Case Report
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Gingival Leiomyosarcoma in a Young Woman: Report of a Challenging Case

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Abstract

Leiomyosarcoma (LMS) is an uncommon malignant mesenchymal tumor. This tumor arises from smooth muscle cells and mostly occurs behind the peritoneum area, including pelvis and uterus. Oral LMS is exceedingly rare and occurs mainly in the bones and the soft tissues. It generally involves adults, without any considerable specific age or sex predominance. This tumor has a clinically aggressive growth pattern and is usually presented as a non-ulcerated, painless, destructive mass with a relatively firm consistency. LMS can be easily mistaken for other more common spindle cell lesions by only light microscopic evaluation; consequently, immunohistochemical study is helpful for the definitive diagnosis. The suggested treatment modalities include excision, radical surgery, radiation therapy, chemotherapy. Wide surgical resection is the most preferred treatment protocol. Herein, we report a clinically controversial case of gingival LMS in a 32-year-old female and also discuss the diagnostic procedures and treatment modalities in this malignancy.

Keywords: Gingiva, Leiomyosarcoma, Mandible, Tumor

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Introduction

Leiomyosarcoma (LMS) is a relatively uncommon mesenchymal neoplasm composed of cells that represent smooth muscle differentiation. It accounts for 5%-10% of the soft tissue sarcomas.[1-4]

The occurrence of leiomyosarcomas in the oral cavity is exceedingly rare. Majority of them are located in

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the retroperitoneum, including pelvis and uterus. Oral leiomyosarcomas arise commonly in the bones and the soft tissues and are reported mainly among adults, without any specific age or gender predilection. They have an aggressive growth pattern and a high rate of recurrence due to their dissemination in the bloodstream.[1,5-7] Clinically, LMS appears as a non-ulcerated painless mass with a well-defined structure.[1,8]

Histopathologically, leiomyosarcomas typically is presented with intersecting fascicles of spindle cells; furthermore, the immunohistochemical evaluation is often helpful to distinguish them from other tumors with similar light microscopic features. Due to their rarity, Leiomyosarcomas can be easily mistaken for other more common spindle cell lesions in the oral cavity.[4,9]

Several modalities are suggested for the treatment of LMS, including excision, radical surgery, radiotherapy, and chemotherapy. Wide surgical resection is the most preferable treatment protocol for LMS.[2,5,8]

In this article, we report a case of gingival LMS in a young woman.

Case Presentation

A 32-year-old woman was referred to the department of oral and maxillofacial medicine at Tehran University of Medical Sciences with the complaint of a mass at the right side of mandibular gingiva. The lesion was developed approximately 3 months earlier and was smaller at the time; however, the patient was visited by a general dentist approximately 2 months prior and the mass was excised without any histopathological examination. Unfortunately, the lesion recurred some time later.

The patient's medical history mentioned a nasolabial cyst in the right nasolabial fold (causing septal deviation and facial asymmetry), Bartholin's cyst surgery, and occasional diarrhea and constipation. Thus, she was referred to an internist for medical evaluation. It should be noted that she was receiving acetaminophen codeine for facial pain on the right side of the submandibular region at the time.

Oral cavity examination revealed a smooth, firm, reddish-pink, painless, homogenous, rapid growing gingival mass around the lingual side of teeth numbers 46 and 47, measuring about 2.5 × 2 × 1 cm, with an eroded surface (Figure 1. A, B). The mass was non-hemorrhagic and dome-shaped. There was a small nodule on the buccal side of the interdental area (about 0.5 × 0.5 cm). The second right mandibular molar (tooth number 47) had grade II mobility. The rest of the mouth was normal. The radiographic examination

