

CASE REPORT

Primary leiomyosarcoma of the maxilla: An investigative loom—report of a challenging case and review of literature

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ABSTRACT

Leiomyosarcoma (LMS) is a malignant neoplasm composed of cells showing distinct smooth muscle features. Majority of the tumors are located in the retroperitoneum, including the pelvis and the uterus but are rare in the oral and pharyngeal region. Intraorally, they are present as painless, lobulated, fixed masses of the submucosal tissues in middle-aged or older individuals. Lesions are usually slow growing and are less than 2 cm in diameter at the time of diagnosis. Here we report the clinico-pathological findings of a case of primary LMS of the maxilla in 63-year-old male patient with an emphasis on the judicious use of ancillary diagnostic modalities to arrive at a definitive diagnosis.

Keywords: Leiomyosarcoma, malignant spindle cell tumor, nuclear palisading, pleomorphic undifferentiated sarcoma, rhabdomyosarcoma, soft tissue sarcoma

INTRODUCTION

Leiomyosarcomas (LMSs) are relatively rare mesenchymal neoplasms composed of cells that exhibit smooth muscle differentiation. They account for 5–10% of soft tissue sarcomas.^[1,2] These adult soft tissue sarcomas are far outnumbered by the more commonly occurring liposarcomas. Majority of these tumors are located in the retroperitoneum, including the pelvis and the uterus. LMS is a rare malignancy of the oral and pharyngeal region of unknown etiology. According to a review of smooth muscle tumors, only 0.06% occurred in the oral region.^[1,3] Although no definite causative factor has yet been identified, correlation has been established between LMS and exposure to radiations and chemicals and possibly trauma. Association of LMSs with hereditary (bilateral) retinoblastoma (Rb), which appears to result directly from mutations or deletions in the Rb 1 gene, suggests the role of chromosomal defects in the etiology of the tumor.^[4]

These tumors arise from smooth muscle cells, especially those found in blood vessel walls and from undifferentiated mesenchymal cells. Often, the tumor completely obliterates its origin in the blood vessel walls.^[2,4]

The sex predilection of LMSs depends on the location of the tumor. Retroperitoneal and inferior vena cava LMSs are more common in women. On the other hand, the oral tumors do not display any significant gender predilection. LMSs present intraorally as painless, lobulated, fixed masses of the submucosal tissues in middle-aged or older individuals but are rarely found in children.^[5-7] Oral lesions are usually less than 2 cm in diameter at diagnosis and are slow-growing, but secondary ulceration of the mucosal surface has been reported in a few cases.^[2,6]

Histological appearances differ according to the degree of differentiation but the presence of a focus of fascicles of elongated brightly eosinophilic spindle cells with vesicular, ovoid to cigar-shaped nuclei showing a strong positivity for smooth muscle actin (SMA) and/or desmin is a minimum requisite.^[8] In well-differentiated lesions, there is fascicular streaming of spindled cells in a manner similar to that seen in a leiomyoma. About 10% of LMSs are anaplastic, which in the extreme cases may show pleomorphic undifferentiated sarcoma (PUS)/malignant fibrous histiocytoma (MFH) like pleomorphism. In these tumors, numerous pleomorphic giant cells with deeply eosinophilic cytoplasm are admixed with a set of more uniform-appearing spindle and round cells.

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The purpose of this compilation is to present a case report of a primary LMS of maxilla in a 63-year-old male with a stress on comprehensive morphological analyses and careful interpretation of immunohistochemical markers to arrive at a correct diagnosis.

CASE REPORT

A 63-year-old male patient reported to the out-patient department of the Dental School with a 6-month history of progressive and continuous enlargement in the anterior maxilla. The growth was painless and non-tender and had loosened the upper front teeth. On examination, the growth was bosselated and arose from the anterior maxillary alveolus extending from right canine to left lateral incisor. There was a palatal extension in the antero-posterior direction. It was firm on palpation and demonstrated a patchy bluish discoloration. Based on the above findings, a clinical diagnosis of malignancy of maxilla was made. Computerized tomography scan showed an osteolytic lesion extending in the nasal chamber but not laterally in the maxillary sinuses. The axial section showed a diffuse soft tissue mass obliterating the anterior nasal chamber completely destroying the anatomy of the anterior palate and the nasal cartilaginous skeleton [Figure 1].

