

# CASE REPORT

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## MYOFIBROBLASTIC SARCOMA: A CASE REPORT

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**ABSTRACT:** Myofibroblastic Sarcoma is a rare or uncommon neoplasm characterized by myofibroblastic proliferation. The tumour may present as single or multiple nodules of soft tissue, bone, or internal organs. When encountered in the jaws, the lesions exhibit clinical and radiographic features suggestive of several odontogenic and non-odontogenic neoplasms. Myofibroblastic Sarcoma of the jaws are usually well-demarcated, but sometimes can be poorly delineated or infiltrated that may result in misdiagnosis and mistreatment<sup>1</sup>. A painless, enlarging mass is the most common clinical presentation, but a definitive diagnosis requires both histopathological and immunohistochemical analyses. In this paper we are going to discuss a case of 64 years old male with Myofibroblastic Sarcoma of left side of face. The patient underwent excision of the lesion in toto with reconstruction of the floor of the orbit using prolene mesh.

**KEYWORDS:** Myofibroblastic Sarcoma, Myofibroma, Maxilla, Alveolus, Floor of Orbit, Prolene mesh.

**INTRODUCTION:** Defining neoplastic Myofibroblastic Sarcoma (MS) as a distinct entity was controversial. With increasing case reports, it became clear that Myofibroma was a distinct entity in soft tissue sarcomas. Even though only low-grade Myofibroma was classified as a distinct entity in the newly published World Health Organization classification of soft tissue tumours, intermediate- and high-grade Myofibroma cases were documented in the literature. Some cases of MYOFIBROBLASTIC SARCOMA were easy to misdiagnose as reactive lesions because of the common existence of myofibroblasts in reactive granuloma. In addition, the frequent exhibition of bland cytologic features in MYOFIBROBLASTIC SARCOMA is an important factor in persuading pathologists toward a benign diagnosis. MYOFIBROBLASTIC SARCOMA, especially low-grade MYOFIBROBLASTIC SARCOMA, is easily confused with myofibroblasts composing nodular fasciitis because of their morphologic similarity and the overlapping immunophenotype.

**MYOFIBROBLASTIC SARCOMA** is a common mesenchymal neoplasm that occurs most frequently in the uterus and gastrointestinal tract. The occurrence of primary MYOFIBROBLASTIC SARCOMA in the oral soft tissues or jawbones is very unusual. 1) Only 31 cases have been reported in the literature. 2) Among cases of oral soft tissue MYOFIBROBLASTIC

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SARCOMA, six arose in the cheek, five in the gingiva, five in the tongue and the remainder were located in the floor of the mouth, soft palate, hard palate, mandibular alveolar mucosa, posterior maxillary soft tissue and maxillary sinus. Among the jawbone lesion cases, six occurred in the mandible and four in the maxilla<sup>2</sup>.

This paper discusses an unusual case of primary MYOFIBROBLASTIC SARCOMA of the oral cavity, which arose in the maxillary buttress and had the growth pattern of a polyp.

**DESCRIPTION OF CASE:** A 64-year-old Male patient visited the Department of Oral & Maxillo-facial Surgery at A. J. Institute of Dental Sciences, Mangalore with the chief complaint of a swelling over the left side of the face region since 2 months duration. Swelling was insidious in onset, initially small in size and associated with pain which was mild in intensity and continuous type. Extent

Anteriorly: Left lateral aspect of the nose.

Posteriorly: 3 cm in front of the tragus.

Superiorly: Infraorbital rim

Inferiorly: Line joining corner of the mouth to tragus of the ear on the left side.

Size: 5 cm x 4 cm.

Patient had previously visited a dentist with complaints of pain and swelling and underwent extraction of the two upper left molars as they were mobile. After extraction swelling rapidly increased in size to its present size. Pain was moderate type and not subsiding with medications. Patient is hypertensive and is on medication. On examination bucco-palatal expansion was noted in the upper left molar region and 1<sup>st</sup> premolar region. Physical examination revealed a firm swollen region, which showed tenderness, pain at palpation. Surface was smooth, irregular in shape with ill defined margin with normal overlying skin. Gingival swellings extended from the left maxillary canine to the 3<sup>rd</sup> molar. However, the patient had not experienced any symptoms. On radiographic examination an extensive radiolucent lesion with sclerotic borders was revealed above the left maxillary canine and molar region. Root resorption of involved teeth was observed with grade III mobility of the 2<sup>nd</sup> and 3<sup>rd</sup> molar teeth for which patient was treated with extraction and Incision & Drainage was done for the swelling as the swelling was believed to be a periodontal abscess. On cross-sectional view of occlusal projection expansion of buccal and palatal cortical bone of the maxilla with partial resorption of buccal cortical bone was observed. Skin over the swelling was normal in appearance but stretched. Obliteration of the nasolabial fold was seen. Watery discharge was seen from the left eye.