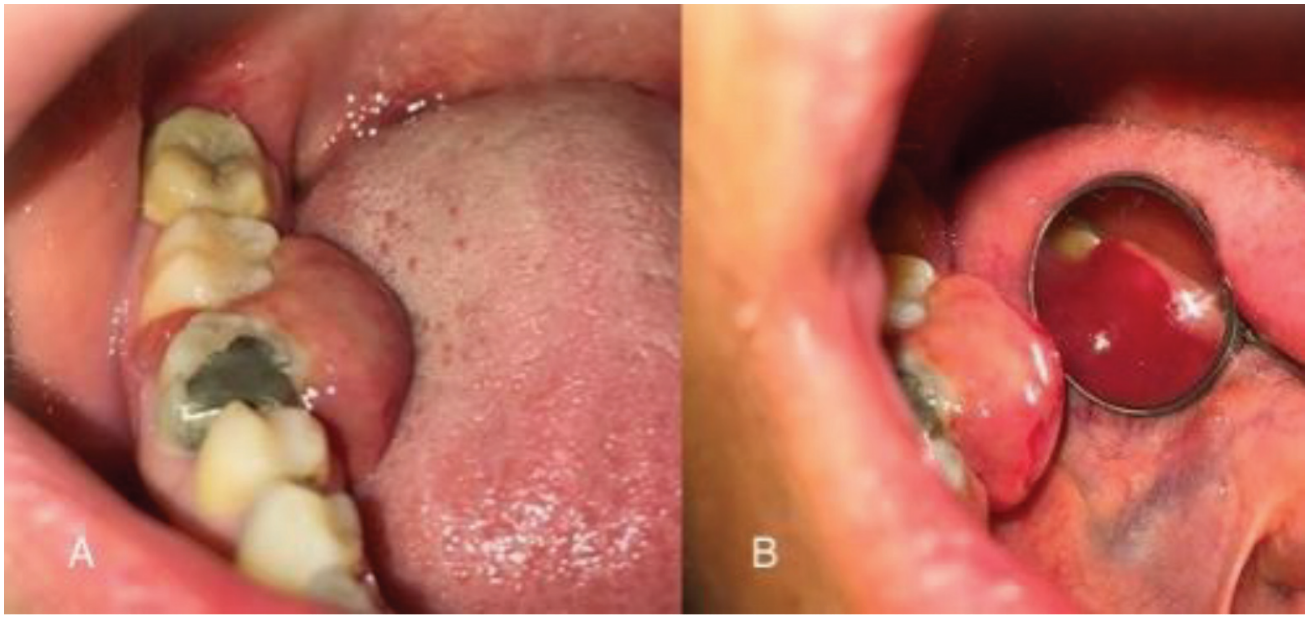

Figure 1. (A, B) Clinical examination showed a reddish-pink and homogenous gingival mass.

Table 1. Case reports of oral leiomyosarcoma

Author (year)	Age (years)	Gender	Location of the lesion	Clinical appearance	Radiographic view
Amarapala (2006)[2]	13	M	Tongue	A lump on lateral border	N/A
Amarapala (2006)[2]	17	F	Mandible	A buccal swelling, oval, soft, non-tender, anesthesia, mobility of teeth, tooth ache	Not reported
Amarapala (2006)[2]	23	M	Mandible	A swelling, pinkish, irregular, hard gradual enlargement	Not reported
Amarapala (2006)[2]	24	M	Maxilla	A swelling	Radiolucency n 678 region
Amarapala (2006)[2]	30	F	Maxillary buccal gingiva	A swelling, asymptomatic, slightly reddish, smooth surface, well-defined margins	Not reported
Rodini (2007)[13]	54	F	Mandibular gingiva	A painful pericoronal gingival swelling	An ill-defined area of resorption in the alveolar bone
Rodini (2007)[13]	63	M	Palate	A nonulcerated, painless, nontender soft palatal mass with elastic consistency	Homogeneous loss of trabecular architecture in form of a nondelineated radiolucent change (the appearance of a tooth "floating in space")
Crossman (2008)[12]	46	F	Tongue	A well-demarcated firm nodule, painful to palpate; the overlying mucosa was indurated and ulcerated	N/A
Kim (2009)[15]	56	F	Maxillary buccal gingiva	A painful swelling with a necrotic ulcer	An ill-defined radiolucency in the area and a slightly increase of radiopacity in the maxillary sinus
Fernandez-Barriales (2013)[10]	49	F	Mandible	Hemifacial swelling, trismus, and an alveolar, mucosal, ulcerated mass in the ipsilateral posterior mandible	A poorly circumscribed, osteolytic, mandibular ramus lesion
Patel (2013)[11]	23	M	Mandible (intraosseous	A moderately firm, vestibular swelling, mobility of teeth, pain	Lytic lesion displacing the root apices of teeth
Sandhu (2014)[1]	63	M	Maxilla	A bosselated growth, firm, painless, non-tender, patchy bluish discoloration,mobility of teeth	Not reported; however, the CT scan showed an osteolytic lesion extending in the nasal chamber
Rahrotaban (2015)[14]	73	M	Maxilla	An exophytic ulcerated mass	An ill-defined radiolucent lesion which seemed to invade the maxillary sinus
Viviano (2017)[5]	22	F	Mandibular lingual gingiva	a hard, non-painful, reddish-pink, non-haemorrhagic gingival swelling	Normal
Ko (2018)[4]	86	M	Buccal mucosa	A swelling with ulceration	N/A
Bayramoglu (2018)[8]	19	M	Maxilla	A well-demarcated, firm nodule without ulceration, slightly tender to palpation, not fixed to the adjacent tissue, mobility of teeth	A small amount of bone resorption