Because of the non-specific clinical presentation, based on the age of the patient and location of the tumor, the differential diagnoses include a salivary gland tumor, a sarcoma and squamous cell carcinoma and incisional biopsy was planned under local anesthesia. Analysis of the biopsy specimen revealed a tumor mass composed of fascicles of interlacing spindle-shaped cells with abundant eosinophilic cytoplasm and moderately large nuclei exhibiting atypia. Based on the above features, a provisional diagnosis of a malignant spindle cell tumor was made. The differential diagnosis of spindle-shaped lesions included malignant lesions such as PUS, rhabdomyosarcoma (RMS), LMS,

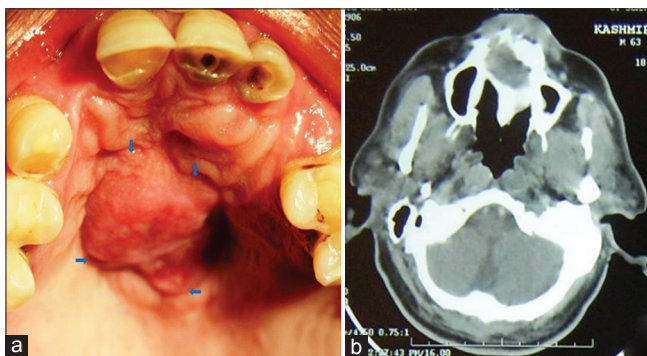


Figure 1: (a) Intraoral photograph showing bosselated growth in the anterior maxilla with patchy bluish discoloration extending from right canine to left lateral incisor with palatal extension in the anteroposterior direction involving the midline. (b) Computed tomography (CT) axial section showing a diffuse soft tissue mass obliterating the anterior nasal chamber. The lesion is also seen extending in the anterior maxillary alveolus area completely destroying the anatomy of the anterior palate and the nasal cartilaginous skeleton

malignant peripheral nerve sheath tumor (MPNST) and fibrosarcoma.

Since the lesion showed aggressive clinical and radiological features suggestive of malignancy, resection of tumor with safe wide margins was planned under general anesthesia. The tumor was safely resected with approximately 1.5 cm of safe margin in the healthy tissue. A 2.4-mm titanium reconstruction plate was affixed in the residual maxilla to support a prosthesis in future. In order to rule out any primary or secondary foci of malignancy, a body scan was performed and the results were negative. The patient has been on regular follow-up for the last 2 years with no signs of recurrence. A tooth bearing obturator was delivered and the patient is presently satisfied with its appearance and function.

Gross findings

The excised dentate hemimaxilla measuring 8 cm × 7.5 cm in dimension; reddish-brown in color and firm in consistency was received [Figure 2]. The cut surface of the tumor mass was solid, fibrous and yellowish-white in color with foci of hemorrhage and necrosis.

Histopathological findings

Histopathologically, the tumor mass was composed of elongated spindle cells arranged in interlacing bundles in interweaving fascicles of varying size. The cells showed abundant eosinophilic cytoplasm and moderately large centrally located blunt ended nuclei with mild atypia. Cytoplasmic vacuolation was also seen. Hyperchromatism of nuclei was pronounced and numerous normal and abnormal mitotic figures were scattered throughout the lesion. Mitotic figures (MF) were abundant and ranged up to 22 MF/10 HPF (high power fields). Necrotic foci were scattered throughout the tumor. The stroma was composed of a delicate network of capillaries [Figure 3]. Numerous pleomorphic giant cells with deeply eosinophilic cytoplasm was intimately admixed with more typical cells. Pleomorphic areas imparting a pleomorphic appearance were abundant in the tumor hence a provisional diagnosis of pleomorphic sarcoma was made



Figure 2: Gross appearance of the excised dentate hemimaxilla. The tumor mass measured 8 cm × 7.5 cm in dimensions and was creamish brown in color with color variegation ranging from brown to yellow and was firm in consistency

