Chondrosarcoma of Maxilla - A Rare Case Report

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Abstract

Rationale: Chondrosarcoma, although being a rare entity in jaws, may turn fatal if left untreated or inadequately excised. Prognosis in terms of 5-year survival rate ranges from 90% for Grade I, 81% for Grade II and 43% for Grade III respectively. **Patient Concerns:** A 35-year-old male patient reported with a gradually progressive hard painless growth over right maxillary molar region. His main concern was removal of pathology without long-term morbidity. **Diagnosis:** Computed tomography revealed ill-defined mass with internal calcification involving posterior half of upper right alveolus. **Treatment and Outcomes:** Mandatory biopsy suggested benign chondroma, however wide excision and infrastructural maxillectomy revealed Grade II chondrosarcoma. **Take-away Lessons:** Complex anatomy of maxilla renders surgical excision of chondrosarcomas with histological clear margins, a daunting task. Due to misdiagnosis of preoperative biopsy, suboptimal excision of malignant mass may lead to local recurrence and occasional distant metastasis. This necessitates further therapy and long term follow up, with occasional poor patient compliance.

Keywords: Biopsy, chondroma, chondrosarcoma, maxilla

Introduction

Chondrosarcomas constitute a rarity in entire head and neck region accounting for 5.6% cases of malignant tumours.[1] Few cases have been reported in cervicofacial region in descending order-anterior maxilla, skull base, cervical vertebrae and nasal cavity.[2] They are malignant mesenchymal tumours which arise from remnants of embryonic cartilage precursors from nasal septum in anterior part of maxilla and from Meckel's cartilage in posterior aspect of mandible.[3]

Diagnosis of chondrosarcoma is amongst the trickiest in tumour pathology. Depending upon size of biopsy, diagnosis may vary. This discrepancy between biopsy and wide excision is highlighted in this paper, where a seemingly benign tumour of maxilla, upon infrastructural maxillectomy without neck dissection, revealed Grade II chondrosarcoma.

Case Report

Patient concerns

A 35-year-old male reported to Department of Oral Maxillofacial Surgery with a history of painless growth in right maxillary region since 6 months which later progressed slowly over a period of 2 months. His primary concern was removal of pathology, no external scar, preservation of facial contours and no long-term morbidity. There was no associated systemic illness, nor any destructive habits.

Extraoral examination revealed right malar disfigurement, nasolabial fold obliteration and intact visual acuity. Paraesthesia was present on right infraorbital region with no cervical lymphadenopathy. Intra-oral examination revealed lobulated growth of approximately 4 cm × 3 cm in size, involving right maxillary canine to molar region obliterating buccal vestibule. It was multinodular, fixed, nontender, firm in consistency and overlying mucosa appeared inflamed [Figure 1]. There was no pus discharge although root pieces of right maxillary molars were seen with marked mobility.

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Diagnostic aids

Routine blood investigations were within normal limits. Panoramic radiograph revealed a mixed lesion with diffuse margins, which had displaced the roots of the right maxillary molars. Contrast enhanced computed tomography (CT) revealed ill-defined tumour mass with internal calcification, involving posterior half of upper right alveolus, projecting along antero-inferior aspect of right maxilla without crossing the midline [Figure 2]. Chest X-ray showed no abnormality.

Incisional biopsy revealed benign chondroid tissue, having lacunae with round nuclei. Calcification along with fibrous and osteoid tissue having occasional nuclear and cellular pleomorphism was observed.

Treatment and outcomes

Surgery was planned under general anaesthesia after biopsy confirmation of chondroma, where wide excision and infrastructural maxillectomy was performed intraorally [Figure 3]. The surgical defect was secondarily closed with medicated gauze pack placed under palatal surgical splint fabricated preoperatively [Figure 4]. Histopathological examination revealed moderately differentiated chondrosarcoma-Grade II. Immunohistochemistry was suggested to the patient which he denied.

Followup

Patient was referred to oncology centre due to one positive surgical margin for further treatment and according to them, no local recurrence or distant metastasis was observed after 1 year follow up.

Macroscopic and microscopic findings

Macroscopically specimen measured 7 cm × 5.5 cm × 4 cm whereas exophytic tumour mass identified measured 4 cm × 3 cm × 2 cm. Microscopic examination revealed moderately differentiated tumour which had plump cartilaginous matrix cells with hyperchromatic nuclei. Tumour cells were arranged in lobulated architecture with abundant cartilaginous matrix separated by fibrovascular stroma suggestive of moderately differentiated chondrosarcoma-Grade II [Figure 5].