M: Male; F: Female; N/A: Not applicable

revealed underlying bone destruction (an interdental rarefaction between teeth numbers 46 and 47) and also PDL widening (Figure 2). The root of the first right mandibular molar (tooth number 46) was also treated. On extra-oral examination, no swollen lymph nodes were

detected in the cervical and submandibular regions.

Differential diagnoses based on the patient's history and clinical appearance of the lesion included reactive lesions (such as peripheral giant cell granuloma) and mesenchymal tumors. It should be noted that due to the non-specific clinical presentation, the diagnosis was more in favor of the reactive lesions. To further confirm this, an excisional biopsy was performed under local anesthesia along with the extraction of tooth number 47.

The sample was sent to a pathologist to establish the diagnosis. Hematoxylin and eosin (H&E) stained slides showed the neoplastic proliferation of spindle-shaped cells arranged in interlacing fascicles within a fibrous to myxoid background containing branching vessels. The tumor cells revealed nuclear polymorphism and increased mitotic figures. The overlying oral epithelium was ulcerated in some parts (Figure 3).

The tumor cells show diffuse positive staining for SMA (Figure 4. A) and weak scattered staining for S100 (Figure 4. B), CD 34 (Figure 4. C), and β-catenin (Figure 4. D). MIB-1 (Figure 4. E) labeling revealed a 25% positivity in tumor cells.

Histologic findings and immunohistochemistry (IHC) staining were in favor of LMS (grade one: since no lymphovascular invasion or necrosis was found and the mitotic rate was less than four in ten high-power fields). Neither necrosis nor the lymph-vascular invasion was identified.

Given this diagnosis, more extensive surgery seemed necessary. Hence, a preoperative spiral face computed tomography (CT) scan with contrast was done. The CT showed a 28 × 21mm mild enhancing oval soft tissue mass in the right nasal nostril, which caused scalloping of underlying maxillary bone without any evidence of invasion, right-sided septal deviation with spur formation, and bony structures that appeared unremarkable. Spiral chest and neck CT scan with and without contrast revealed no abnormalities. The patient was also referred to a gynecologist for possible metastatic uterine leiomyosarcoma,[10] which was negative.