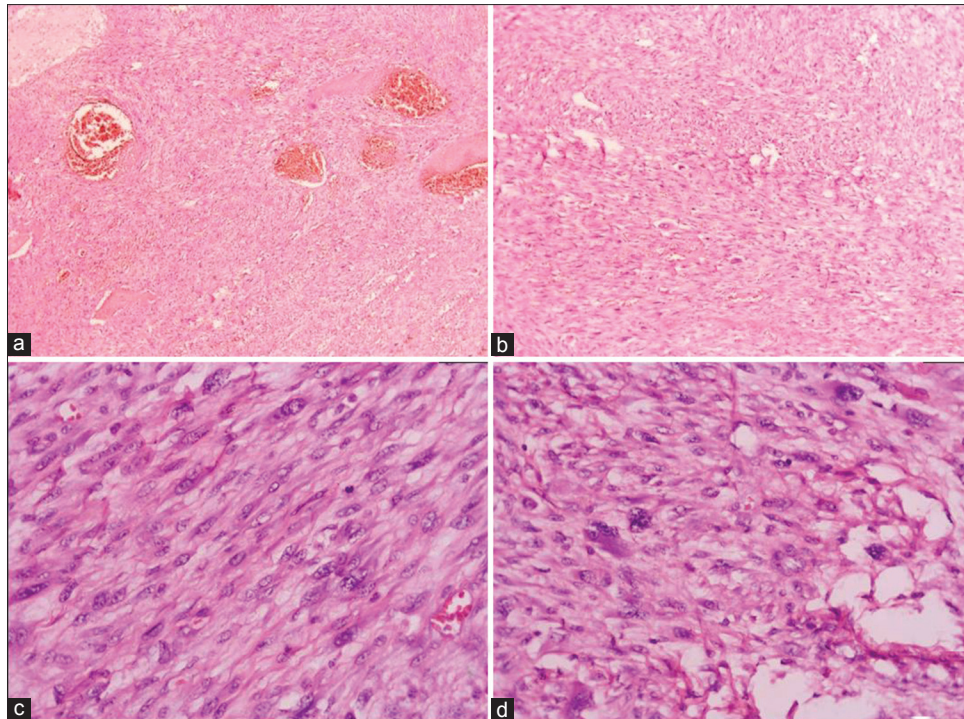


Figure 3: Photomicrographs showing a) Low power view of a tumor mass with numerous engorged vascular channels (H&E stain, x40). (b) Tumor mass composed of fascicles of interlacing spindle-shaped cells with abundant eosinophilic cytoplasm and moderately large nuclei exhibiting atypia (H&E stain, x40). (c) Elongated cells with abundant cytoplasm exhibiting centrally placed blunt ended nuclei (H&E stain, x100). (d) Bizarre tumor cells with nuclear pleomorphism interspersed among regular spindle-shaped cells (H&E stain, x100)

and the spectrum included tumors exhibiting prominent pleomorphism like PUS (storiform-pleomorphic MFH), pleomorphic liposarcoma, pleomorphic leiomyosarcoma and pleomorphic RMS [Figure 4]. The tumor was graded according to the French grading system as described by Coindre *et al.*^[9] With this system, the differentiation grade of tumors, the number of MF/2 mm² and the amount of necrosis are scored. All tumors were divided into grade 1, grade 2 or grade 3 tumors. The mitotic index (number of MF/2 mm²) was determined on hematoxylin and eosin (H and E) stained sections of the tumors; the areas with the highest rates of mitosis were selected.

The histopathological uncertainty prompted an immunohistochemical analysis. For immunohistochemical staining, the Strept Avidin Biotin (SAB) method was used with primary antibodies to vimentin, SMA, Muscle Specific Actin (HHF35), pancytokeratin, desmin, myogenin, h-caldesmon, CD31, CD34, S-100 protein, human melanoma black (HMB) 45 and α 1 anti-chymotrypsin. The tumor cells showed a strong immunopositivity for vimentin with focal positivity for SMA and muscle HHF-35 [Figure 5]. The tumor cells were negative for pancytokeratin, Epithelial Membrane Antigen (EMA), desmin, myogenin, h-caldesmon, CD31, CD34, S-100 protein and HMB45 [Table 1].

Based on the above parameters, a diagnosis of Pleomorphic Leiomyosarcoma Grade 3/3 (Coindre Grading System) was made.

