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RESEARCH ARTICLE

HIGH-GRADE MYOFIBROBLASTIC SARCOMA OF THE BUCCAL MUCOSA: A RARE ORAL MALIGNANCY WITH RAPID FATAL PROGRESSION

Ansueya Sharma[1], NDVN Shyam[2], Kiran Kumar Gundamaraju[3], Vaishali Narayen[3], Paremala K[3], Radhika Ramgundamwar[4], Guruvarshni V[1] and T. Maria Alphonsa Deepthi[1]

1. Postgraduate, Department of Oral and Maxillofacial Pathology, Government Dental College & Hospital, Hyderabad, Telangana, India.
2. HOD & Professor, Department of Oral and Maxillofacial Pathology, Government Dental College & Hospital, Hyderabad, Telangana, India.
3. Associate Professor, Department of Oral and Maxillofacial Pathology, Government Dental College & Hospital, Hyderabad, Telangana, India.
4. Senior Resident, Department of Oral and Maxillofacial Pathology, Government Dental College & Hospital, Hyderabad, Telangana, India.

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Abstract

Myofibroblastic Sarcoma is a rare malignant neoplasm of soft tissue origin defined by the proliferation of spindle-shaped mesenchymal cells with myofibroblastic differentiation. The present report describes the case of a 55-year-old male with a four-month history of progressive swelling in the left posterior mandibular region. Clinical assessment demonstrated a mass arising from the left buccal mucosa,while histopathological examination revealed a densely cellular spindle-cell neoplasm arranged in elongated fascicular patterns within a collagenized and hyalinized stroma. Immunohistochemical evaluation substantiated myofibroblastic differentiation and when interpreted in conjunction with the histopathological findings, supported the diagnosis of Myofibroblastic Sarcoma.The patient ultimately succumbed to the disease within 4 months, underscoring the potentially fulminant biological behaviour of this rare oral mesenchymal malignancy.

Introduction:

Myofibroblastic Sarcoma (MS) is a rare malignant soft-tissue neoplasm characterized by the proliferation of mesenchymal spindle cells with myofibroblastic differentiation. In 1998, Mentzel et al. first described this tumour entity following the clinicopathological evaluation of 18 myofibroblastictumours. [1] Current knowledge of MS remains limited because most published data are derived from isolated case reports. [2–6] In 2002, the World Health Organization (WHO) recognized Low-Grade Myofibroblastic Sarcoma as a distinct entity in its classification of soft tissue tumours, and it continues to be recognized in the current WHO classification. However, Intermediate- and High-Grade forms remain poorly characterized owing to their rarity and heterogeneous clinicopathological features, resulting in the absence of universally accepted grading criteria. [3,5,6] Consequently, Intermediate- and High-

Corresponding Author: Paremala K
Address: Associate Professor, Department of Oral and Maxillofacial Pathology, Government Dental College & Hospital, Hyderabad, Telangana, India.

Grade Myofibroblastic Sarcomas are graded using the FNCLCC system, which evaluates tumour differentiation, mitotic activity, and necrosis.[7] Immunohistochemistry complements grading by confirming myofibroblastic differentiation, excluding spindle-cell mimics, and assessing tumour aggressiveness, thereby improving diagnostic accuracy and prognostic stratification.

Case presentation:

A 55-year-old male presented to the Government Dental College and Hospital, Hyderabad, with a chief complaint of swelling in the lower left posterior tooth region for 4 months. The lesion began as a small, inconspicuous mass and progressively expanded to its current dimensions. The swelling was accompanied by mild pain. Extraoral evaluation revealed a subtle yet diffuse fullness involving the left middle and lower third of the face, producing mild facial asymmetry and noticeable restricted mouth opening. The overlying skin exhibited a normal hue without surface alterations. On palpation, the swelling was firm and non-fluctuant, suggesting a solid underlying pathology. On intraoral examination, a well-defined, nodular exophytic growth measuring approximately 3.0 × 4.0 cm^2 was identified on the left buccal mucosa, extending from the retrocomissure to the retromolar region. The lesion was firm and mildly tender on palpation, with no evidence of surface ulceration or bleeding. (Figure 1)