Subsequently, segmental bone resection was performed under general anesthesia. The extraoral cutaneous incision in the neck area was done starting from the submental region to the right mastoid, 2 cm below the mandibular angle. The next step involved a horizontal incision on the platysma to reach the right submandibular gland, followed by the ligation of the facial vein and artery. Upon pushing the submandibular gland further down, the base of the mandible became reachable from the menton to sigmoid notch area. Thereafter, a soft tissue intraoral incision with a 1 cm margin was done extending from the mesial side of tooth 46 to the distal side of 48, followed by a monocortical bone cut of the buccal cortex

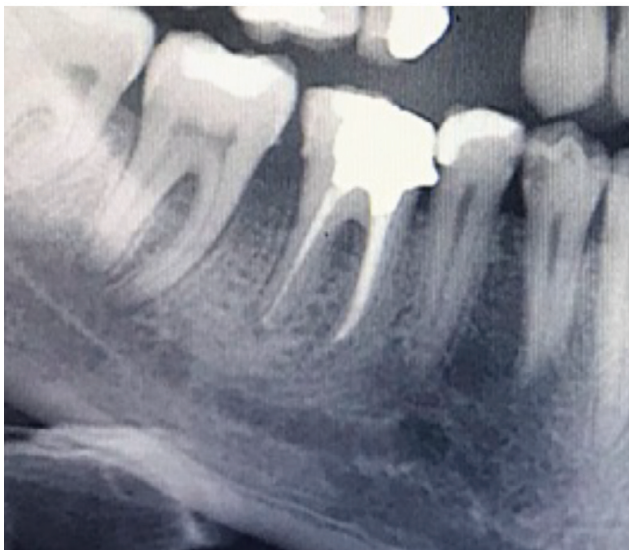

Figure 2. Radiographic examination showed an interdental rarefaction.

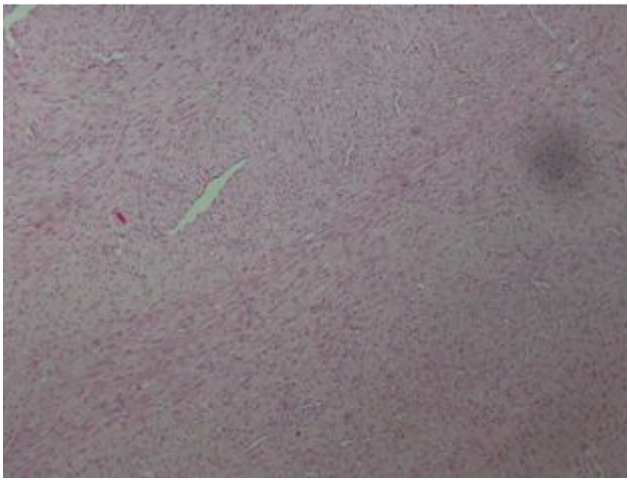

Figure 3. H&E staining revealed interlacing bundles of spindle-shaped cells with blunt-ended nuclei (×100).

from the distal side of tooth 45 to the distal side of tooth 48. After the insertion of a reconstruction plate, complete bicortical segmental resection of mandible in the anterior and posterior monocortical bone cuts areas were achieved; therefore, the pathologic lesion was entirely excised in one piece with a minimum margin of 1 cm (Figure 5. A, C). Following irrigation of