DISCUSSION

LMSs are malignant neoplasms composed of cells showing distinct smooth muscle features.^[1] They comprise a heterogeneous group of neoplasms, each with unique clinical, histological and radiographic characteristics.^[10]

LMS is listed as a “rare disease” by the Office of Rare Diseases (ORD) of the National Institute of Health (NIH).^[11] It is rarer in the head and neck region and most commonly affects the nose and paranasal sinuses, skin, subcutaneous tissue and the cervical esophagus.^[1] In the oral cavity, the maxillary sinus, mandible and maxilla appear to be the predilection sites for LMS, but other reported intraoral locations are the cheek, floor of the mouth, tongue, hard and soft palate, lips and gingiva.^[1,5] The rare occurrence of LMS in the oral cavity has been correlated to the scarcity of smooth muscle structures in this location, as compared with their abundance in other sites. LMSs occurring in the oral tissues in all probability arise from tunica media of blood vessels, arrectores pilorum, circumvallate papillae, myoepithelial cells and pluripotential undifferentiated mesenchymal cells.^[1,4]

The tumor lacks specific age or sex predilection.^[3,5] Oral and maxillofacial LMSs are rare and have no specific signs and symptoms, usually presenting as non-ulcerated painless masses.^[6] As a result, some of these lesions are occasionally mistaken for the other common lesions affecting the oral cavity and correct diagnosis is made only after a judicious histologic

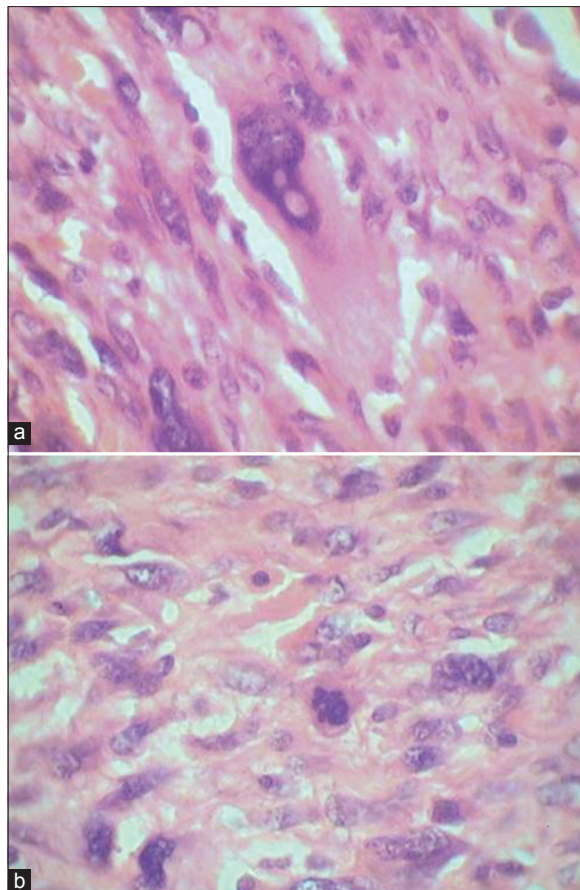


Figure 4: Photomicrographs showing pleomorphism in leiomyosarcoma. (a) Bizarre spindle cell exhibiting deeply eosinophilic cytoplasm with pleomorphic nuclei and nuclear vacuolization (H&E stain, ×400). (b) Cells exhibiting extreme degree of pleomorphism and occasional multinucleation resembling pleomorphic undifferentiated sarcoma/malignant fibrous histiocytoma (H&E stain, ×400)

examination. The clinical differential diagnosis for a palatal lesion includes benign and malignant salivary gland tumors (pleomorphic adenoma, mucoepidermoid carcinoma, adenoid cystic carcinoma, polymorphous low-grade adenocarcinoma) and benign and malignant mesenchymal tumors.^[5]

Because of lack of a distinct radiographic presentation, these tumors can simulate any expansive or destructive lesion of the jaw.^[6] They can present as lytic lesions with ill-defined margins, periosteal elevation, calcification and cortical destruction.^[12]

Microscopic diagnosis of the smooth muscle lesions is difficult due to the small biopsy sizes and the diversity of the lesions.^[13] Morphological features in favor of smooth muscle

Table 1: Immunohistochemical expression of the lesional cells

IHC marker reaction

Vimentin: Strong positivity
Smooth muscleactin (SMA): Focal positivity
Muscle specific actin (HHF-35): Focal positivity
Pancytokeratin: Negative
EMA: Negative
Desmin: Negative
Myogenin: Negative
Caldesmon: Negative
CD31: Negative
CD34: Negative
S100: Negative
HMB45: Negative
α1-antichymotrypsin: Negative