Provisional diagnosis was Peripheral Ossifying Fibroma, Fibrosarcoma, Osteosarcoma. Incisional biopsy was performed under local anaesthesia with adrenaline (1:200000) and the sample was sent to Department of oral pathology for histopathological examination. Histopathological examination suggested the lesion to be Osteosarcoma (Chondroblastic Variant).

Patient was admitted in our hospital and all the necessary blood investigations were carried out along with ECG, ECHO, Chest X ray and C. T. Scan of the Head. After obtaining a medical clearance, the patient was taken up for Tumour excision under general anaesthesia. Reconstruction of the orbital floor was done using prolene mesh which was suture using 3.0 vicryl. Intra orally suturing was done using 3.0 vicryl and skin suturing using 5.0 prolene. The excised tumour was then sent for histopathological examination.

Furthermore, patient was advised for therapeutic radiotherapy.

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**HISTO PATHOLOGICAL ASSESSMENT:** Sections show a cellular spindle cell tumour arranged in intersecting fascicles and slight swirling striiform and whorl-like pattern. The tumour cells are spindly to lump to stellate shaped showing mild pleomorphism, with vesicular nucleus some showing conspicuous nucleolus and ill-defined pale eosinophilic cytoplasm. Focal nuclear hyperchromasia is also seen. The stroma is myxoid at places and contains many thin walled dilated and congested blood vessels. Large areas of hemorrhage are also seen in the studied sections. Mitosis is low. A sparse chronic inflammatory infiltrate is also seen. Tumor tissue is seen infiltrating the surgical resected margin.

Histological features are suggestive of low grade malignant SPINDLE CELL SARCOMA. Immunohistochemistry for further evaluation and confirmation of histopathological changes (CD34, S- 100 CD 99, Desmin, Actin).

### **IMMUNOPHENOTYPING REPORT:**

SMA : Positive in some of the tumour cells.

Desmin : Negative in tumour cells.

S-100 : Negative in tumour cells.

CD99 : Negative in tumour cells.

CD34 : Negative in tumour cells.

**IMPRESSION:** With above features, Myofibroblastic Sarcoma has to be considered.

**DISCUSSION:** Myofibroblasts are modified fibroblasts, which can occur in normal tissues (e.g. periodontal ligaments), reparative granulation tissue and reactive soft-tissue lesions. The cause of Myofibroma is presently unknown. A number of authors have suggested that the tumors are inherited in an autosomal dominant or alternatively in an autosomal recessive trait. However, its low familial incidence suggests that there are probably factors other than genetics that play an important role in the cause of this disease<sup>7</sup>. They are, however, also a principal cell type in benign and malignant soft-tissue tumours. Myofibroblasts are spindle-shaped or stellate cells with ovoid pale nucleus and distinct nucleolus. The cytoplasm is usually amphophilic and there are indistinct cell borders. Ultrastructurally, myofibroblasts can be distinguished from fibroblasts and smooth muscle cells by the findings of indentation of nucleus, presence of peripheral or subplasmalemmal bundles of thin cytoplasmic filaments, termed stress fibres, and a distinctive cell-stromal attachment termed fibronexus<sup>4</sup>. Myofibroblastic Sarcoma is a well defined malignant tumour composed of myofibroblasts. It can occur at any age with slight male predominance; the tumour size varies from 1.5 cm to 17 cm. MYOFIBROBLASTIC SARCOMA can arise in soft tissues of various anatomic sites, including extremities, trunk (e.g. retroperitoneum, breast and heart) and genital tract, but there is a predilection for the head and neck region. Oral cavity (especially tongue, cheek and gingiva), nasal cavity, salivary glands and bones (e.g. maxilla and mandible) can be affected. Grossly, most cases were described as firm, gray-white coloured tumours with ill-defined margins. Histologically, the tumour cells show diffuse fascicular or storiform growth pattern and they infiltrate surrounding tissues (e.g. skeletal muscle). The cytological and ultrastructural characteristics of neoplastic cells are consistent with myofibroblasts (described above). Increased mitotic activity of tumour cells and tumour necrosis is associated with more aggressive behaviour of MYOFIBROBLASTIC SARCOMA<sup>5</sup>. Tumour cells in MYOFIBROBLASTIC SARCOMA show a variable immunophenotype: actin positive/desmin negative, actin negative/desmin positive, and actin positive/desmin positive