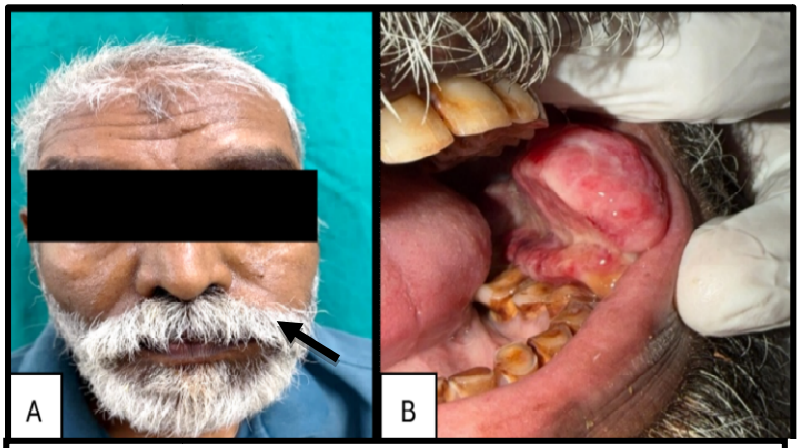

FIGURE 1: A) Clinical image showing a mild diffuse swelling noticed on left middle and lower third of the face. B) Well-defined, nodular exophytic growth of 3.0 x 4.0 cm^2 on left buccal

Radiographic examination of left posterior mandibular region revealed no underlying bony destruction. (Figure 2)

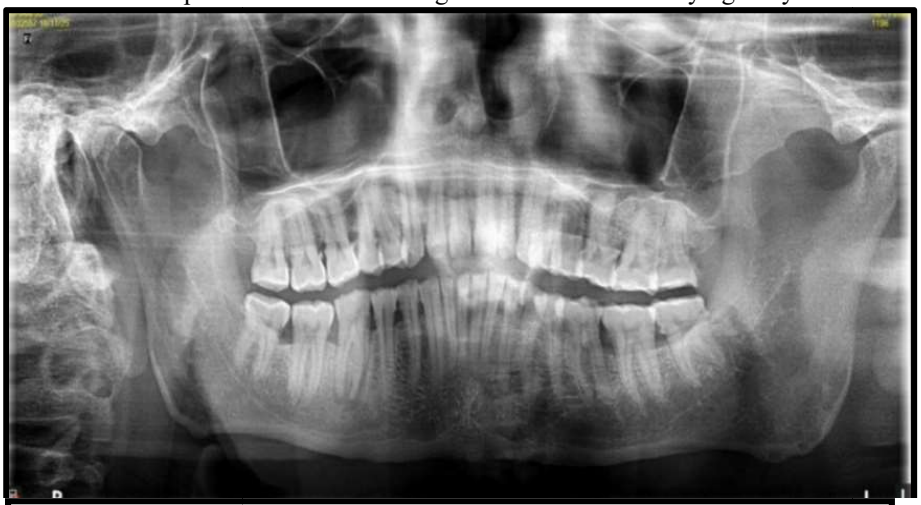

FIGURE 2: Orthopantomogram (OPG) of left posterior mandibular region with no obvious destruction of the bone.

An incisional biopsy of the lesion was subsequently performed, and the tissue specimen was submitted to the Department of Oral and Maxillofacial Pathology, Government Dental College and Hospital, Hyderabad, for histopathological evaluation. On gross examination, two cream-brown soft-tissue bits measuring about 1.1 × 0.8 cm^2 were received, which were firm in consistency. (Figure 3)

FIGURE 3:Two cream-brown soft-tissue bits measuring a bout 1.1 × 0.8 cm^2 were received, which were firm in consis

Microscopically, the lesion showed highly cellular stroma composed of spindle-shaped cells arranged in long fascicles within a collagenous and hyalinized background. These fascicles are admixed with pleomorphic cells showing vesicular nuclei and pale cytoplasm. The lesional cells showed marked atypia in the form of cellular and nuclear pleomorphism, nuclear hyperchromatism with prominent nucleoli and increased and abnormal mitosis. Less than 50% tumour necrosis and up to 6 mitotic figures per high-power field (HPF) were observed.Mild chronic inflammatory infiltrate and vasculature were also evident. (Figure 4)

