleoli. The patient is still on follow up and has no recurrence of the symptoms and is undergoing fixed partial denture prosthesis.



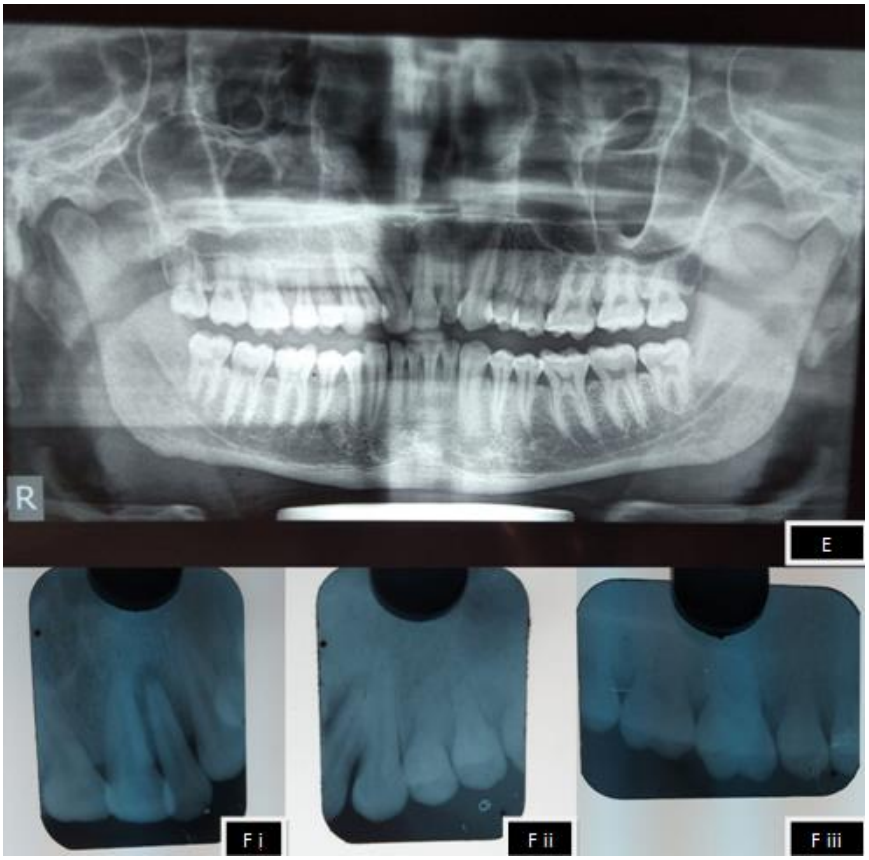
A(i), A(ii) AND A (iii) EXTRAORAL PREOPERATIVE APPEARANCE
B(i), B(ii) AND B(iii) EXTRAORAL POST OPERATIVE APPEARANCE



C(i) AND C (ii) INTRAORAL PRE OPERATIVE APPEARANCE
D(i) AND D(ii) INTRAORAL POST OPERATIVE APPEARANCE

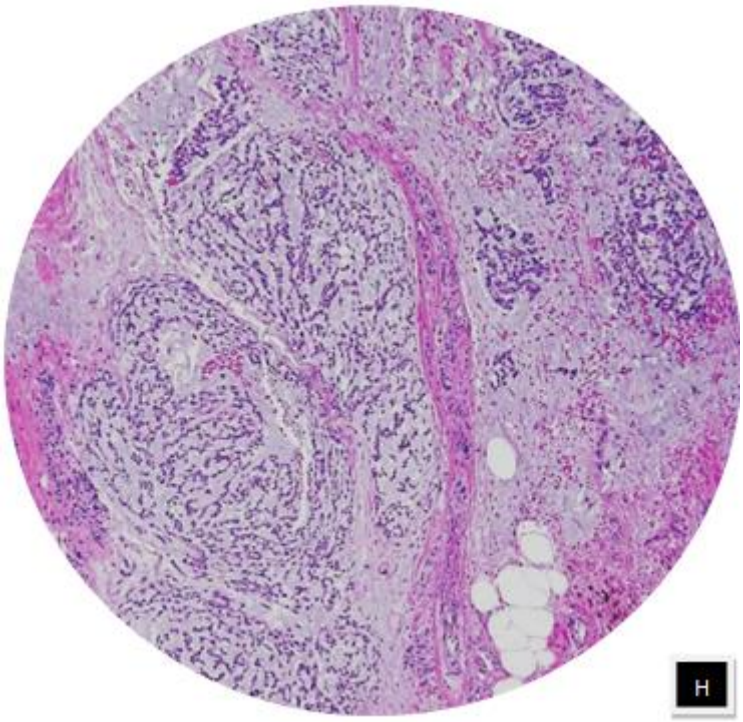


CT SCAN PHOTOS SHOWING EROSION OF THE MAXILLA AT THE SITE OF THE LESION



E: PANORAMIC IMAGE OF THE PATIENT
F(i): INTRAORAL PERIAPICAL IMAGE OF 14

F(ii): INTRAORAL PERIAPICAL IMAGE OF 14, 15 AND 16.
F(iii): INTRAORAL PERIAPICAL IMAGE OF 16 AND 17.



