CASE REPORT

Mesenchymal Chondrosarcoma of Maxilla: A Rare Case Report and Review of Literature

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ABSTRACT
Mesenchymal chondrosarcomas are rare malignant neoplasms that can arise from both soft and hard tissues. They are distinct tumors arising in unicentric or multicentric locations. They reveal unusual clinical behavior, characteristic histopathological features, and poor prognosis with late recurrences. Here is a case report of a rare case of mesenchymal chondrosarcoma arising in a 19-year-old female patient’s right maxilla.

Keywords: Mesenchymal chondrosarcoma, De-differentiated chondrosarcoma, Condrosarcomas of jaw, Hemangiopericytoma, Recurrent lesion, Metastasis.

INTRODUCTION
Mesenchymal chondrosarcoma is one of the most unusual, rare malignant cartilaginous tumors with unique histopathological appearance and biological behavior.1 Only around 50 cases affecting the jaws were reported in literature.2,3 It was first described by Lichtenstein and Bernstein in 1958 as a separate entity.1 It develops from pluripotent, mesenchymal stem cells and can differentiate into angioblastic, fibroblastic or cartilaginous structures.1 Mesenchymal chondrosarcoma arises from soft tissues or bone in the ratio of 1:2 to 1:6.4 These lesions affect females more commonly than males (4:1).2 The most affected region is the facial skeleton, especially the jaws, other bones are also affected. Jaw lesions appear in the 2nd and 3rd decades of life.5 Histologically characterized by clusters or sheets of highly undifferentiated small ovoid cells with zones of neoplastic cartilage.4 They reveal local aggressive behavior and highly metastatic and poor prognosis, which require wide surgical excision with adjunct radiotherapy and chemotherapy. These neoplasms occasionally may arise multicentrically.6

CASE REPORT
A 19-year-old female patient was reported to the Department of Oral Medicine and Radiology, Ame’s Dental College and Hospital, Raichur with a complaint of painless swelling in the right maxillary posterior region since eight months. History revealed that the patient was aware of swelling in the maxillary posterior region, which was gradually increasing in size since eight months. The swelling was not associated with pain except for discomfort. There was no history of discharge from the swelling. She had no significant dental, medical and family history. Extraoral examination reveals asymmetry of face due to diffuse swelling in the right middle 1/3 of face causing slight obliteration of nasolabial fold. Skin over the swelling appears normal; on palpation no local rise of temperature, nontender, and firm to hard in consistency. Intraorally, a well-defined swelling was present in the palatal aspect of right premolar and molar region measuring about 4 × 3 cm in diameter extending anterior posteriorly from canine to tuberosity and from palatal marginal gingiva to midline (Fig. 1). Mucosa over the swelling appears slightly inflamed with no discharge, and margins of the swelling was well defined. Hard tissue examination reveals no caries teeth, no pathological mobility of teeth, nontender on percussion, except for mild stains and calculus. After a thorough clinical evaluation, a provisional diagnosis of ameloblastic fibroma was made. After negative aspiration, an incisional biopsy was done under LA from right maxillary palatal mucosa, which revealed ameloblastic fibroma. 3D CT and plain CT scan in both axial and PNS section shows a wide radiolucency with areas of fine and coarse calcifications involving right maxilla extending towards ethmoid sinus medially, and into the orbit superiorly, into the pterygoids posteriorly and into the skin laterally through the infraorbital foramen (Figs 2 and 3). The patient was treated with right total maxillectomy. The excisional biopsy report concluded the diagnosis of mesenchymal chondrosarcoma (Fig. 4). Later, patient was treated with chemotherapy and radiotherapy. The patient was followed for the period of one year and no recurrence was observed. Later she did not return for follow-up; after two years, it was known from her relatives that she expired of lung metastasis.

Fig. 1: Intraoral photograph showing swelling in the right premolar region
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Odontogenic fibroma, chondromyxoid fibroma, fibro sarcoma and angiosarcoma on incisional biopsies. They show no specific clinical signs, symptoms and usually have painless swellings (53%) as in the present case, however painful mass (16%) has also been reported. Few patients develop neurological disturbances, such as facial paresthesia and lip pareses. In some cases, dental extractions, any local surgery or even biopsy may provoke rapid growth. The most common radiographic appearance of mesenchymal chondrosarcoma of jaw is radiolucent shadow, which is difficult to distinguish from other sarcomas. In present case, there is wide radiolucency with fine and course radiopacities involving complete right maxilla. It was reported in previous literature that on CT scan, the tumor presents as a well-defined mass with multiple areas of fine and course calcification. FNAC features have been also less described in literature, in the present case, there was negative aspiration. Mesenchymal chondrosarcoma is usually considered to be a well-delineated mass and can be easily demarcated from the surrounding tissue, as in our case, it was nodulated and easily separated from surrounding bone. Histologically, diagnosis of mesenchymal chondrosarcoma is not difficult because of characteristic presence of highly cellular, undifferentiated zones with islands of chondroid differentiation. A correct diagnosis, however can be difficult in small sample of biopsy, which may contain only one of the two components. Because of rich vascular component, it may be misdiagnosed as hemangiopericytoma. In present case, there was malignant cartilage surrounded by an islands of anaplastic small, round or ovoid cells.

There is diversity of opinion in the treatment of mesenchymal chondrosarcoma, the general consensus suggests that surgical excision with wide margins along with chemotherapy and radiotherapy is suggested. The present case was treated with total right maxillectomy followed by chemotherapy and radiotherapy.

Prognosis of mesenchymal chondrosarcoma is poor because the tumor has a tendency for late recurrence either locally or metastasis by hematogenous route. With lungs being the most common site.

REFERENCES


DISCUSSION

Mesenchymal chondrosarcomas are rarely reported in literature due to their rarity of incidence. Both skeletal and extraskeletal lesions have been reported in the head and neck region. Extraskeletal tumors commonly arise from the orbit, meninges, nasal and paranasal mucus. Among the facial skeletal region, jaws are most commonly involved with more preference to maxilla than mandible as presented in this case. They are misdiagnosed as...