IHC: Immunohistochemistry, EMA: Epithelial membrane antigen

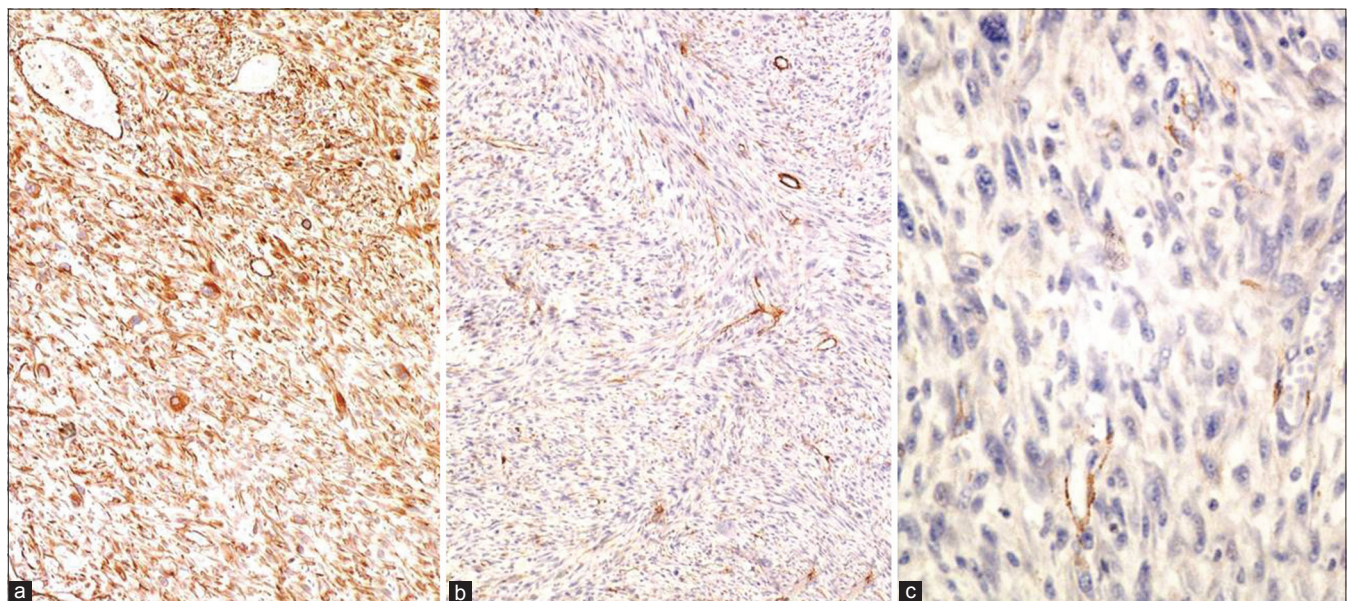


Figure 5: Photomicrographs showing (a) Strong positive immunohistochemical reaction to antibodies against vimentin (IHC stain, ×100), (b) Focal expression of smooth muscle actin (SMA) in the tumour cells (IHC stain, ×100), (c) Focal reaction to muscle specific actin (HHF-35) by the tumor cells (IHC stain, ×400)

differentiation include intersecting fascicles of spindle cells, eosinophilic cytoplasm, paranuclear vacuoles and blunt-ended nuclei. Para nuclear vacuole is seen at one end of the nucleus, causing a slight indentation, so the nucleus assumes a concave rather than a convex contour. Size, cellularity, atypia, necrosis and mitoses per HPF are indicators that help define the difference between a benign smooth muscle tumor and LMS. Of these indicators, mitoses per HPF is considered to be the most reliable.^[1,14]

In poorly differentiated LMS, lesional cells are less elongated, more fusiform or rounded, enlarged and pleomorphic.^[5] Hyperchromatism of nuclei is often pronounced and numerous normal and abnormal mitotic figures are scattered throughout the lesion. Focal areas may contain giant cells with multiple, pleomorphic, even bizarre nuclei.^[6,8] The stroma is typically sparse, but cellular streaming is usually far less regular than in the low grade lesions, although the fascicles may be as uniform as those of well-differentiated lesions.^[15] Hemorrhage, focal necrosis, increased vascularity and focal myxoid change are not uncommon features of poorly differentiated lesions.^[1,6,12]

