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A Benign Mask for a Malignant Face: Primary Leiomyosarcoma of the Maxillary Anterior Region

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Abstract

Background: Leiomyosarcoma of the oral and maxillofacial region is a rare malignant neoplasm that often present diagnostic challenges due to overlapping clinical, radiographic, and histopathological features with other spindle cell tumors. The maxillary anterior region is a particularly uncommon site for this tumor.

Case presentation: A 31-year-old female presented with a progressively enlarging swelling in the left maxillary anterior region. The patient had undergone an incisional biopsy four months earlier, which was reported as a benign reactive fibrous lesion. Clinical examination and Cone Beam Computed Tomography (CBCT) revealed aggressive features, including diffuse osteolytic bone destruction and periosteal reaction. Excisional biopsy with segmental bony resection was performed. Histopathological examination demonstrated a malignant spindle cell neoplasm and initial immuno-histochemical evaluation supported a diagnosis of spindle cell rhabdomyosarcoma. The patient was subsequently referred to a higher oncologic center where hemi-maxillectomy with reconstruction was performed using an anterolateral thigh flap. Histopathological evaluation of the wide excision specimen demonstrated smooth

muscle differentiation, confirming a final diagnosis of primary leiomyosarcoma of the maxilla.

Conclusion: This case highlights the diagnostic complexity of spindle cell malignancies of the oral cavity the potential for diagnostic revision between closely mimicking entities on limited sampling and the importance of clinico-radiographic correlation, adequate tissue sampling and multidisciplinary management in achieving an accurate final diagnosis.

Keywords: Leiomyosarcoma, Spindle Cell Sarcoma, Maxillary Anterior Region, Oral Malignancy, Diagnostic Dilemma, Rhabdomyosarcoma.

Introduction

Leiomyosarcoma is a malignant neoplasm arising from smooth muscle differentiation, most commonly encountered in the uterus, gastrointestinal tract, and retroperitoneum. Primary involvement of the head and neck is rare and intraosseous leiomyosarcoma of the jaws is exceptionally uncommon, likely arising from smooth muscle within vascular walls given the paucity of intrinsic smooth muscle in maxillofacial tissues [2,5]. The maxilla is affected more often than the mandible and lesions confined to the anterior maxillary region are especially rare [2].

Spindle cell malignant neoplasms of the oral and maxillofacial region are uncommon and represent a diagnostic challenge due to their overlapping clinical, radiographic, and histopathological characteristics.[1] These tumors frequently display heterogeneous morphology, making precise classification difficult, particularly when biopsy specimens are limited. The maxilla is an unusual site for primary spindle cell sarcomas and lesions involving the anterior region are exceedingly rare.[2]

Clinically, such tumors may present as firm, painless swellings that resemble benign reactive proliferations, resulting in potential delay in definitive diagnosis. Clinically, leiomyosarcoma and its histologic mimics may present as firm, painless swellings that resemble benign reactive proliferations, resulting in potential delay in definitive diagnosis.[3] Radiographic evaluation may reveal aggressive features such as poorly defined osteolytic destruction and periosteal reaction; however, definitive diagnosis requires comprehensive histopathological examination supplemented by immunohistochemistry, since leiomyosarcoma closely mimics other spindle cell malignancies both morphologically and to a lesser extent, immunophenotypically. [1]

The differential diagnosis of leiomyosarcoma in the maxillofacial region includes spindle cell rhabdomyosarcoma, malignant peripheral nerve sheath tumor and other undifferentiated spindle cell sarcomas.[4] Because desmin positivity is shared between smooth muscle and skeletal muscle lineages, distinguishing leiomyosarcoma from rhabdomyosarcoma can require a broader immunohistochemical panel — smooth muscle actin and h-caldesmon favoring smooth muscle differentiation, myogenin and MyoD1 favoring skeletal muscle differentiation — and variation in interpretation may occur, particularly in limited incisional biopsy specimens [1]. The present case illustrates this diagnostic complexity: an initial impression favoring spindle cell rhabdomyosarcoma was revised to a final diagnosis of leiomyosarcoma on evaluation of the complete resection specimen, underscoring the importance of adequate tissue sampling and multidisciplinary evaluation in suspicious intraoral lesions.

