# SARCOMA MAXILLAE SUPERIORIS

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**Abstract: Sarcoma of the maxilla (Sarcoma maxillae superioris)** is a group of malignant, rapidly growing, and highly invasive tumors of mesenchymal origin that arise from the maxillary bone and adjacent soft tissues, characterized by destruction of bone, rapid spread to surrounding anatomical structures, and early hematogenous and lymphogenous metastasis. Sarcomas of the jaws most often originate from the medullary (bone marrow) portions of the jaw bones. Sarcomas developing from the elements of bone tumors are relatively rare.Among jaw sarcomas, spindle-cell (fusiform) and round-cell sarcomas, reticulosarcomas, chondrosarcomas, fibrosarcomas, and myxosarcomas are encountered more frequently.Sarcomas usually develop in younger patients compared to carcinomas. They are characterized by rapid growth, less pronounced inflammatory signs, and an early tendency to metastasize. Determining the stage of sarcoma development is of great diagnostic importance.

**Keywords:** malignant, lymphogenous metastasis, chondrosarcomas, fibrosarcomas, myxosarcomas, reticulosarcomas.

## Introduction

Maxillary sarcoma (Sarcoma maxillae superioris) is a malignant mesenchymal tumor arising in the maxillary bone and adjacent soft tissues, characterized by rapid growth, destruction of surrounding bone and soft tissues, and invasive behavior, with the potential for early hematogenous and lymphogenous metastasis.This tumor typically produces osteolytic lesions, causes facial skeletal deformity, and loosens teeth. Histologically, it may present as osteosarcoma, chondrosarcoma, fibrosarcoma, or rhabdomyosarcoma. Treatment usually involves radical surgical resection, often combined with chemotherapy or radiotherapy, and requires long-term follow-up.

## Main Part

Sarcomas are malignant neoplasms derived from cells of mesenchymal origin. The originating tissue is diverse and includes bone, cartilage, muscular, fibrous, vascular, fatty, and neural tissue. Sarcomas of the head and neck are rare tumors, accounting for 4–10% of all sarcomas [1,2, 3, 4, 5,] and fewer than 1% of neoplasms in this region [3, 6]. Sarcomas of the oral and maxillofacial region are

even rarer. Gorsky and Epstein [6] reviewed 11,250 head and neck malignancies, among which there were 139 cases of sarcomas (1.24%), and among these there were only 16 cases (0.14%) of intraoral soft tissue sarcoma [6]. There are some previous reports on sarcomas of the head and neck, but only few reports focus on sarcoma of the oral and maxillofacial region [7,8, 9].

**Classification:** Broadly speaking, sarcomas can be classified into two categories: soft tissue and hard tissue (bone and cartilage) sarcomas. This, however, is an oversimplification since precise classification is fraught with significant difficulties. For example, fibrosarcoma and malignant fibrous histiocytoma are soft tissue sarcomas that arise in hard tissues [10, 11]. In contrast, chondrosarcomas are classified as soft tissue tumours. Additionally, some hard tissue tumours display extraosseous extension [12,13,14]. Ten or more main histological types and even more subtypes of sarcomas are described. Osteosarcoma, angiosarcoma and malignant fibrous histiocytoma make up the majority tumours in adults. In children, rhabdomyosarcoma is the most common sarcoma [12,13,14].

**Pathology & Oncology Research.** Sarcomas of the Oral and Maxillofacial Region (SOMR) are rare lesions which pose diagnostic and management challenges. We analyzed 26 cases of SOMR with respect to clinical presentation, histopathological subtype, treatment modalities, recurrence, and treatment outcome. In our series, Osteosarcoma (OS) was the most common type of sarcoma (7 cases), followed by 5 cases of Ewing's Sarcoma (ES), 3 cases each of Chondrosarcoma (CS) and Leiomyosarcoma (LMS), 2 cases each of Malignant Peripheral Nerve Sheath Tumor (MPNST), Pleomorphic Undifferentiated Sarcoma (PUS), Myeloid Sarcoma (MS)and Rhabdomyosarcoma (RMS). Surgery was the primary treatment modality in most cases and was combined with adjuvant chemo/ radiotherapy in few cases. 24 of the 26 cases were followed up for an average period of 40.67 months. Adverse disease outcomes like recurrence were seen in 2 cases whereas death due to the disease was reported in 7 cases. In view of the diagnostic challenges faced in SOMRs, it appears practical to stress on the underlying genetic aspects of the disease process rather than histological subtyping to improve disease outcome. [15.]