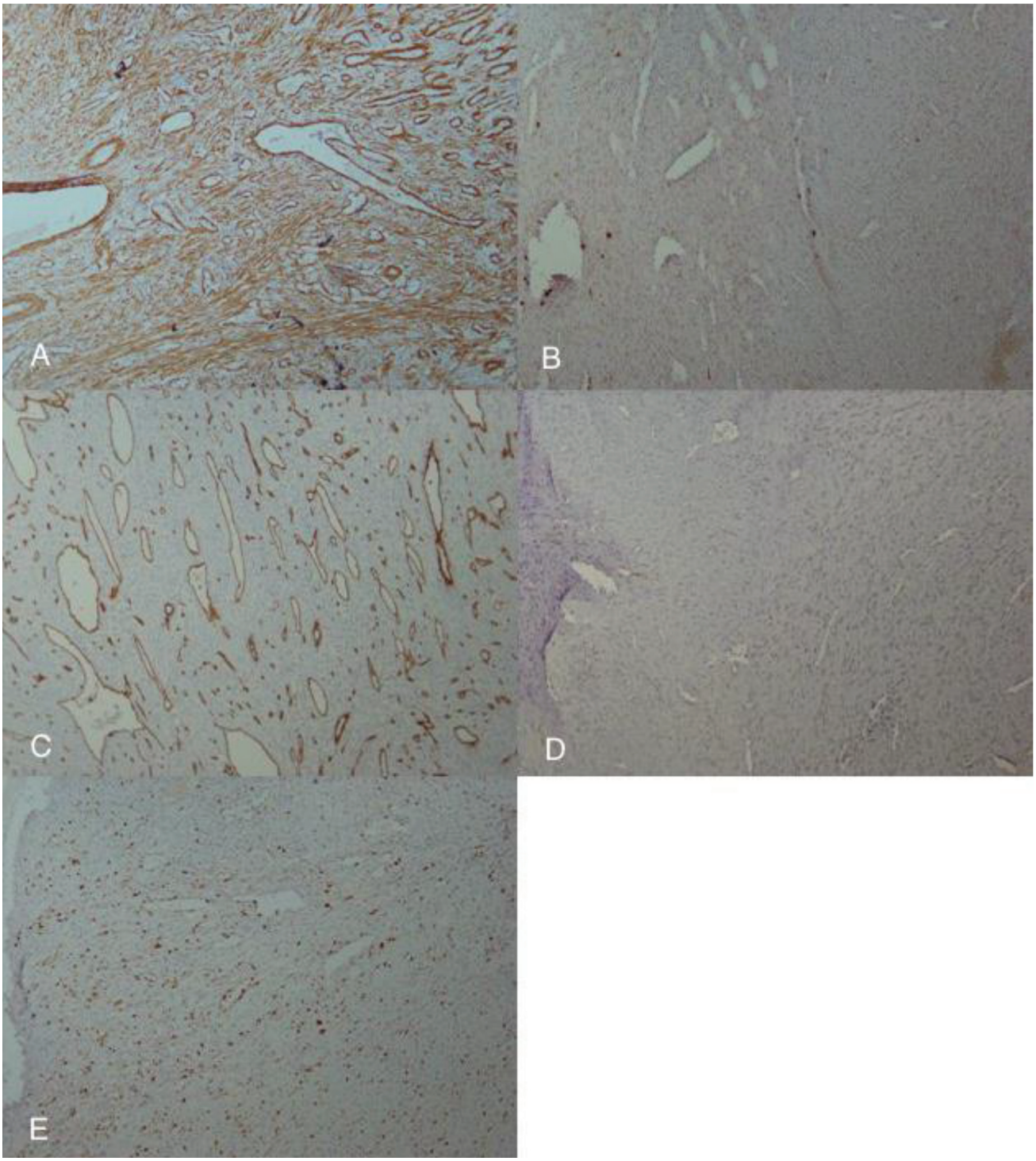

Figure 4. A. SMA staining demonstrates a positive cytoplasmic staining in almost all tumor cells (×100); B. S100 staining reveales a few scattered positive cells (×100); C. CD 34 immunoreactivity is not observed in tumor cells; note positive CD 34 staining in tumor vessels which serves as internal control (×100); D. IHC staining for β-catenin shows a few scattered positive cells (×100); E. IHC staining for MIB-1 labeling depicts more than 20% positive nuclear staining in hot spots (×100).
IHC: Immunohistochemistry

the area with normal saline solution, intraoral mucosa was initially sutured using a vicryl 3-0 suture. The neck incision was also sutured using a vicryl 3-0 suture. The extraoral cutaneous incision was finally sutured with a nylon 4-0 suture.

According to the preliminary pathology and official permanent reports, all margins were tumor-free; therefore, other treatment modalities were not applied.

Simultaneous surgery of the swelling of the nasolabial fold was performed and the result of the histopathological exam confirmed as a nasolabial cyst.

The patient was requested to monthly visit the surgeon and has a CT scan every 6 months.

Currently, ten months have passed since the radical surgery, with no signs of metastasis. However, the patient has some other problems such as a mild paresthesia in the area and the fracture of the mandibular reconstruction plate.