Case Presentation

A 31-year-old female patient reported to the Department of Oral and Maxillofacial Surgery, with a chief complaint of swelling in her upper front tooth region since one

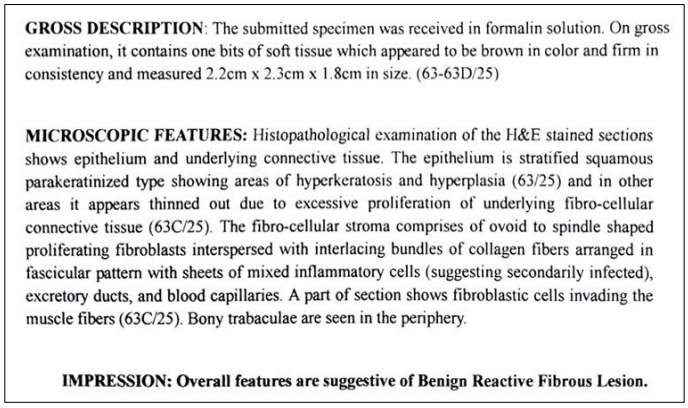
month. The patient gave a history of gradually enlarging swelling in her left upper anterior region, which was initially small in size and progressed to its present dimensions over a period of one month. The swelling was associated with mild intermittent pain with no specific aggravating or relieving factors.

The patient had no significant medical history and no known drug allergies. Past dental history revealed a history of a lesion in the same region approximately four months prior, for which an incisional biopsy had been performed at a private dental clinic. The histopathological report at that time was suggestive of a benign reactive fibrous lesion; however, the swelling persisted and subsequently increased in size.

On extra oral examination, a diffuse swelling was noted over left middle third of face, extending from philtrum to corner of mouth anteroposteriorly, obliterating philtrum and left nasolabial fold. The swelling was non tender on palpation, firm in consistency, non-compressible, non-fluctuant and no local rise in temperature noted.

Intraoral examination revealed a solitary swelling measuring approximately 4*2 cm noted in labial vestibule extending from midline, associated with teeth-21,22,23,24 and till mesial aspect of 25. The swelling involved the labial vestibule, extending into interdental papilla and marginal gingiva of involved teeth. The surface appeared smooth, globular, non-erythematous. On palpation, swelling is firm in consistency, non-tender and no active bleeding on probing noted. Grade 1 mobility noted irt 22.

Based on clinical findings, a provisional diagnosis of fibrous lesion was made. Routine haematological investigations were advised and Cone Beam Computed Tomography (CBCT) was obtained.



GROSS DESCRIPTION: The submitted specimen was received in formalin solution. On gross examination, it contains one bits of soft tissue which appeared to be brown in color and firm in consistency and measured 2.2cm x 2.3cm x 1.8cm in size. (63-63D/25)

MICROSCOPIC FEATURES: Histopathological examination of the H&E stained sections shows epithelium and underlying connective tissue. The epithelium is stratified squamous parakeratinized type showing areas of hyperkeratosis and hyperplasia (63/25) and in other areas it appears thinned out due to excessive proliferation of underlying fibro-cellular connective tissue (63C/25). The fibro-cellular stroma comprises of ovoid to spindle shaped proliferating fibroblasts interspersed with interlacing bundles of collagen fibers arranged in fascicular pattern with sheets of mixed inflammatory cells (suggesting secondarily infected), excretory ducts, and blood capillaries. A part of section shows fibroblastic cells invading the muscle fibers (63C/25). Bony trabaculae are seen in the periphery.

IMPRESSION: Overall features are suggestive of Benign Reactive Fibrous Lesion.

Figure 1: Initial histopathological report of incisional biopsy suggestive of Benign Reactive Fibrous Lesion

Radiographic findings

CBCT was advised to assess the extent and nature of the lesion. CBCT revealed a diffused osteolytic lesion in relation to teeth 22 and 23, exhibiting a moth-eaten pattern of bone destruction with poorly defined margins. Destruction of the labial cortical plate was evident. A periosteal reaction suggestive of a sunburst appearance was noted in the affected region. Circumferential widening of the periodontal ligament space was observed in relation to teeth 21, 22, and 23, with associated crestal and apical bone loss. No evident crown or root pathology was detected in the involved teeth.