HISTOPATHOLOGICAL SECTION

DISCUSSION

Chondrosarcoma is the malignant form of chondroma, may occur in maxilla or mandible or any other bone in the body with special predilection to be in the maxilla. This tumor shows slight predilection for men in the ratio of 2:1. Large portions of this tissue may appear myxomatous, calcified or even ossified. Chondrosarcomas have slow clinical evolution, may arise from osteochondromas. Usually affected ages between 30-60years. 16% occurs in patients age 20years or less, may be higher grade and at different sites. Conventional tumors are divided by location into central, peripheral and juxtacortical forms. Metastasis is rare and often occurs late. Chondrosarcoma with hyaline cartilage are extremely rare, most of them are myxoid chondrosarcomas.[13] Usually 10-15% of the chondrosarcomas arise secondary to pre-existing condition may be an exostosis, chondrodysplasia and multiple chondromas.

Although the exact etiology of this tumour is unknown, the basic proliferating tissue is cartilage throughout, large portions of it may become myxomatous, calcified or even ossified, sometimes fibrosarcoma like tumours are also found. Pathophysiology of this tumor states that majority of this variant are driven by gene infusions.[3]

The characteristic feature of this tumour is that the malignant cells produce chondroid lacework or cartilaginous network even in small foci, it has slow clinical evolution. Metastasis of chondrosarcoma composed of hyaline cartilages is very rare in somatic soft tissues and occurs very late. Secondary chondrosarcoma arises from osteosarcoma, if primary and secondary chondrosarcoma appears together it causes dedifferentiated chondrosarcoma.[4]

RADIOGRAPHICAL FEATURES

It is seen only when it is long standing, occasional tumors appear radioopaque due to fluffy calcification of neoplastic cartilage, these have poorly defined margins, erosion or thickening of the cortex with usually no periosteal bone formation.[5]

ORAL MANIFESTATIONS

Both primary and secondary chondrosarcoma appears as an expanding lesion which is painless, may occur in maxilla or mandible with primary involvement of the alveolar ridge or sometimes in the maxillary antrum, it also causes resorption or exfoliation of the teeth being invasive, destructive and also metastasizing readily. The mucosa is often stays intact which becomes classic feature of destruction.[4-5]

HISTOLOGICAL FEATURES

Histological features show poorly differentiated cells with a myxoid pattern. These cells exhibit a round or stellate form. They are scattered in an abundant stromal reaction. The only differentiation is chondroid; there is no osteoid formation in tumoral areas. These patterns are quite different from osseous

myxomas which do not exhibit chondroid differentiation and pleomorphism of nuclei.[2]

Grading: based on cellularity and nuclear changes in chondrocytes; well, moderate or poorly differentiated correspond to grades 1 - 3; grade 4 is spindled tumor representing either chondroblastic osteosarcoma or dedifferentiated chondrosarcoma

Well differentiated: less cellular with only a few double nucleated cells and mild / moderate atypia; not well circumscribed, lobulated architecture with abundant cartilaginous matrix separated by narrow fibrovascular bands; tumor cells resemble chondroma; permeate existing trabecular bone and fill marrow space; lie in lacunar space surrounding hyaline cartilaginous matrix; malignant features more obvious at growing edge of tumor; may have reactive thickening of cortex.[2]

Poorly differentiated: marked hypercellularity, extreme pleomorphism with markedly hyperchromatic nuclei; bizarre tumor giant cells and small cells, frequent mitotic figures; usually mixed with other grades; tumor cells destroy cortex and form soft tissue mass.

Grade I chondrosarcomas (50%) can be distinguished only by growth criteria. The nuclei are small and show high chromatin density.

Grade II chondrosarcomas (42%) have medium-sized, regular nuclei with loose chromatin structure.

Grade III Chondrosarcomas (8%) The chondrocytes of grade III cases show polymorphic nuclei. Binucleate forms, the number of mitoses and cellularity all show considerable overlap for all three grades. So far there are no immunohistological and molecular biological methods for reliable differentiation. These have extreme cellularity, large bizarre nuclei, and small foci of spindling at the periphery of the lobule. Sheets of spindle cells with hyaline material and mucous is seen in chondromyxosarcoma.[2,6]

TREATMENT

Surgery is the best possible treatment plan for this tumour as radiotherapy and chemotherapy is not preferred due to the radioresistant nature of this tumour. The prognosis for chondrosarcomas depends on the size, location, grade, and surgical respectability of the tumor. Complete resection is the most effective treatment for conventional chondrosarcomas.[7] Maxillary and antral tumors are more difficult to eradicate and therefore are less amenable to cure. Local recurrence leads to death by direct extension of the tumor into vital structures of the head and neck. Patients presenting with mandibular tumors and with tumors of better differentiated histologic grade enjoy a better survival time. Mesenchymal and dedifferentiated chondrosarcomas are usually treated by chemotherapy because of their aggressive clinical course.[7,8]

CONCLUSION

Thus chondromyxosarcoma being exceedingly rare tumour involves a small percentage of the population in any age group. Being an orphan disease chondromyxosarcoma poses difficulties in identifying indisputable etiological risk factors. These rare tumours get less scientific consideration and financial support than their frequent counterparts. Hence, patients with rare cancers pose particular challenges due to their low prevalence, including mostly incorrect and often late diagnosis, difficulty in accessing clinical skill and proper treatment. Thus enhanced multicentric experimentation is necessary for diagnostic accuracy and better treatment.

DECLARATION OF PATIENTS CONSENT

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published but the anonymity is no guaranteed.

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CONFLICTS OF INTEREST

There are no conflicts of interest

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