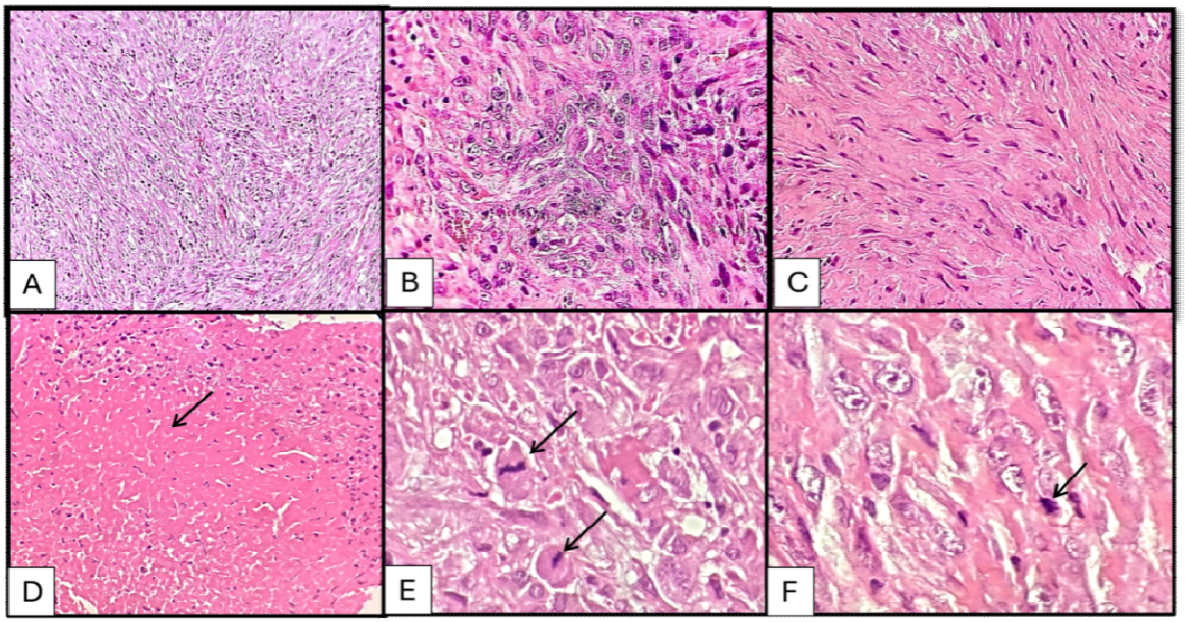

FIGURE 4: A) Spindle shaped cells arranged in long fascicles within a collagenous and hyalinized background admixed with pleomorphic cells. (H&E 20X) B) Pleomorphic cells showing vesicular nuclei and pale eosinophilic cytoplasm with marked atypia and mitotic figures. (H&E 40X) C) Streaming fascicles of spindle shaped cells with marked cellular and nuclear pleomorphism. (H&E 40X) D) High-magnification photomicrograph showing extensive acellular eosinophilic, glassy areas consistent with tumour necrosis. E) & F) High magnification photomicrograph showing mitotic figures.

Immunohistochemical analysis was performed to determine the lineage of the spindle cells. The immunohistochemical panel included vimentin, alpha-smooth muscle actin (α-SMA), p53, Ki-67, desmin, CD68and S-100. Immunoreactivity was assessed using standardized scoring criteria, and quantitative analysis of positively stained cells was performed using ImageJ software. The tumour cells demonstrated strong positivity for vimentin, α-SMA, p53and Ki-67, while they were negative for desmin, CD68and S-100. (Figure 5)