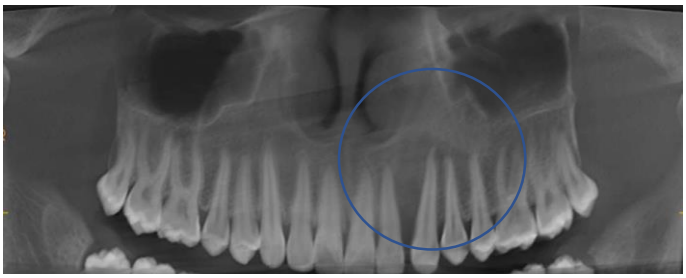


Figure 2: Orthopantomogram (OPG) showing radiolucent lesion in the left maxillary anterior region with associated bone destruction in relation to teeth 22–24

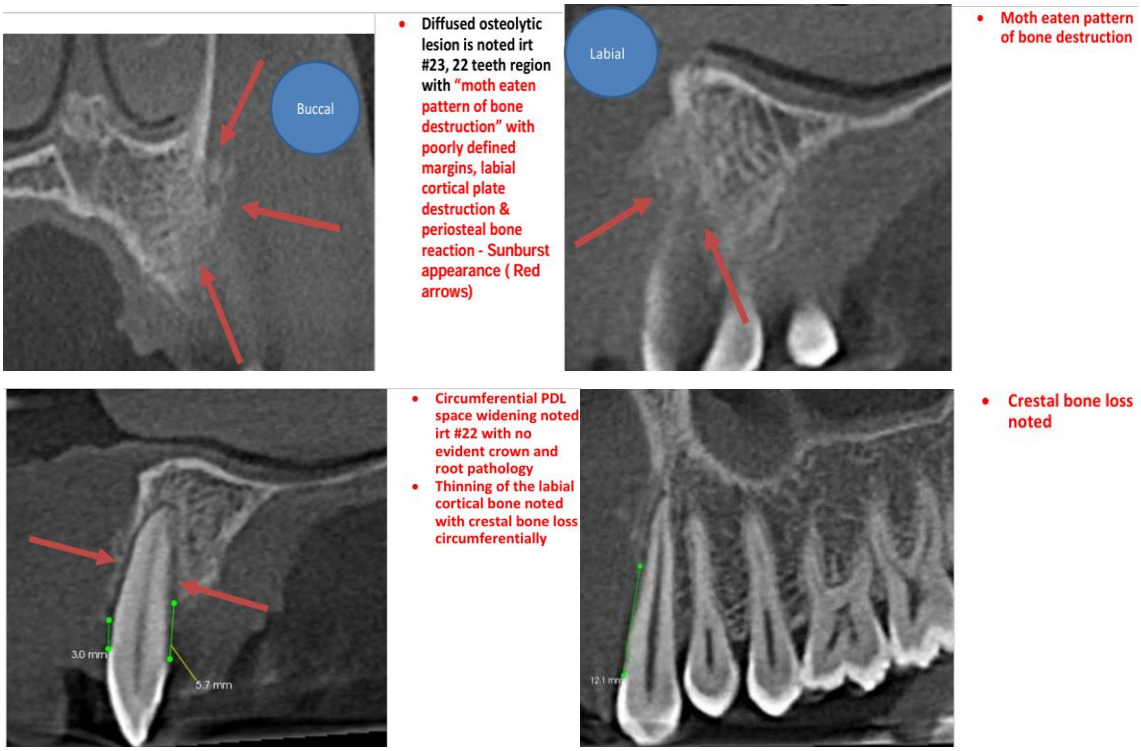


Figure 3: CBCT images demonstrating diffused osteolytic lesion in the left maxillary anterior region (22–23) with moth-eaten pattern of bone destruction, poorly defined margins, labial cortical plate destruction, periosteal reaction (sunburst appearance), circumferential periodontal ligament space widening, and associated crestal bone loss.