Discussion

Chondrosarcomas are malignant mesenchymal tumours characterized by formation of cartilage by tumour cells. Primary chondrosarcomas arise independently, whereas secondary chondrosarcomas arise from preexisting enchondroma or osteochondroma. In contrast to osteosarcoma, it is uncommon in first 2 decades of life. The mean patient age ranges from 35 to 45 years and male to female ratio is 1.2:1.[1,3]

The most common clinical finding is painless expansile swelling of both cortices with exfoliation of teeth. Painful ulceration at later stages with rare lymphnode involvement can be seen. Headache, blurred vision, proptosis and diplopia are less frequent findings. It can also cause nasal stiffness or epistaxis. Clinical features of chondrosarcomas like paresthesia and dysesthesia, are used to differentiate a malignant neoplasm from osteomyelitis.[4,5]

Conventional radiographic findings include irregular intramedullary radiolucencies interspersed with punctuate radiopacities, expansion and destruction of cortical plates, periodontal ligament space widening and occasional sunburst appearance at periphery. CT scan demonstrates an ill-defined cloud-like matrix with calcified whorls and arcs.[4] The tumour may grow in a lobular pattern with minimal or no foci of calcification in some cases and it may appear as a multilocular radiolucency mimicking a benign lesion.[1]

Histologically, chondrosarcoma can be classified according to the microscopic appearance into conventional, clear cell, myxoid, mesenchymal and dedifferentiated.[2] Histological grading given by Evans *et al*. is an important determinant of prognosis and progress of the disease [Figure 6].[6]

In chondrosarcoma, bone formation takes place on framework of preexisting cartilage matrix; whereas in osteosarcoma, it is directly by the malignant stromal cells.[1] Distinguishing chondroma from low-grade chondrosarcoma is sometimes challenging, as seen in our case. Chondroma is a tumour composed of mature hyaline cartilage, usually occurring in mid-line of axial skeleton and is generally asymptomatic. Histological appearance of enchondroma could be similar to Grade I chondrosarcoma but in higher grades, chondrosarcoma displays significant hyper-cellularity and atypia.[7]

Chondrosarcoma is generally treated with multimodal approach like wide en-bloc resection, local curettage, cryotherapy, chemotherapy, radiotherapy and immunotherapy. A wide surgical resection with clear margins, however remains the gold standard for treatment.[4] These lesions are radio-resistant and therefore radiotherapy is not generally recommended as a primary modality, however it may be indicated for incompletely resected (Positive surgical margin) high-grade or unresectable tumours as well as an adjunct or a form of palliative therapy for recurrent lesions. Doses of 50 Gy preoperatively and 60–66 Gy postoperatively for close or positive margins are typically used.[5] Spot-scanning beam proton therapy and intensity modulated radiation therapy were recently tested in chondrosarcoma of the skull base. These techniques are relatively safe and can be an effective adjuvant to surgery when used at high doses.[8]

The intralesional excision – curettage of large lesions, combined with local radiation therapy can be cautiously advocated, but only in low grade variants. Cryosurgery is also suggested for treatment of Grade I chondrosarcoma but it can be associated with complications such as infection, embolism, and neuropathy.[2]

Chemotherapy, before or after surgery is controversial and is usually reserved for mesenchymal, undifferentiated high-grade tumours. As a neoadjuvant treatment, it inhibits tumour

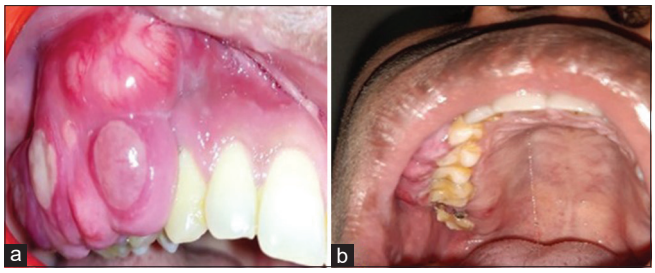

Figure 1: Intra oral view shows extension of lesion (a and b)

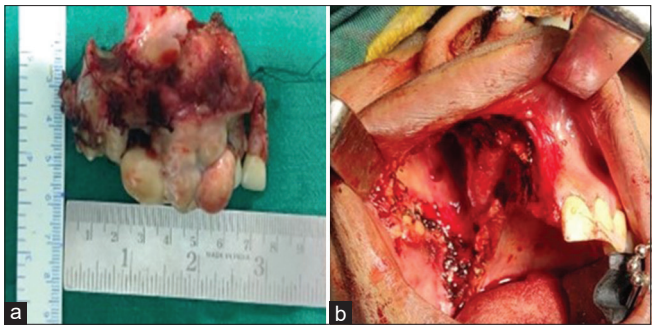

Figure 3: Intraoperative view shows specimen (a) residual cavity (b)