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cases. In addition, neoplastic cells can express fibronectin, calponin, CD 34, CD 99 and CD 117, whereas S-100 protein, epithelial markers and h-caldesmon are negative. MYOFIBROBLASTIC SARCOMA is usually a tumour of low grade malignancy, prone to local recurrence (about 30% in general), even after many years<sup>5</sup>. However, metastases to lungs have been also reported. Therefore, close clinical follow-up of the patient is mandatory. Differential diagnosis of MYOFIBROBLASTIC SARCOMA includes reactive myofibroblastic lesions, benign myofibroblastic lesions and other Myofibroblastic Sarcomas. In these cases, immunohistochemistry is not helpful and the differential diagnosis is based on careful examination of the slides stained with hematoxylin-eosin. On the other hand, many histogenetically different spindle-cell tumours can be distinguished from MYOFIBROBLASTIC SARCOMA using immunohistochemistry. Nodular fasciitis is a rapidly growing lesion, histologically with variable cellularity and myxoid stroma. It is less cellular and uniform than MYOFIBROBLASTIC SARCOMA. Although mitoses can be present, the cells lack nuclear atypia, which is usually present in MYOFIBROBLASTIC SARCOMA. Fibromatoses are composed of uniform, collagen-producing myofibroblasts without nuclear pleomorphism. Inflammatory myofibroblastic tumour (IMT) is histologically characterized by fasciitis-like, fascicular and sclerosing areas with a prominent chronic inflammatory infiltrate with numerous plasma cells. In addition, anaplastic lymphoma kinase (ALK) can be immunohistochemically detected in 30–40% of Inflammatory myofibroblastic tumours<sup>6</sup>. The inflammatory infiltration in the presented case of MYOFIBROBLASTIC SARCOMA was, however, due to tumour ulceration. High grade Myofibroblastic Sarcoma is often difficult to distinguish from other high grade sarcomas. This diagnosis should be established in those cases, where the presence of myofibroblasts is confirmed by electronmicroscopy. Histogenetically different spindle-cell tumours can be distinguished from MYOFIBROBLASTIC SARCOMA using immuno-histochemistry. Spindle-cell carcinoma shows positivity for cytokeratins, fibrosarcoma is SMA negative, leiomyosarcoma has a different microscopic appearance and shows desmin and h-caldesmon positivity. Spindle-cell rhabdomyosarcoma shows SCMA positivity, angiosarcoma shows positivity for endothelial markers (e.g. F VIII, CD 31 and CD 34) and malignant peripheral nerve sheath tumour shows at least focal positivity for S-100 protein. In summary, MYOFIBROBLASTIC SARCOMA is a well-defined myofibroblastic malignancy with predilection for head and neck region, which is prone to local recurrency, rather than metastasing. For the diagnosis, a careful examination of routinely stained slides is crucial. Immunohistochemistry can be useful in differential diagnosis from some other spindle cell lesions and tumours. Electronmicroscopic examination can support the diagnosis of MYOFIBROBLASTIC SARCOMA by proving the origin of tumour cells.

### LEGENDS:

1. Frontal pre-operative photograph.
2. Birds eye view pre-operative photograph.
3. Intra oral lesion pre-operative photograph.
4. Intra operative photograph after excision of the lesion.
5. Excised lesion in toto.
6. Frontal view Post operative photograph.
7. Birds eye view Post operative photograph.
8. Histopathology Photograph.
9. C. T. Scan Pre operative sub mento vertex view.
10. Occlusal Radiograph

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Fig 1: Frontal pre-operative photograph



Fig 2: Birds eye view pre-operative photograph



Fig 3: Intra oral lesion pre-operative photograph



Fig4: Intra operative photograph after excision of the lesion



Fig 5: Excised lesion in toto.



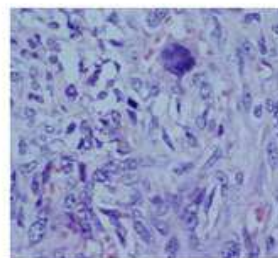
Fig 6: Frontal view Post operative photograph



Fig 7: Birds eye view Post operative photograph



Fig 8: Histo pathology Photograph



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Fig 9: C.T.Scan Pre operative sub mento vertex view

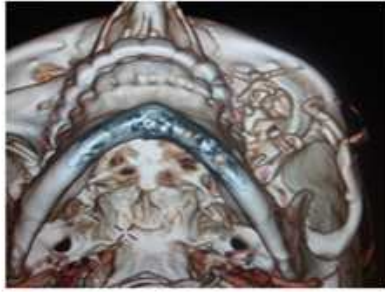


Fig 10: Occlusal Radiograph



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