Surgical procedure and Biopsy findings

In view of the previous incisional biopsy report suggestive of a benign reactive fibrous lesion and the persistent progressive enlargement of the swelling, an excisional biopsy was undertaken to establish a definitive diagnosis.

The patient was prepared and draped under universal aseptic precautions. Local anaesthesia was achieved using bilateral infraorbital and nasopalatine nerve blocks. A crevicular incision was placed extending from tooth 12 to 25 with releasing incisions and a full-thickness mucoperiosteal flap was reflected. A soft tissue mass, clinically resembling fibrous tissue was identified on the labial aspect extending from the 21 to 24 region. The lesion was carefully dissected, excised and sent for histopathological examination.

On further exploration, irregularities of the buccal cortical plate were noted extending from 22 to 24 region. Considering the extent of bony involvement, teeth 22, 23, and 24 were extracted, and segmental bony resection was performed up to approximately 2 mm below the nasal floor in relation to 22–24 region. The resected specimen was submitted for histopathological analysis.

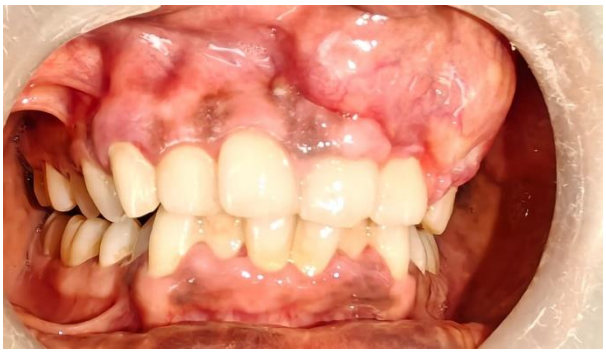


Figure 4: Preoperative intraoral photograph showing solitary swelling in the left maxillary anterior labial vestibule extending from 21 to 24 region

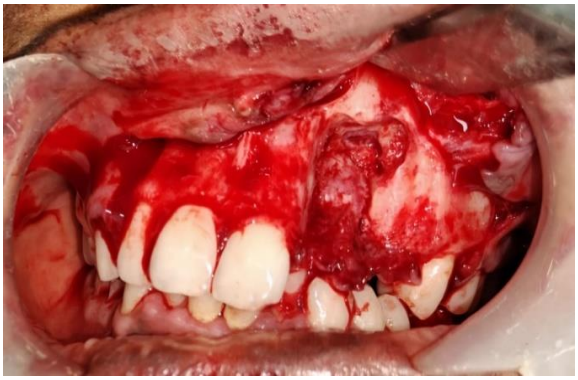


Figure 5: Intraoperative view showing mucoperiosteal flap reflection with extraction of teeth 22, 23, and 24 and segmental bony resection performed up to approximately 2 mm below the nasal floor in the left maxillary anterior region

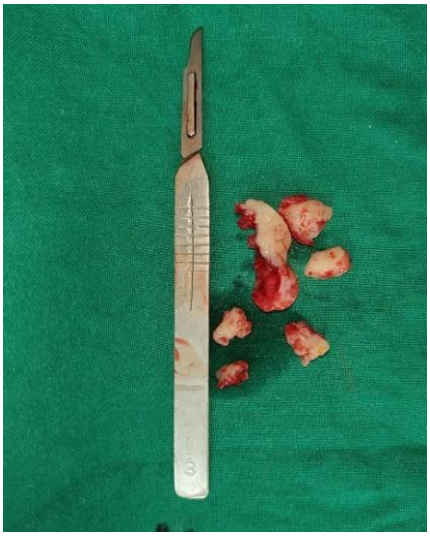


Figure 6: Excised soft tissue mass specimen

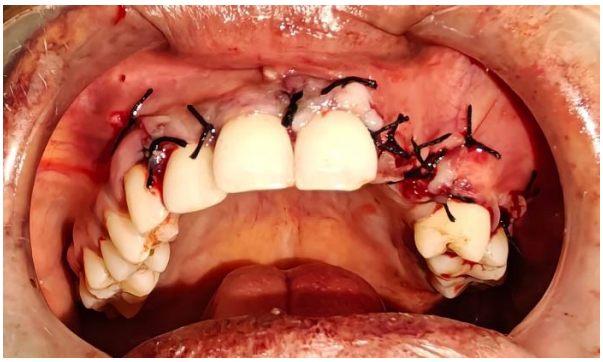


Figure 7: Postoperative intraoral view after segmental resection and primary closure in the left maxillary anterior region.