Discussion

LMS is an exceedingly uncommon neoplasm in the oral cavity which rarely occurs in the head and neck owing to the confined amount of smooth muscle in the region.[4] However, several cases of this neoplasm in the oral cavity have been reported, some of which are listed in table 1. Sandhu in 2014 reported an LMS of the maxilla in a 63-year-old male as a painless bosselated growth.[1] This patient's case comprises several differences compared with ours, since our patient was a 32-year-old female with a smooth mandibular mass.

Viviano in 2017 described an LMS in a 22-year-old woman. The lesion was an asymptomatic, reddish-pink, non-haemorrhagic swelling on the mandibular gingiva. The author stated that the radiograph of the patient's jaw was normal;[5] thus, it is different from the current study's case. However, features such as the women's young age and the clinical appearances and location of the

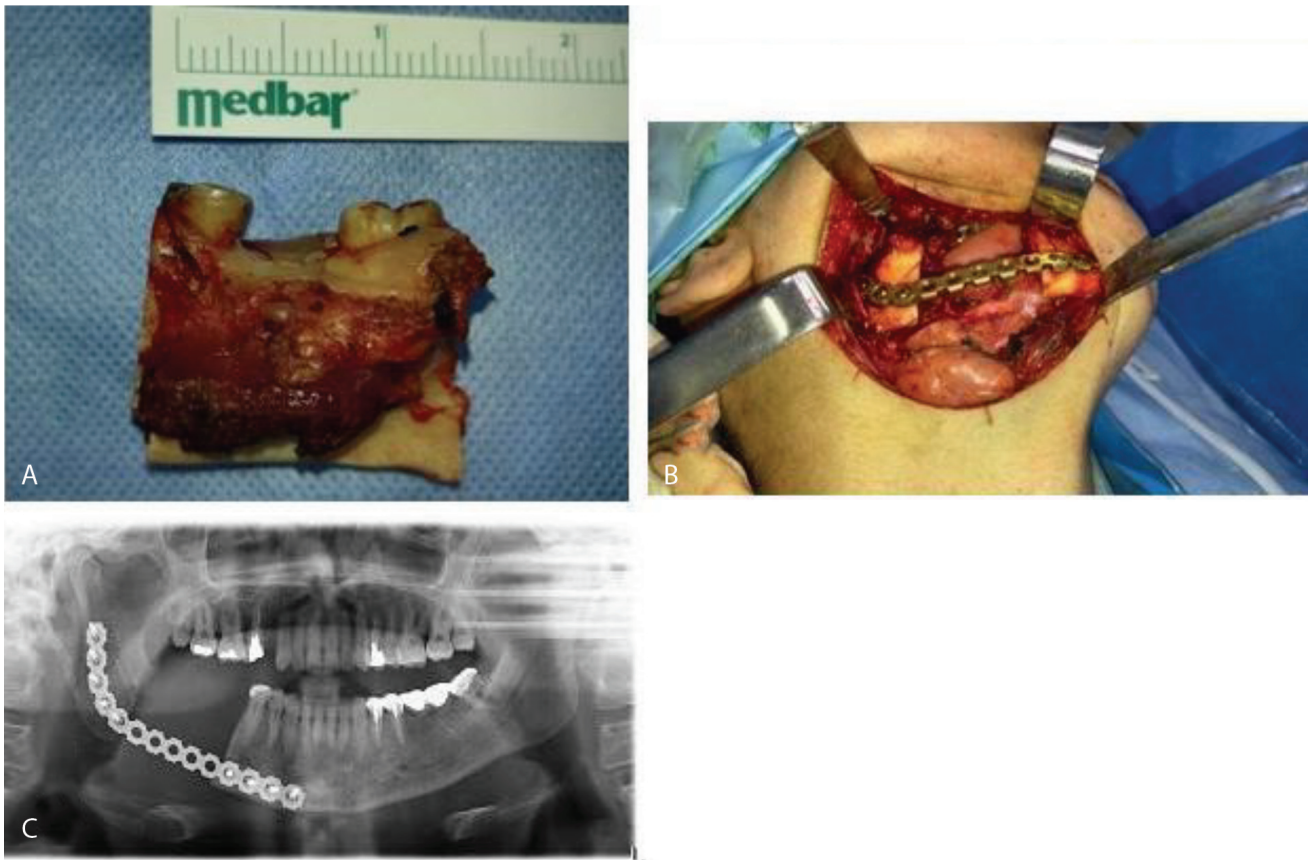

Figure 5. A. The pathologic lesion is entirely excised in one piece; B. Clinical view during bone resection; C. Radiographic view after bone resection.

lesions, indicate similarities between the two cases.