A score of 1 is currently assigned to sarcomas closely resembling normal adult tissue to such a degree as to be confused with benign tumors, such as a well-differentiated LMS. A score of 3 is given to embryonal and poorly differentiated sarcomas, sarcomas of doubtful histologic type, synovial sarcoma, primitive neuro-ectodermal tumor, osteosarcoma, pleomorphic RMS and pleomorphic liposarcoma. Other sarcomas of certain histologic type such as myxoid liposarcoma are scored 2. Mitotic count is obtained in 10 successive HPFs in most mitotic areas. This count is taken to establish the score: Score 1 for 0 to 9 mitoses; score 2 for 10 to 19 mitoses; and score 3 for more than 19 mitoses per 10 HPF.^[9]

Our case represented Pleomorphic LMS Grade 3/3 (Coindre Grading System). It is a morphologic variant rather than a distinctive subtype and is usually associated with areas of undifferentiated pleomorphic sarcoma along with fields showing morphological, immunohistochemical or ultrastructural evidence of smooth muscle differentiation.

Although the present case displayed features of smooth muscle differentiation to some extent, the correct light-microscopic diagnosis was difficult, mainly because the tumor was pleomorphic, resembling a PUS (MFH).

The differential diagnosis of LMSs includes other sarcomas composed of fascicles of moderately differentiated spindle cells, such as fibrosarcoma and MPNST. Although the low-power appearance of all three can be similar, there is a greater tendency to see a close juxtaposition of longitudinally and transversely cut fascicles in a LMS. The cytological features play an important role in the differential diagnosis. Compared with the cells of LMS, those of a fibrosarcoma tend to be tapered and those of a MPNST are wavy, buckled and distinctly

asymmetric.^[1,5,6,15,16] Vimentin, SMA, desmin, h-caldesmon are positive in LMS but fibrosarcoma is diagnosed by exclusion as it is S-100 positive [Table 2]. Thus, careful scrutiny of cytological details in multiple sections, clinico-pathological correlation and immunohistochemistry (IHC) are mandatory for a definitive diagnosis.

In pleomorphic LMS, the neoplastic cells have more bizarre features with numerous tumor giant cells, high mitotic activity and a prominent storiform pattern. The latter is sometimes so overwhelming that a diagnosis of PUS (MFH) is made on H and E. We encountered a similar problem in our initial diagnosis where the diffuse and prominent storiform arrangement of neoplastic spindle cells favored a diagnosis of PUS. Based on the IHC results a diagnosis of LMS was reached thus emphasizing its role in arriving at a final diagnosis in case of spindle cell tumors.

Originally the tumor was diagnosed as PUS (MFH) based on the whorled structural pattern and considerable cellular pleomorphism. The distinction between pleomorphic LMS and PUS (MFH) is probably the most difficult and controversial. Unless there are light microscopic areas that are diagnostic of smooth muscle differentiation, decision is based upon the immunoreactivity for various myogenic markers reflective of smooth muscle differentiation.^[10] Significant amounts of actin or some degree of both actin and desmin immunoreactivity is required for the diagnosis of pleomorphic LMS.^[1] Peripheral expression of actin immunoreactivity of myofibroblasts in PUS (MFH) contrasts with the diffuse actin immunoreactivity of smooth muscle cells in LMSs and careful evaluation is required to differentiate the two entities. PUS (MFH) are usually positive for α 1-antichymotrypsin and vimentin and are negative for myoglobin and desmin.^[1,4]

Table 2: Tumor immunoreactivity reference chart^[17]

Tumor	IHC marker
Spindle cell carcinoma	AE1, AE3, CK1, CK18, EMA, SMA, Desmin+
Malignant myoepithelioma	CK, SMA, GFAP, CD10, calponin, and smooth muscle Myosin heavy chain+
Melanoma	Vimentin, tyrosinase, melan-A, S-100 protein, and HMB 45 +
Anaplastic large cell lymphoma	CD 30 +
Fibrosarcoma	Diagnosis by exclusion -S-100 protein-
Rhabdomyosarcoma	Myogenin, desmin, and actin+
Pleomorphic undifferentiated sarcoma/malignant fibrous histiocytoma	α 1-antichymotrypsin and vimentin+, focally actin+
Leiomyosarcoma	Vimentin, SMA, desmin, h-caldesmon and HHF35, transgelin+

EMA: Epithelial membrane antigen, CK: CytoKeratin, SMA: Smooth muscle actin, HHF-35: Muscle specific actin

Table 3: Clinical data of the reported cases of maxillary leiomyosarcoma (LMS)