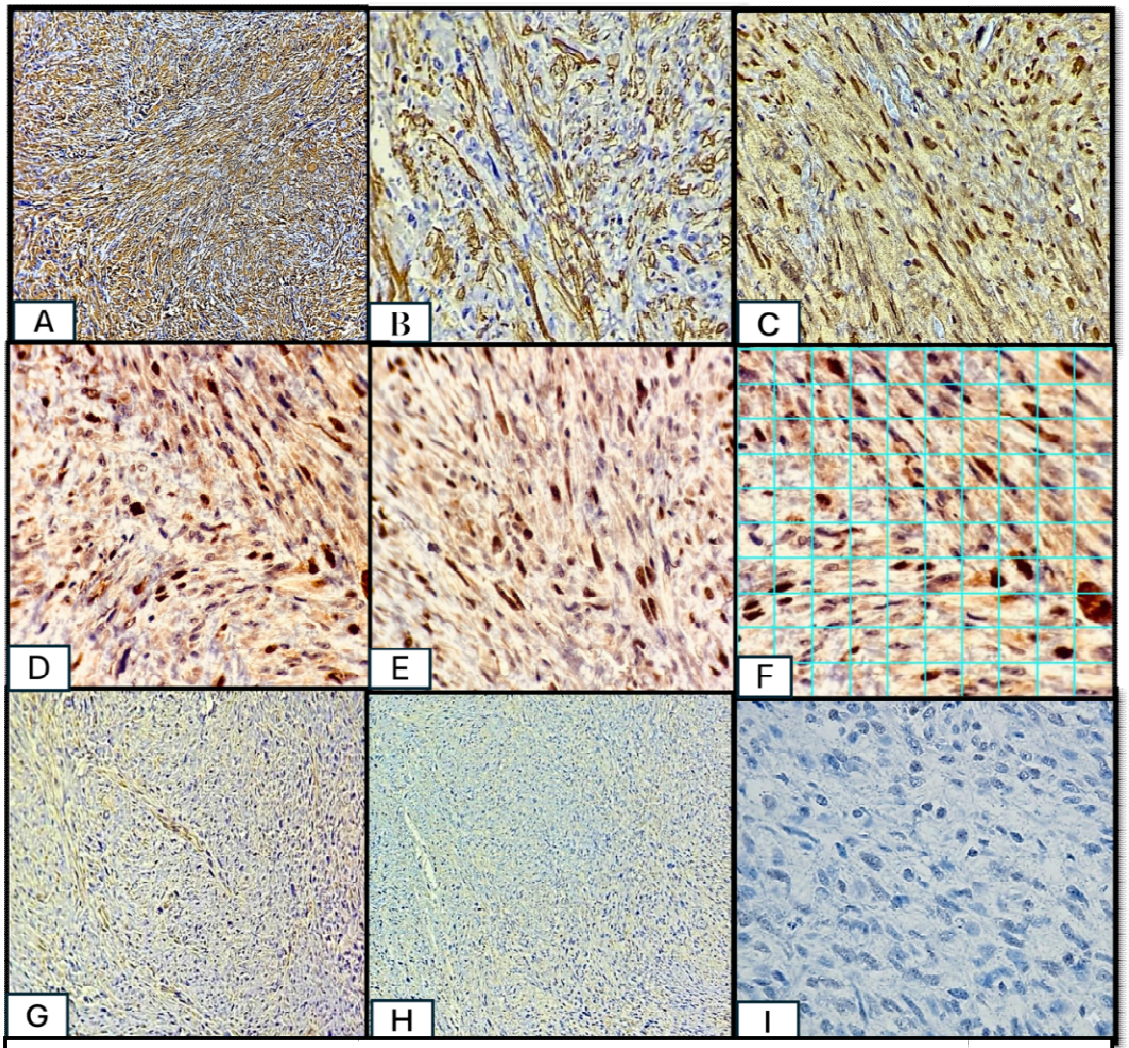

FIGURE 5: A) Immunohistochemistry revealed strong positivity for Vimentin. (20X) B) Immunohistochemistry revealed positivity for α-SMA. (40X) C) Immunohistochemistry revealed positivity for P53. (40X) D) & E) Immunohistochemistry revealed strong positivity for Ki 67. F) ImageJ-assisted quantification of immunopositive cells using a standardized cell-counting method. G) Immunohistochemistry revealed negativity for Desmin. (20X) H) Immunohistochemistry revealed negativity for CD68. (10X) I) Immunohistochemistry revealed negativity for S100. (10X)

Based on the cumulative histopathological, immunohistochemical, and clinicopathological findings, a final diagnosis of High-grade Myofibroblastic Sarcoma was established. Histologically, the tumour was assigned an FNCLCC Grade 2 based on tumour differentiation, mitotic activity, and extent of tumour necrosis. Although the FNCLCC grading system classified the lesion as Grade 2, the marked cytological atypia, atypical mitotic activity, tumour necrosis, diffuse p53 overexpression, high Ki-67 labeling index, and rapidly progressive clinical course indicated a biologically aggressive phenotype, supporting the diagnosis of High-grade Myofibroblastic Sarcoma. Owing to financial limitations, the patient was referred to MNJ Cancer Hospital, Hyderabad, where a multidisciplinary tumour board recommended single-agent doxorubicin (60 mg/m² every 21 days). Despite initiating chemotherapy, the patient deteriorated rapidly and died after the first treatment cycle, approximately four months

after the initial presentation. This fulminant course was consistent with the tumour's adverse clinicopathological features, including marked cellular atypia, a high Ki-67 proliferative index, and diffuse p53 overexpression.