Ko in 2018 reported an oral LMS in an 86-year-old male with a swelling on his buccal mucosa.[4] This case is different from our patient in terms of gender, age, and the location of the lesion.

Bayramoðlu in 2018 reported an LMS case of the maxilla in a 19-year-old male. The lesion was a well-demarcated, firm, non-ulcerated nodule with a slight tenderness. It caused a small amount of bone resorption and tooth mobility.[8] The similarities between the cases are the clinical features of lesion and radiographic view; whereas, differences in terms of gender, age, and the location of the lesion are also visible.

Given that the LMS usually appears to be a painless mass, it is frequently mistaken for a more common lesion of the oral cavity.[4,5,8,11] Our patient's lesion was a painless, homogenous, and dome-shaped mass (however, she stated some pain on the right side of the submandibular region); and according to clinical appearance, we became more suspicious of the benign reactive lesions including peripheral giant cell granuloma.

Oral LMS can occur in the bone or the soft tissue, causing lytic lesions with ill-defined margins, periosteal elevation, calcification and cortical destruction.[1,5,6] Our patient's lesion was observed in the soft tissue (mandibular gingiva); however, radiographically showed interdental rarefaction between teeth numbers 46 and 47.

Although the precise aetiology of LMS is unknown, various cases relating to trauma, chemicals, estrogenic stimulation, ionizing radiation, and the Epstein-Barr virus have been reported in the literature.[5,8,12] However, herein, none of the etiologic factors appear to play a role in the occurrence of LMS.

The diagnosis of LMS may be challenging and should be established by IHC study. Moreover, decisive diagnosis of LMS depends on the light microscopic and immunohistological findings. According to the observation of spindle cells, the differential diagnosis should include rhabdomyosarcoma, spindle cell carcinoma, fibrosarcoma, myofibrosarcoma, malignant schwannoma, and so on.[8,13,14] In our patient, the histopathologic view revealed the neoplastic proliferation of spindle-shaped cells. Moreover, to confirm the diagnosis, a wide range of lesions had to be ruled out through IHC examination. Accordingly, we requested a panel of IHC markers including SMA, S100, CD34, β-catenin, and MIB-1. These investigations led to the final diagnosis.

The usual route of metastatic dissemination for LMS is the bloodstream to the lungs. LMS has higher metastatic potential than some other sarcomas including fibrosarcomas and liposarcomas. In oral LMS, metastasis to regional lymph nodes is relatively uncommon.[1,8,14,15] In our patient, fortunately, no metastasis has been observed during the 10 months after radical surgery.

The prognosis for LMS is usually poor due to its high metastasis and recurrence rate. Its prognosis is relevant to the site of involvement. In the head and neck region, anatomic structures confine a complete resection of tumor, so some difficulties in tumor management are present. Therefore, local recurrence is under expectance specially in the first two years of treatment.[8,14,16]

Excision, enucleation, curettage, radical surgery, radiation therapy, and chemotherapy have all been proposed as treatment modalities. Wide surgical excision is the most superior treatment protocol for LMS, although it is not always possible in the head and neck region.[2,5,8,11,13] In our patient, according to the initial pathologic findings following the bone resection, all the margins were tumor-free, so no other treatment modalities were used.

In our opinion, given that our patient's lesion was completely removed, the possibility of local recurrence is negligible; nevertheless, the patient follow-up will continue if she cooperates.

Informed Consent

The patient declared her informed consent to submit this report.

Conflicts of Interest

None declared.

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