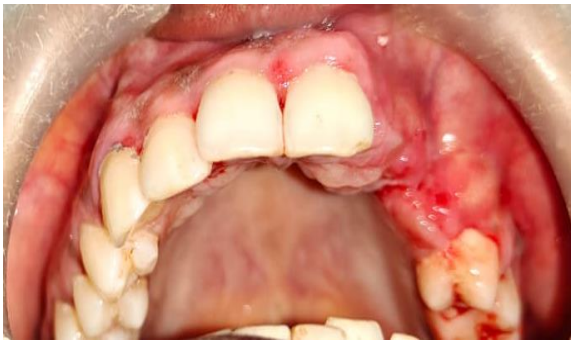


Figure 8: One-month postoperative clinical follow-up after doing excisional biopsy

Histopathological and Immuno-histochemical findings

Microscopic examination revealed a highly cellular spindle cell neoplasm arranged in interlacing fascicles. The tumor cells exhibited elongated nuclei with nuclear pleomorphism and occasional atypical mitotic figures. The lesion demonstrated an infiltrative growth pattern with extension into adjacent skeletal muscle. No epithelial differentiation was identified. The initial impression was suggestive of a malignant spindle cell tumor and immunohistochemical evaluation was advised for further characterization.

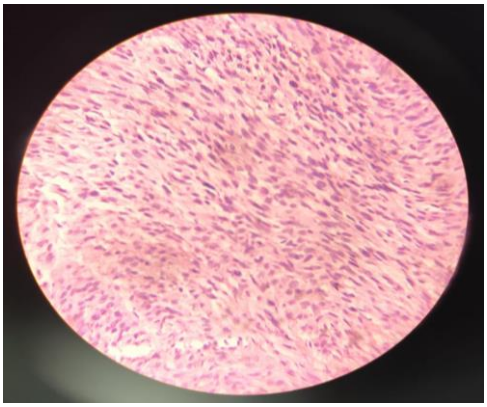


Figure 9: Photomicrograph showing highly cellular spindle cell neoplasm arranged in interlacing fascicles with nuclear pleomorphism and atypical mitotic figures (H&E stain, ×40), initially suggestive of malignant peripheral nerve sheath tumor.

Immuno-histochemical analysis of the excisional biopsy specimen- the neoplastic spindle cells are positive for CK, Bcl2, Desmin, Myogenin, MyoD1, TLE1 and SMA

supported a diagnosis of spindle cell rhabdomyosarcoma. The proliferative index (Ki-67) was approximately 25%, indicating active tumor proliferation.

Further Management and Follow- up

The patient was referred to a higher oncologic center for definitive management. She subsequently underwent hemimaxillectomy and reconstruction was performed using an anterolateral thigh flap.

Histopathological evaluation of the wide excision specimen revealed a spindle cell sarcoma infiltrating the maxillary bone. Repeat/extended immunohistochemical evaluation- PANCK, H CALDESMON, SATB2- demonstrated smooth muscle differentiation, confirming final diagnosis of leiomyosarcoma.

The patient has been placed under regular postoperative surveillance and continues to be on close follow-up.

Discussion

Leiomyosarcoma of the oral and maxillofacial region is an uncommon malignant neoplasms that frequently presents significant diagnostic challenges due to overlapping clinical and histopathological characteristics with other spindle cell sarcomas.[1] These tumors often demonstrate heterogeneous morphology and limited biopsy samples may not fully represent the aggressive component of the lesion. In the present case, the lesion initially mimicked a benign reactive fibrous proliferation both clinically and histologically and the malignant spindle cell neoplasm on excisional biopsy was initially believed, on immunohistochemistry, to represent spindle cell rhabdomyosarcoma rather than leiomyosarcoma — a distinction that was only resolved on the wide excision specimen. This highlights the potential for diagnostic revision in heterogeneous, closely mimicking spindle cell malignancies, particularly on limited tissue sampling.