**Histopathological Challenges in Sarcoma Diagnosis**

Histopathology is the main stay in sarcoma diagnosis but is frequently problematic with an abundance of di- agnostic pitfalls. Oral lesions that may be misdiagnosed as sarcomas are benign bony lesions like fibrous dysplasia and ossifying fibroma for a low-grade osteosarcoma, lymphomas for round cell

tumors and fibromatosis for low-grade fibrosarcomas. Epithelial malignancies like sarcomatoid carcinoma may be misinterpreted as an epitheloid sarcoma in the absence of surface epithelium in a small biopsy sample. Thus conventional histology needs to be supplemented with ancillary methods like immunohisto-chemistry and molecular diagnostic techniques for accurate diagnosis

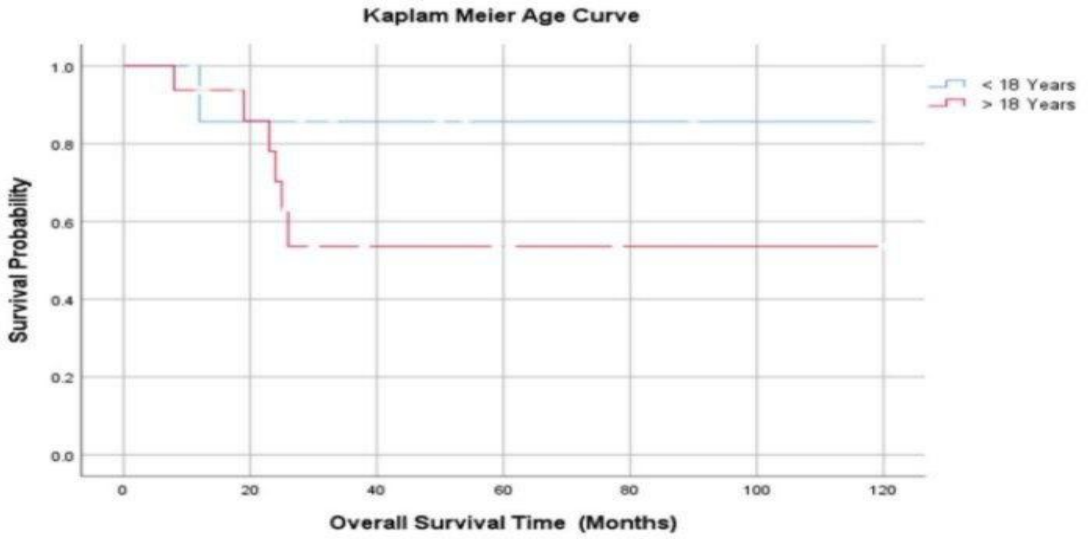


**Fig. 2** Kaplan-Meier Age curve

**Osteosarcoma**

Osteosarcomas are primary malignant bone tumours in which mesenchymal cells produce osteoid [16-17]. Although osteosarcoma is the most common malignant bone tumour, lesions of the jaw are uncommon, representing about 4% of the osteosarcomas [18]. Osteosarcoma of the jaw (JOS) occurs over a wide age range, with a peak in the fourth decade of life [18-21]. Although a male predilection has been reported [18,19,20], some authors have observed a slight predominance in females [20]. The chief clinical features are swelling, pain, and ulceration [18-21.] Radiologically, the findings may include radiolucency, radiopacity, or a mixture of both with poorly defined irregular margins [18-21]. JOS differs from osteosarcoma of the long bones in its biological behaviour, presenting a lower incidence of metastasis and a better prognosis [18,22]. Early diagnosis and adequate surgical resection are the keys to high survival rates [20].

