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Chondrosarcoma of the maxilla: a case report and literature review

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INTRODUCTION.

Maxillofacial chondrosarcoma (MC) is a malignant maxillofacial bone and cartilage tumor, and account for approximately 3-4% of all chondrosarcomas. MCs are highly invasive. The symptoms are non-specific, depending on the site of origin. Local relapse or persistent tumor are the main causes of death, so they must be excised as early and as widely as possible. The study describes a rare case of maxillary chondrosarcoma and reviews the literature of maxillary chondrosarcomas, from 1962 to 2017.

CASE REPORT.

A 27 year-old white woman was referred to the Department of Odontostomatologic and Specialized Clinical Sciences, Marche Polytechnic University, Ancona, complaining swelling and pain at the left anterior maxillary elements 2.1-2.2-2.3, since several months. The patient reported having previously undergone the extraction of element 2.2. The residual cavity and the alveolus were filled with heterologous material and a Maryland bridge was fabricated as a temporary prosthetic solution. No histopathological examination of the removed sample was required . To our observation, the extraoral examination revealed a moderate swelling on the left side of the lip. On intraoral investigation, an irregular lobulated swelling was observed, involving the anterior left side of the palate, extended from element 2.1 to 2.3. The Computed Tomography (CT) imaging, showed an irregular soft tissue mass, causing osteolytic destruction of left upper maxillary alveolus. The intraoral radiograph showed a radiolucent area around the elements 2.1-2.2-2.3. An excisional biopsy was performed and the material was sent to the Institute of Pathology, Marche Polytechnic University, Ancona, for histological examination. Furthermore, a contrast CT was requested. Macroscopically, the major specimen appeared as a whitish nodule of cm 1. Microscopically, the lesion showed a lobular growth of cartilaginous differentiation cells with focal presence of osteoid, marked atypia and atypical mitoses. It was partially surrounded by fibrous tissue, focally interrupted and passed. Among the most probable hypotheses (chondroblastic osteosarcoma and chondrosarcoma), the diagnosis of Chondrosarcoma grade II was released (4th Edition WHO, 2017). The last CT imaging showed an expansive, osteolytic and infiltrative lesion in the median e para-median portion of the left upper maxillary bone, extending on the underlying alveolar arch, from the element 2.1 to 2.4. The osteolytic component of the lesion determined the erosion of the maxillary left frontal process. The incisive canal was involved and the lesion reached until the left nasal fossa. The fleshy component was about cm 3.2 of diameter, deforming the nose wing, the sulcus naso-labialis and the left cheek. Moreover, it caused deviation of the left nasal septum. The patient underwent to left total and right subtotal maxillectomy and bone reconstruction was performed using an autologous graft harvested from fibula. Several plastic surgeries followed the reconstructive therapy.

RESULTS.

The literature review (1962-2017) revealed 58 cases of MCs. The tumor prefers the third decade (38.9%) and the female gender (59.3%). Swelling is the most common presenting manifestation, reported in all cases. The duration of the symptoms range from 2 to 720 weeks. Macroscopically, most cases appear as a hard and firm mass. The maximum tumor size range from cm 0.5 to cm 12, with a mean maximum diameter of cm 4.5. Histological examination reported the diagnosis of mesenchymal chondrosarcoma, chondrosarcoma, peripheral chondrosarcoma and dedifferentiated chondrosarcoma in 71.2%, 17%, 10.2% and 1.7% of cases, respectively. Total relapses amount to 23.4%, during a follow-up ranged between 5 and 320 months. 45.5% of dead patients (18.6%) had disease recurrence.

CONCLUSIONS.

The appropriate management requires an early and widely excision, which guarantees adequate healthy tissue margins. Chondrosarcomas only occasionally respond to radiotherapy and the chemotherapy appears to be of little value. The prognosis of the MC is generally worse than that for other sites, due to the difficulties of adequate excision. Maxillary tumors have a even worse prognosis than mandibular ones, because of the diagnostic delay, the proximity to the central nervous system, and the difficulty to resection. The 30-year survival rates is 43% for patients with localized disease, 22% for patients with regional metastasis, and less than 10% for patients with distant metastasis. Histological grade and complete resection with unharmed surgical margins are the most important prognostic factors. Since relapses often are a late sequela, patients must be followed for their lifetime.

REFERENCES.

1. *Mahajan AM, Ganvir S, Hazarey V, Mahajan MC. Chondrosarcoma of the maxilla: A case report and review of literature. J Oral Maxillofac Pathol. 2013 May; 17(2):269-73.*

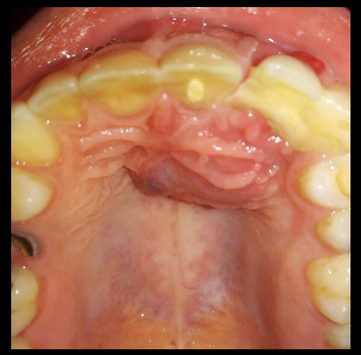

Fig.1: Palatal swelling from 2.1 to 2.4.

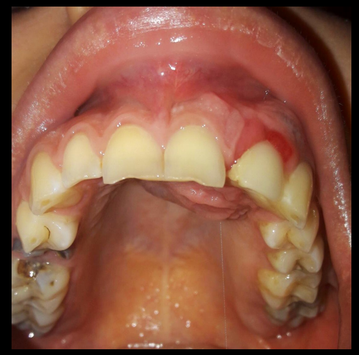

Fig.2: Buccal aspect of the lesion.

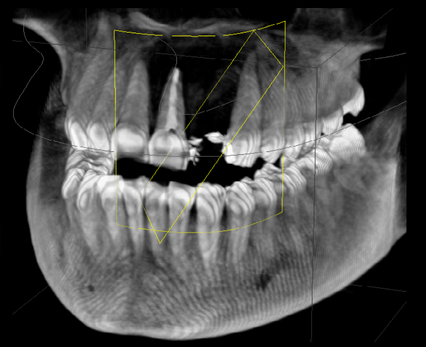

Fig.3: 3D-CT imaging

Features	
Gender	• F: 59.3% ; • M: 41.7%;
Age (y)	• Range: 8-102; • Mean:31.9; • Peak: III decade; 38.9%.
Symptoms	• Swelling: 100%; • Teeth displacement: 32.2% • Epistaxis: 20.3% • Nasal obstruction: 18.6% • Pain: 11.9%
Presentation	• Hard and firm mass: 80%; • Ulcerated mass: 10% • Soft mass: 10%
Duration	• Range: 2 weeks- 60 years;
Size (cm)	• Range: 0.5-12; • Mean:4.5
Histological diagnosis	• Mesenchymal chondrosarcoma: 71,2%; • Chondrosarcoma: 17% • Peripheral chondrosarcoma: 10.2% • Dedifferentiated chondrosarcoma: 1.7%
Follow-up (m)	• Range: 4 – 320;
Recurrence (%)	• Yes: 23.4 • No: 57.4 • Died: 18.6

Table 1. Main features of maxillary chondrosarcoma literature cases.

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