Author	Year	Age/sex	Site	Histopathological and immunohistochemical profile
TagakiM, et al. ^[3]	1972	56/M	Maxillarygingiva	LMS
WeitznerS ^[10]	1980	39/F	Anteriormaxillary alveolar ridge	LMS
Kawakami, et al. ^[25]	1987	63/F	Maxilla	LMS
TandonD, A et al. ^[14]	1991	35/M	Zygoma and maxillary sinus	LMS
Freedman, et al. ^[15]	1993	27/F	Maxillary gingiva	LMS
MizutaniH, et al. ^[16]	1995	17/F	Upper gingiva	LMS desmin+ve
Izumi, et al. ^[20]	1995	70/M	Maxillarygingiva	LMS, desmin, HHF-35, and myosin+ve
LoMuzioL, et al. ^[8]	2002	31/F	Upper alveolar mucosa	LMS (coindre grade 2) HHF-35 and alpha SMA, desmin and vimentin+ve
Wada S, et al. ^[18]	2002	71/F	Maxilla	LMS
RodiniCO, et al. ^[2]	2007	63/M	Maxillary bone	LMS vimentin, desmin, SMA, laminin, HHF-35+ve S-100, AE1/AE3proteins -ve
Qureshi SS, et al. ^[13]	2008	15/M	Maxilla andethmoid	LMS vimentin and SMA+ve, S-100, LCA -ve
ChewYK, et al. ^[21]	2009	36/M	Maxilla	LMSvimentin and SMA -ve
Park S, et al. ^[19]	2010	64/M	Anterior maxilla	LMS
Arora P, et al. ^[22]	2010	21/M	Gingivoalveolar sulcus	PLMS, vimentin, desmin, CK
ChiuYW, et al. ^[23]	2011	58/M	Maxilla and pterygoid plate	High-gradeLMS HHF-35, SMA and h-Caldesmon+ve
Azevedo RS, et al. ^[24]	2012	71/M 69/M	Tongue and mandible	LMS
Ahn JH, et al. ^[26]	2012	54/F	Tongue	LMS
Iwaia S, et al. ^[27]	2013	24/M	Maxillary gingiva	LMS
Sağlam, et al. ^[28]	2013	20/M	Soft palate	LMS
PapoiianV, et al. ^[29]	2014	83/F	Maxilla (Sinonasal)	LMS

LMS: Leiomyosarcoma, SMA: Smooth muscle actin, HHF-35: Muscle specific actin, CK: Cytokeratin

In this case, discrimination of LMS from RMS was necessary.^[15] The tumors that originate from muscle stain positive for desmin, HHF35 and vimentin; however, this case was positive for α -SMA and the diagnosis of RMS was ruled out. Immunohistochemical demonstration of muscle-specific actin and smooth-muscle-specific actin is a strong indicator for LMS. Also, desmin may be helpful in the diagnosis of LMS, since the intermediate filament desmin is present in both smooth muscle and striated muscle. Vimentin is considered a major component of the intermediate filaments of smooth muscle cells derived from vasculature. Spindle-shaped cells of LMS are usually negative for CKs^[2,8,12-14,16] [Table 2].

Intraoral LMSs are very aggressive tumors with high local recurrence and/or distant metastasis and the survival rate is very low.^[4] Clinical data of reported cases of maxillary leiomyosarcoma is presented in Table 3.

Generally, a complete surgical resection with tumor-free margins is recommended to control local recurrence and adjuvant radiotherapy or chemotherapy is considered to have little beneficial effect on decreasing recurrence of LMS or increasing survival time.^[4] However, in some specific anatomic locations such as the vicinity of the infratemporal fossa, the maxillary sinus, the pterygoid plates and the mandibular condyle, it may be technically less feasible to achieve tumor-free margins because of difficult access,

possibly resulting in residual microscopic disease leading to the local recurrence of the tumor and a poorer prognosis.^[10,16,18] A few cases of metastatic LMS have been described in the oral cavity.^[1] Distant metastases of intraoral LMS appear in up to 39% of cases. In oral LMS, metastasis to regional lymph nodes was relatively rare and the most common site of metastasis was the lungs.^[19,20]

LMS is a relatively rare tumor in the oral and maxillofacial region and has a poor prognosis as a result of high local recurrence.^[4,5] A thorough morphological analyses and careful interpretation of immunohistochemical markers is necessary to arrive at a correct diagnosis.^[21] Accurate diagnosis, classification and multi-modality treatment approach is essential for favorable outcome.^[22-25]

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