Sarcomas of the head and neck constitute a small proportion of all malignancies in this region and intraoral involvement is relatively rare.[7] The maxilla, particularly the anterior region, is an unusual site for primary spindle cell sarcomas.[2,5] Spindle cell/ sclerosing rhabdomyosarcoma, the closest histologic mimic in this case, represents a rare variant that may involve the oral region and show considerable histologic overlap with leiomyosarcoma.[6]

Clinically, both leiomyosarcoma and spindle cell rhabdomyosarcoma often present as painless, progressively enlarging swellings that may resemble reactive or inflammatory lesions.[3] This overlap can delay definitive diagnosis, particularly when initial biopsy findings are inconclusive. In the present case, the earlier incisional biopsy was reported as a benign reactive fibrous lesion, which underscores the limitations of sampling in heterogeneous spindle cell neoplasms.[1] The aggressive clinical progression and radiographic findings prompted further surgical intervention and comprehensive histopathological evaluation.

Radiographically, malignant spindle cell tumors may demonstrate poorly defined osteolytic destruction and cortical plate breach, reflecting their infiltrative nature.[3] Such features were evident in this case, supporting suspicion of malignancy despite the previous benign report. Correlation between clinical behavior and radiographic findings is therefore critical in identifying lesions that warrant more extensive surgical management.

The differential diagnosis of spindle cell malignancies in the oral cavity includes leiomyosarcoma, spindle cell rhabdomyosarcoma, malignant peripheral nerve sheath tumor, and other undifferentiated sarcomas.[4] Since desmin is expressed in both smooth muscle and skeletal muscle differentiation, an initial panel weighted toward skeletal-muscle markers, or a limited biopsy showing only desmin positivity, can favor an initial impression of

rhabdomyosarcoma that is subsequently revised once smooth-muscle-specific markers such as smooth muscle actin and h-caldesmon are evaluated on the complete specimen — the sequence observed in the present case. Studies have emphasized the clinical and molecular heterogeneity of spindle cell rhabdomyosarcoma in the head and neck region, further complicating diagnostic interpretation.[10] Likewise, adult rhabdomyosarcoma has been associated with variable outcomes and aggressive behavior, necessitating multimodality treatment approaches.[9]

Head and neck sarcomas require prompt recognition and coordinated multidisciplinary management to improve outcomes. Wide surgical excision with adequate margins remains the cornerstone of treatment for localized disease.[2] In the present case, the patient underwent definitive surgical management at a higher oncologic center following initial excision, reflecting the importance of timely referral and comprehensive oncologic evaluation.

This case reinforces the diagnostic complexity associated with leiomyosarcoma of the maxillary anterior region and its overlap with spindle cell rhabdomyosarcoma. Persistent or progressively enlarging intraoral swellings should be approached with caution, particularly when clinical and radiographic findings suggest aggressive behavior. Adequate tissue sampling, careful histopathological assessment, and multidisciplinary collaboration are essential for achieving accurate diagnosis and appropriate treatment planning.

Conclusion

Primary Leiomyosarcoma of the maxillary anterior region is a rare but diagnostically demanding group of malignancies, particularly when initial biopsy findings are inconclusive and when its immunophenotype overlaps with spindle cell rhabdomyosarcoma, as in this case. This case underscores the critical importance of correlating clinical progression and radiographic aggressiveness with histopathological interpretation, especially in lesions that persist despite prior benign diagnosis. The evolution of diagnosis in this patient highlights the inherent heterogeneity of spindle cell neoplasms and the limitations of limited tissue sampling. Early suspicion, comprehensive evaluation and timely multidisciplinary referral are essential to prevent diagnostic delay and ensure optimal oncologic management. Oral and maxillofacial surgeons should carefully evaluate persistent or rapidly growing intraoral swellings to avoid overlooking potentially aggressive lesions such as leiomyosarcoma.

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