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Case Report

Chondrosarcoma of the Maxilla: A case report

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ABSTRACT

Chondrosarcoma is a malignant tumour of mesenchymal origin, after osteosarcoma. It is the second most common bone tumour. It is relatively a slow-growing tumour and is mostly detected in the base of the skull and the anterior maxilla. Here, we report a very distinct case of chondrosarcoma in maxillary sinus. A 65-year-old man reported with a tumour on the right maxillary sinus region. CT imaging showed an abnormal mass destructing surrounding tissues. Our case shows that radiography combined with histopathology is necessary to make the final diagnosis. The presented case revealed that chondrosarcoma can be a heterogeneous tumour.

1. Introduction

Chondrosarcoma is a malignant tumour of mesenchymal origin, after osteosarcoma. It is the second most common bone tumour. It usually occurs in iliac bone, ribs, and long bones. Chondrosarcoma most commonly affects adults, with no sex predilection.[1] Chondrosarcomas usually arise de novo; although, these are common in patients with osteochondromas, Maffucci syndrome, Ollier's, and Paget's disease.[2–4] There are three grades of the malignancy, depending on cellularity, atypia, mitotic activity, nuclear size, and surrounding matrix composition.[5] The rate of metastasis depends on the grade of the tumour. There is 70% risk for grade 3 and 10% risk of metastasis for grade 1. It usually metastasise to the lungs.[5] The survival rate is about five years in 90% cases for grade 1, 81% for grade 2, and 43% for grade 3.[6]The diagnosis of chondrosarcoma is among the most difficult in tumour pathology. Histological changes in the small biopsy specimen may be insufficient for definitive diagnosis. In such cases, CT and MRI are required. 12% of all chondrosarcomas are located in the head and neck region and occur mostly in adults between the third and sixth decade of life. Patients may present nonspecific symptoms, which vary greatly depending on the location of the tumour.[5–7] Primary sites of the head and the neck include larynx, thyroid cartilage and arytenoids.[8] However, chondrosarcomas can occur in all other sites of the craniofacial compartment in which cartilage is found, such as mandible, paranasal sinuses, nasopharynx, base of the skull, and maxilla.[9,10] The treatment of choice is wide surgical excision of all the involved structures with negative margins and preservation of function if possible. To prevent the local recurrence Radiotherapy is indicated. The crucial factors determining the prognosis of chondrosarcoma of the maxilla are the anatomic localisation, size, extension, and histological grade of the tumour.[11] Five-year survival rate for chondrosarcomas of the jaw and facial bones in only 67.6% individuals. However, life-long follow-up is essential, because chondrosarcomas show a wide variation in time of recurrence and metastasis.[12]

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2. Case Report

A 65-year-old male patient reported to the Department of Oral & Maxillofacial Surgery with the chief complaint of swelling and pain in the right zygomatic region for last two months. The history of present illness revealed that the patient was apparently asymptomatic two months back, after then patient felt the pain and noticed swelling over the right side of his jaw. The swelling has been increasing in size gradually since past two months and painful.

The patient had no positive, associated medical and family history. He had a history of tobacco chewing in the form of gutkha 4-6 packets/day for the last 30 years.

On extra oral examination, a diffused swelling was seen in the right middle third of the face (zygomatic region) measuring approximately 5 cm x 6 cm. It was tender on palpation. Swelling was extended superiorly upto infraorbital margin, inferiorly upto the angle of mouth, anteriorly upto nasolabial fold and posteriorly upto the anterior border of ramus. Overlying skin appeared normal and tender on palpation (Figure 1).

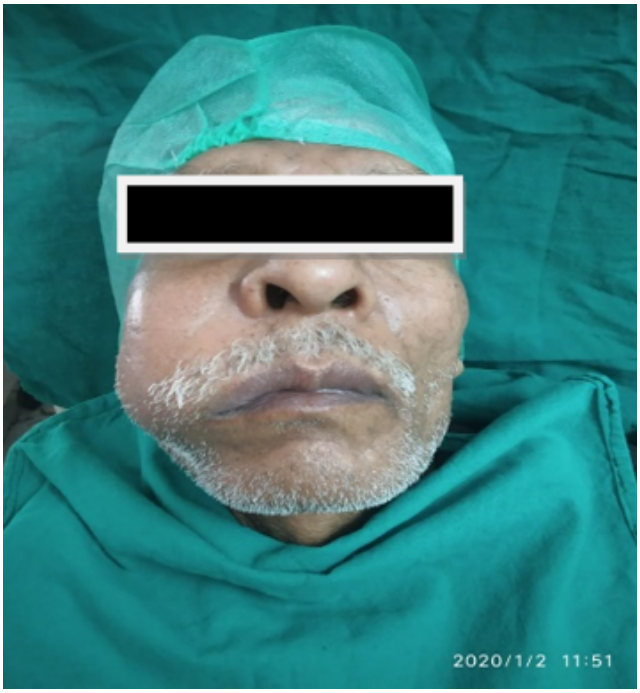

Fig. 1:

The intraoral examination revealed a growth measuring 5 cm x 6 cm in dimension, with respect to 14, 15, 16 and 17 teeth buccally and it also extends palatally. Mobility was also present in these teeth. The necrotic slough was present over the growth and it was bleeding spontaneously on examination (Figure 2).

The patient was advised for NCCT face with 3D CT reconstruction

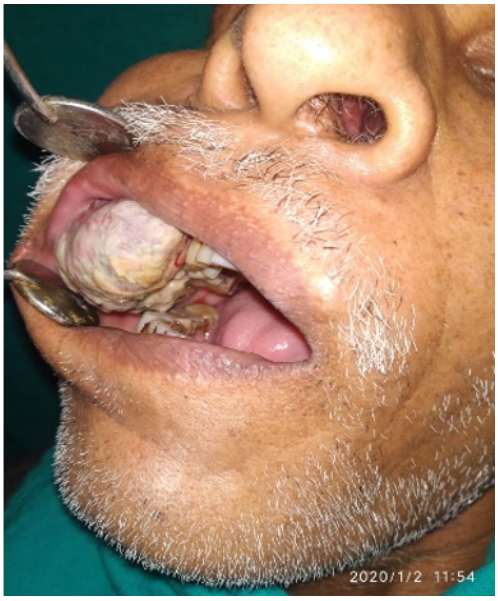

Fig. 2:

3. Radiographic Investigation

A computed tomography (CT) scan showed right maxillary soft tissue mass occupying entire maxillary cavity causing anterior and lateral bony wall breach and extending to the soft tissue of the cheek (Figure 3). It was breaching the medial wall inferiorly into the oral cavity in the right canine region and extending to the right side of upper lip and nasal spine region. However, no orbital and nasal extension of this soft tissue mass is seen. Bone is breached into anterior and inferior nasal compartment. It was also involving the maxillary sinus, right maxillary space, the nasal cavity, and hard palate. It was also inferiorly extending into the oral cavity (Figure 4).

Considering the patient history, clinical examination and radiographic examination a provisional diagnosis was made of peripheral giant cell granuloma, central giant cell granuloma and carcinoma of maxillary antrum.

A biopsy was deemed mandatory. An incisional biopsy was performed under local anaesthesia in the department of oral and maxillofacial surgery.

4. Histopathological Examination

On microscopic examination, the given haemotoxylin and eosin (H&E) -stained soft tissue section showed connective tissue stroma that is richly cellular. The cells are round to spindle shaped with hyperchromatic nuclei, nuclear pleomorphism, altered N/C ratio and few abnormal mitotic figures.

Few double nucleated cells are seen with mild atypia and surrounding cartilaginous matrix. There are numerous haemorrhagic areas surrounded by acute and chronic

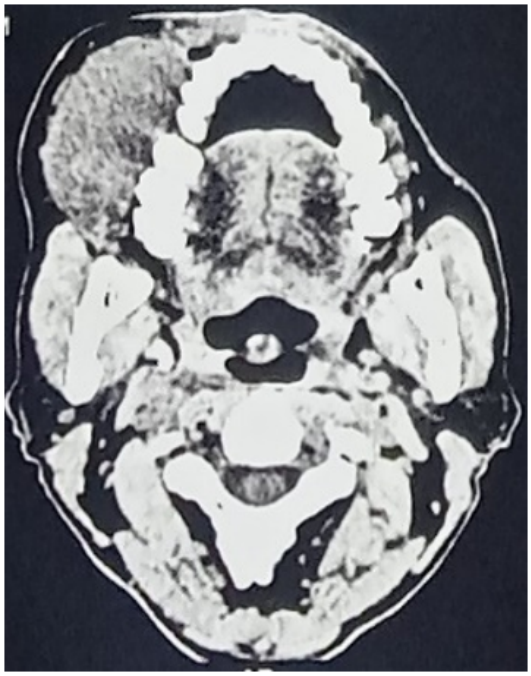

Fig. 3:

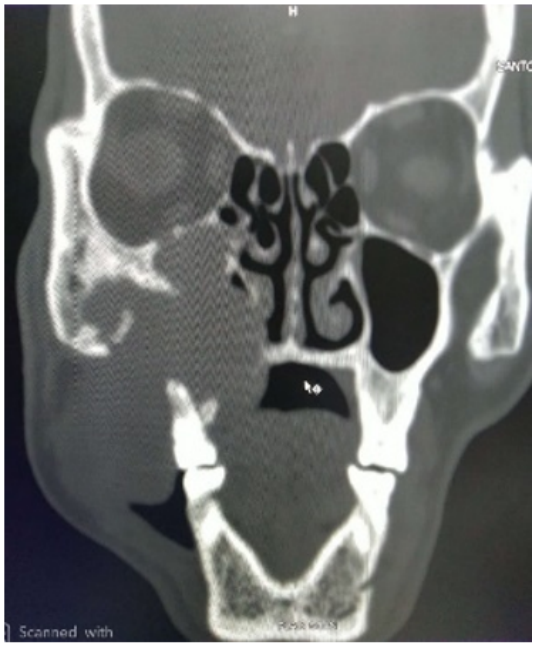

Fig. 4:

inflammatory infiltrate.

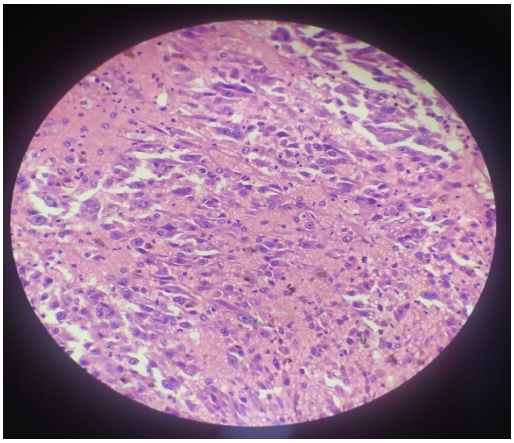

Fig. 5: (High magnification)

The overall histopathological features are suggestive of malignant connective tissue tumour– Chondrosarcoma.

5. Discussion

Patients with chondrosarcoma of the jaws have no sex predilection. Maxillary tumours are difficult to eradicate. Local recurrence leads to death by direct extension of the tumour into vital structures of the head. The characteristic features of chondrosarcoma of the maxilla is painless swelling, paraesthesia, trismus, and mobility of the teeth.[11] Diagnosis of the chondrosarcoma is among the most difficult in head and neck tumour pathology. This bone tumour is usually discovered on radiographs. In some cases, it can be difficult to differentiate between benign and malignant lesions. Additional techniques such as CT scan and MRI can deliver more information about the nature of the tumour mass. In the majority of cases, biopsy of a tissue sample and histopathological examination brings the final diagnosis of chondrosarcoma. In some cases there are problems in distinguishing chondroma from low-grade chondrosarcoma. Chondroma is a tumour composed of mature hyaline cartilage with the highest prevalence in the fifth to seventh decade of life. It usually occurs in the mid-line of the axial skeleton and is generally asymptomatic. It can be called enchondroma when it occurs within the medullary cavity of tubular bones. The cells resemble normal chondrocytes and produce cartilaginous matrix. The histological appearance of enchondroma could be similar to grade 1 chondrosarcoma. In both cases, calcification and ossification are common. However, in higher grades, chondrosarcoma displays significant hypercellularity and atypia. Cells of chondrosarcoma are often binucleate with clumps of chromatin within the nucleus. In the Chondrosarcoma, bone is usually infiltrated.[13] Chondrosarcoma is graded from 1 to 3. Grade 1 chondrosarcoma grows relatively slowly and the cells

resemble unaffected ones. In grade 2, dysplasia is more prominent. The amount of chondroid matrix is significantly reduced. In grade 3, pleomorphic cells show the highest mitotic activity.[1,5,14] There is a high tendency towards both local and distant recurrence.[15] Histologically, the tumour shows connective tissue stroma that is richly cellular. The cells are round to spindle shaped with hyperchromatic nulei, nuclear pleomorphism, altered N/C ratio and few abnormal mitotic figures. There are numerous haemorrhagic areas surrounded by acute and chronic inflammatory infiltrate. In this type of critical cases before making the final diagnosis, radiography and histopathology are necessary along with detailed clinical history.

6. Conclusion

In the present case, the patient presented all the necessary diagnostic criteria. The present case is unique as a "Chondrosarcoma" of maxilla which invaded right maxillary sinus, causing anterior and lateral bony wall breach and extending to the soft tissue of the cheek. A thorough lesional examination and a long-term follow-up should be done to ascertain the prognosis.

7. Source of Funding

Nil.

8. Conflicts of Interest

None.

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