**Case Report**

A 38-year-old woman was referred to the Department of Oral and Maxillofacial Surgery, Baleia Hospital, for evaluation and treatment of fibrous dysplasia of the facial bones. The patient was complaining about pain, the loss of visual acuity in the right eye and the limitation of the oral opening.

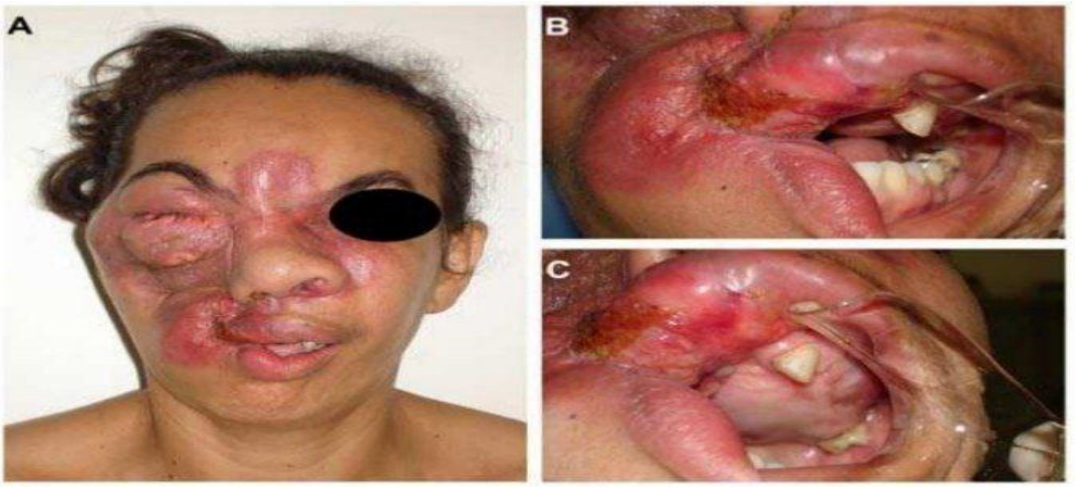


**Fig. 1.** Clinical appearance of the lesion. (A) Redness and swelling of the skin in right perioral, right orbital, glabelar, nasal and left paranasal regions. (B and C) Upper lip ulceration and hard palate swelling covered by normal mucosa.

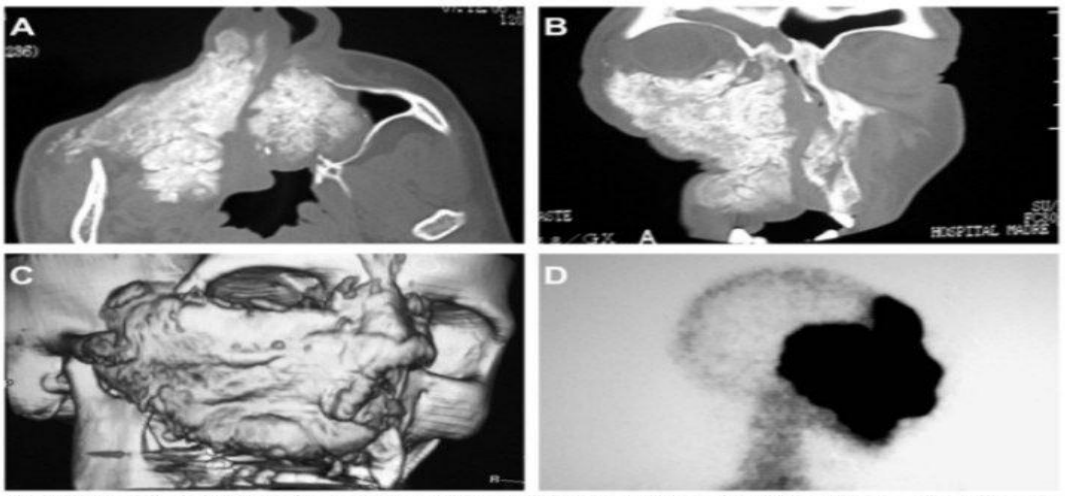


**Fig. 2.** Computed tomography and bone scintigraphy. (A and B) Computed tomography demonstrating large hyperdense mass in right maxilla extending to right orbit and left maxillary sinus. (C) Three-dimensional computed tomography reconstruction showing an expansive lesion in the right hemiface. (D) Bone scintigraphy showing a hypercaptation area in the face.