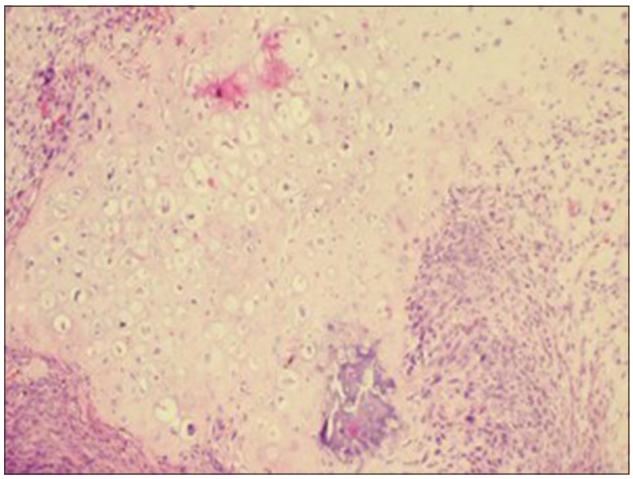

Figure 5: Photomicrograph showing the presence of increased proliferation of chondrocytes within lacunae with increased nuclear atypia (H and E; original magnification, ×10)

growth and progression. However, it is not beneficial in improving long-term survival nor distant metastasis control.[4] Current guidelines of the National Comprehensive Cancer Network (NCCN, version 1.2018) indicate that conventional chondrosarcomas (Grades I–III) have no known standard chemotherapy options.[9]

Positron emission tomography-CT has higher sensitivity and specificity than conventional imaging in detecting distant metastases, with exception of pulmonary nodules. The rate of metastasis depends on the grade of the tumour. There is a 10% risk of metastasis for Grade I and II and 70% for Grade III tumours.[10]

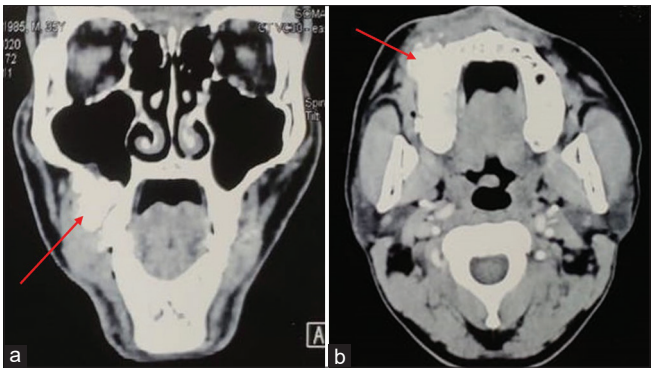

Figure 2: Computed tomography scan shows bucco-palatal extension of lesion and osteolytic destruction of the upper right maxillary alveolus with ill-defined margin which extending into the right maxillary sinus area (a and b)

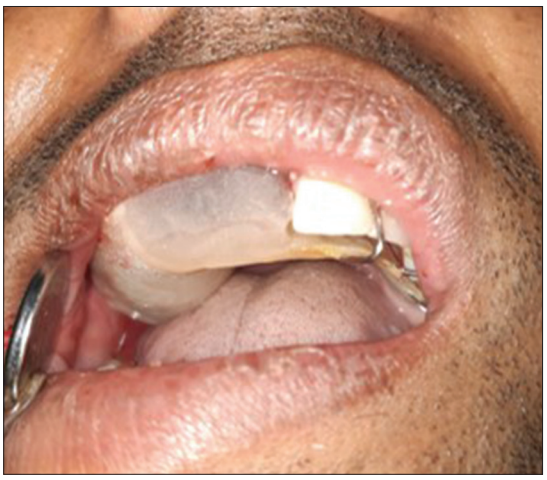

Figure 4: Obturator given

Grade I	Well-differentiated (low-grade)	Small, densely staining nuclei often with multiple nuclei within one lacune
Grade II	Moderately differentiated (intermediate grade)	Increased cellularity, significant amount of cells having moderately sized nuclei, but demonstrate a low mitotic rate of less than 2 mitoses per HPF (also includes myxoid chondrosarcoma)
Grade III	Poorly differentiated (high-grade)	More than 2 mitoses/ HPF, nuclear size generally greater than seen in grade II (also includes dedifferentiated chondrosarcoma)

Figure 6: Histological grading of chondrosarcoma given by Evans *et al.*[6]

Prognostic factors for patients with head and neck chondrosarcoma are histological grading, anatomical location and adequacy of surgical resection. Prognosis in terms of 5-year survival rate ranges from 90% for Grade I, 81% for Grade II and 43% for Grade III chondrosarcoma respectively.[3,7]

Conclusion

Chondrosarcoma of the maxilla is a rare and perplexing neoplasm with a high local recurrence rate. Primary wide excision with clear surgical margins is the treatment of choice, whereas long-term clinical and radiological follow ups are mandatory.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient have 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 and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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