Discussion:-

Myofibroblastic Sarcoma (MS) is an exceptionally rare malignant mesenchymal neoplasm arising from myofibroblasts, accounting for approximately 0.14% of head and neck malignancies. [1,8] Although it most frequently involves the soft tissues of the head and neck, cases have also been reported in the extremities, chest wall, axillary and inguinal regions, abdominal cavity, and bone. [3,5,9,10] The tumour has been described across a wide age range (6–75 years), with a slight male predominance and a mean age of approximately 40 years. [5,9] Clinically, MS usually presents as a slowly enlarging, painless soft-tissue mass, making early diagnosis challenging because of its nonspecific presentation. [3,11] Histologically, MS is characterized by spindle to stellate-shaped cells arranged in intersecting fascicles, sheets or storiform patterns within a collagenous or myxoid stroma. The neoplastic cells typically exhibit indistinct eosinophilic cytoplasm and fusiform nuclei with variable cytological atypia. [7] In the present case, the tumour demonstrated long fascicles of spindle cells with marked cellular and nuclear pleomorphism, atypical mitotic figures and focal tumour necrosis, features indicative of an aggressive spindle-cell sarcoma.

Because no universally accepted grading criteria exist for Intermediate and High-Grade Myofibroblastic Sarcoma, histological assessment is generally performed using established soft-tissue sarcoma grading systems, particularly the French Federation of Cancer Centers Sarcoma Group (FNCLCC) system. [3,12] The FNCLCC system incorporates tumour differentiation, mitotic count, and tumour necrosis to stratify tumours into three prognostic grades. In the present case, the lesion achieved an FNCLCC score of 4 (Grade 2). However, histological grading alone may not fully capture the biological behaviour of rare spindle-cell sarcomas.

To complement conventional histopathological grading, Ki-67 immunohistochemistry was performed to assess tumour proliferative activity. ImageJ-based quantitative analysis, following the method of Li YX et al.,[13] demonstrated a Ki-67 labelling index of 72%, indicating markedly increased proliferative activity. Although the tumour was classified as FNCLCC Grade 2, the Ki-67 index (≥30%, highest proliferative score according to Hasegawa et al.,[14]), together with diffuse p53 overexpression, marked cytological atypia, tumour necrosis, and rapid clinical progression, indicated biologically aggressive behaviour. Therefore, the diagnosis of high-grade myofibroblastic sarcoma was based on an integrated histopathological, immunohistochemical, and clinicopathological assessment rather than the FNCLCC grade alone.

Ultrastructurally, Myofibroblastic Sarcoma (MS) is characterized by clefted nuclei, abundant rough endoplasmic reticulum, microfilaments, subplasmalemmal plaques, discontinuous basal lamina, and occasional micropinocyticvesicles.[9] Immunophenotypically, MS typically expresses α-SMA with variable expression of muscle actin, calponin, collagen IV, laminin, CD99, CD57, BCL-2, CD68, and occasional desmin, while lacking S-100, epithelial markers, nuclear β-catenin, and h-caldesmon.[15] In the present case, vimentin and α-SMA confirmed myofibroblastic differentiation, whereas diffuse p53 overexpression, a high Ki-67 labelling index, and marked cytological atypia indicated aggressive biological behaviour. Negative desmin, CD68, and S-100 excluded major spindle-cell mimics. Although electron microscopy may provide confirmatory evidence, the diagnosis was established by integrating the characteristic morphology with the immunophenotype (vimentin+, α-SMA+, p53+, desmin−, CD68−, S-100−). [2,3,5,8,10]

Complete surgical excision with negative margins remains the treatment of choice; however, local recurrence and distant metastasis are common.[16] Reported recurrence rates range from 32% to 75%, with metastasis occurring in up to 68% of cases, while the benefit of adjuvant radiotherapy or chemotherapy remains uncertain because of limited evidence.[3,5,17] In the present case, despite referral to a tertiary oncology centre and initiation of single-agent doxorubicin, the patient deteriorated rapidly and died approximately four months after presentation, consistent with the tumour's aggressive clinicopathological profile. Thepresent case emphasizes the value of integrating histological grading with proliferative and immunohistochemical markers for accurate risk assessment and highlights the need for prompt multidisciplinary management and larger studies to refine grading criteria and identify reliable prognostic biomarkers.

Conclusion:-

Myofibroblastic sarcoma is a rare malignant spindle-cell neoplasm with significant diagnostic challenges due to its overlapping histopathological features and the lack of standardized grading criteria for aggressive lesions. This case emphasizes the value of integrating histopathological, immunohistochemical, and proliferative markers to accurately diagnose and assess tumour aggressiveness. Despite an FNCLCC Grade 2 classification, the high Ki-67 labelling index, diffuse p53 overexpression, marked cytological atypia, tumour necrosis, and rapid clinical progression indicated aggressive biological behaviour. Early diagnosis, multidisciplinary management, and close follow-up are essential, while larger studies are needed to refine grading criteria and improve prognostic assessment in this uncommon malignancy.

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