According to the anamnesis, the patient had been diagnosed with fibrous dysplasia four years earlier, when she developed a hard palate painless swelling. During these four years,the patient was submitted to three osteoplasties and one partial maxillectomy, all followed by recurrences. The specimens removed in these surgical procedures had been histopathologically diagnosed as fibrous dysplasia. The patient informed that the last recurrence had begun three months prior, presenting rapid growth and skin damage. Extraoral examination showed redness and swelling of the skin in the right perioral, right orbital, glabelar, nasal and left paranasal regions (Fig. 1A). Intraoral examination showed painful upper lip ulceration as well as hard palate swelling covered by normal mucosa (Fig. 1B and C). Computed tomography showed a large hyperdense mass in right maxilla

extending to right orbit and left maxillary sinus (Fig. 2A and B). Three-dimensional computed tomography reconstruction showed an expansive lesion in the right hemiface (Fig. 2C). Bone scintigraphy showed hypercaptation in the entire face (Fig. 2D). An incisional biopsy was performed and the specimen was sent to the Oral Pathology Laboratory, School of Dentistry, Pontifícia Universidade Católica de Minas Gerais. Microscopic examination demonstrated areas of osteoid and chondroid formation surrounded by a cellular stroma (Fig. 3). The diagnosis of osteosarcoma was established, and the patient was recommended for oncologic treatment. Since surgical resection was not feasible, the oncology service decided to use palliative chemotherapy. The patient died six months after the osteosarcoma diagnosis due to uncontrollable local spread.

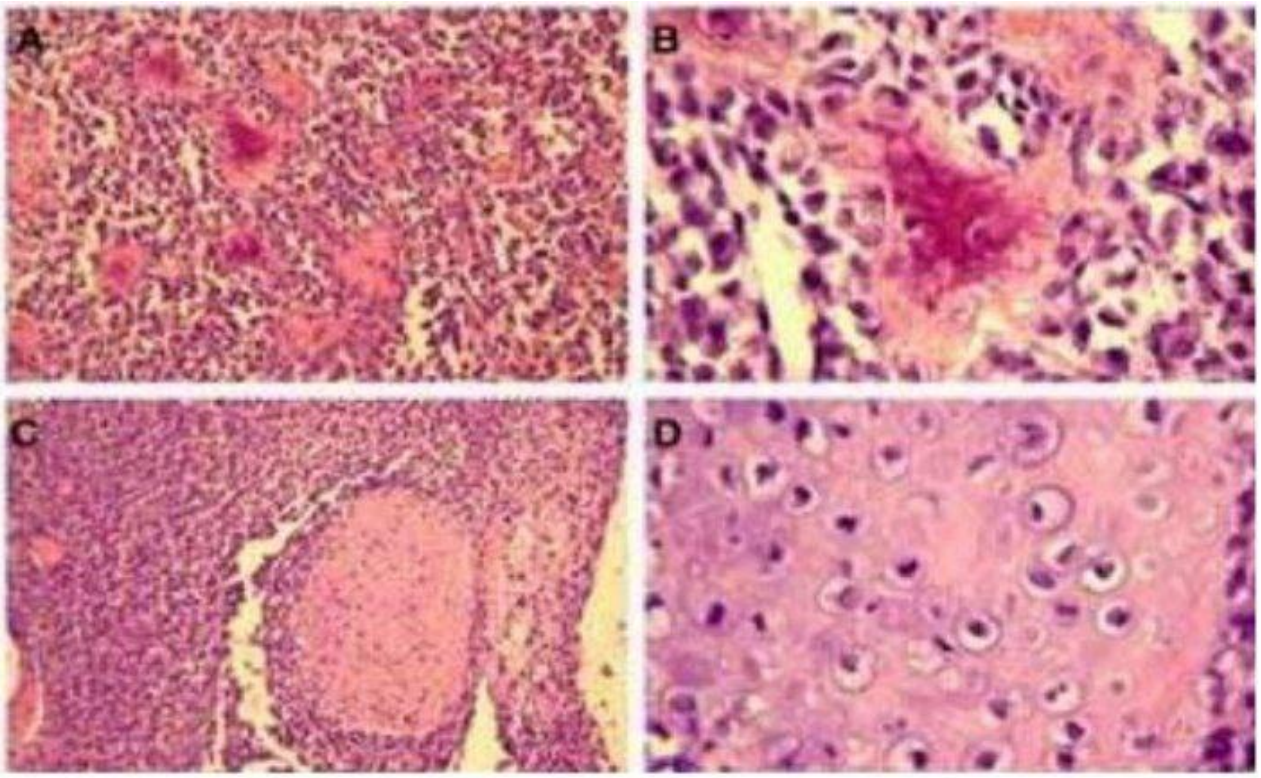


**Fig. 3.** Light microscopic observation of the tumour. Areas of osteoid formation surrounded by a cellular stroma (A and B; HE x100 in "A" and HE x400 in "B"). Area of chondroid formation surrounded by a cellular stroma (C; HE x 100). High magnification of chondroid formation with cellular atypia (D; HE x 400).

**Discussion**

Osteosarcomas are malignant bone tumors which most commonly arise from long bones of the extremity. Osteosarcoma of head and neck region accounts for less than 10% of all cases of osteosarcoma.[23]Osteosarcoma have different

histological variant of which Giant cell-rich osteosarcoma is a rare histological variant accounts for about 3% of all Primary intraosseous osteosarcomas, first described by Bathurst et al in head and neck region mandible is the common site, giant cell rich osteosarcoma of maxilla very uncommon as only two case being published in the literature regarding GCRO of maxilla till now, GCRO is aggressive, poor prognosis commonly seen in older age group and female predominance but in our case it seen in young age male its seen.[24] The differential diagnosis of giant cell lesions can present a diagnostic challenge, especially if they occur in an unusual location. In this case, it was difficult to differentiate between a giant cell granuloma from giant cell osteosarcoma. The major risk factors for development of head and neck osteosarcoma are radiation exposure and Paget's disease.[25]

Radiologically giant cell rich osteosarcoma are different from the conventional osteosarcomas as they mimic non mineralized benign or malignant bone tumors. In the long bones they present as osteolytic lesion with cortical thinning and ballooning without obvious cortical destruction.[26] While in our case CECT Nose PNS showed expansile heterogeneous mass filling the maxilla with extensive destruction of the all the walls of the maxilla, areas of speckled calcification within the tumor and adjacent sclerosis. The tumor was destroying the palate and extending into oral cavity. Radiologically, the differential diagnosis includes giant cell granuloma, aneurysmal bone cyst, chondrosarcoma, Chondromyxoid fibroma and malignant fibrous histiocytoma.[27] Surgical excision with post operative Radiotherapy is the treatment of choice. In our case combined approach radical tumour excision done ie total maxillectomy and reconstruction.Temporalis muscle flap is used for reconstruction in this case as it has a reliable and hearty blood supply, can be tailored to provide reconstruction of most oropharyngeal defects, and arguably presents minimal donor site morbidity if performed correctly.[28]The flap is accessible through a hemicoronal incision that is hidden in the hair-bearing scalp and preauricular crease.The temporalis muscle flap lies in the same operative field as the defect, potentially reducing operative time and reducing motion-induced viability issues during the recovery period. The deep temporal fascia lines the flap and obviates the need for skin grafting. The temporalis muscle develops a mucosal layer with time.[29]

**Conclusion**

Sarcoma maxillae superioris is one of the most aggressive malignant tumors of the maxillary region, characterized by its mesenchymal origin, rapid growth, invasive behavior, and ability to metastasize early. The disease is often diagnosed

at a late stage due to nonspecific clinical manifestations, which reduces treatment effectiveness and worsens prognosis.

Accurate diagnosis relies on radiological imaging and histopathological biopsy, both of which play a decisive role. Effective treatment requires a multimodal approach combining radical surgical resection with chemotherapy and radiotherapy. Therefore, early detection, appropriate differential diagnosis, and comprehensive treatment strategies are crucial for improving patients' quality of life and prolonging survival.

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