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Malignant transformation of leiomyoma of the mandible into leiomyosarcoma: A case report

Masoud Fallahi Motlagh [1*], Yousef Janbaz [2], Zahra Mirzaei [3]

1. Azarbayjan Hospital, Urmia, Iran.

2. Oral & Maxillofacial Pathologist.

3. Department of Oral & Maxillofacial Disease, Medical Sciences of Urmia University, Urmia, Iran.

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**Corresponding author:*
Masoud Fallahi Motlagh

Azarbayjan Hospital, Urmia, Iran.

Tel: +98-44-32754981
Fax: +98-44-32754981
Email: m_fallahi_m@Yahoo.com

ABSTRACT

Leiomyosarcoma (LMS) is an uncommon malignant spindle cell tumor of the head and neck region. It is extremely rare in the oral cavity that arises from smooth muscle differentiation. It may arise as primary, radiation-associated, or metastatic tumor. The clinical appearance of these tumors can be deceptively benign and can be mistaken for non-malignant conditions. Here We present a case with atypical leiomyoma of the mandible in a 40-year- old man who referred with complaint of pain and swelling in his jaw. He underwent surgery and histology and immonohistochemestery studies confirmed the diagnosis. After 6 months recurrence occurred. Histologic examination confirmed leiomyosarcoma so he was managed with surgical excision followed by radiotherapy and chemotherapy without any recurrence or metastasis after 2 years of follow-up.

Keywords: Leiomyosarcoma; Mandible; Spindle cell tumor.

Introduction

Leiomyosarcoma is a malignant neoplasm of mesenchymal origin, which occurs commonly in the uterus and gastrointestinal tract of adults. Because the paucity of smooth muscle content, occurrence of this neoplasm in the oral cavity is extremely rare [1,2]. It accounts for only 4% of the head and neck sarcomas [3]. In the oral cavity, most of the cases are seen in the mandible, maxilla, tongue, cheek, hard and soft palate, floor of the mouth and lip [4].

Clinically, it is very aggressive, and the prognosis is poor [5]. Immunohistochemical assay for actin, desmin, HHF 35 and vimentin helps in confirming the diagnosis of leiomyosarcoma [6]. This paper discusses an unusual case of atypical leiomyoma of the oral cavity with transforming into leiomyosarcoma. Although it hasbeen reported in the utero [7] but we can't find the same report in the oral cavity.

Case report

A 40 y/o man refers to our clinic with swelling in the oral cavity with duration of six weeks associated with moderate pain. Hewasn't a tobacco smoker and past medi-

cal history wasn't significant. Extra oral examination was normal and no palpable lymph node was found in cervical examination. On intraoral examination a gingival bulging and proliferation was seen with normal overlying mucosa andwithout any ulceration (Fig. 1). The patientdidn't have trismus. His laboratory tests were normal.

The panoramic radiograph revealed a lucent, ill-defined lesion extending from left central incisor to molar region (Fig. 2). CT scan showed radiolucency that has destructed borders without obvious expansion (Fig. 3). He underwentan incisional biopsy under local anesthesia. Section showed a nonencapsulated soft tissue tumor that consists of spindle shape cell that formed herring bone pattern and interlacing fascicles. Surface epithelium demonstrates ulcerative parakeratinized stratified squamous epithelium with elongated reteridges. The cells demonstrate cigar-shape morphology with acidophilic cytoplasm, minimal nuclear atypia, eosinophilic nucleoli, low mitotic activity (5-7/10HPF) with occasional atypic-looking configuration. No coagulative necrosis was seen (Fig. 4). Immunohistochemestry (IHC) staining was positive for SMA, Vimentin and positive in 25-28% of neoplastic cells for k167 and faintly positivein some tumoral cells for Myogenin. It was negative for PCK and CD34. Final diagnosis based on Kempson criteria was atypical leiomyoma with low risk of recurrence (AL_LRR).

So the patient underwent complete excision of the tumor with safe margins. Histological examination of main specimens confirmed the diagnosis. 27_30% of neoplastic cells positive for K167, and this indicate the malignant potential of tumor so long term follow up is necessary. The patient came back after six months for follow up and he complained about pain. Cbct revealed destruction in mandible (Fig. 5). Growing of the tumor was fast and it grew up about 3x5cm in a few days. The patient was candidate for surgery and before starting surgery frozen section study revealed spindle cells and malignancy so the patient underwent resection of the mandible and neck dissection and overlying soft tissue with 1cm safe margin and reconstructed with reconstruction plate and Submental flap (Fig. 6).

Microscopic examination revealed a malignant mesenchymal tumor that was composed of spindle cell proliferation forming rough bundles and fascicles with interlacing pattern. Scattered abnormal mitotic figureswere also present (Fig. 7). Immounohistichemestry study for KI67 showed positive up to 50% of neoplastic cells and strongly positive for SMA and Vimentin (Fig. 8). Notumoral necrosis was observed. Then the patient went on chemotherapy in the local hospital, and two year follow-up showed no recurrence of tumor.

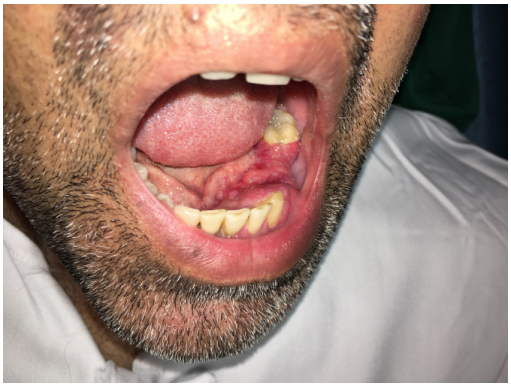


Fig 1. A proliferative lesion without any ulceration in the gingiva.

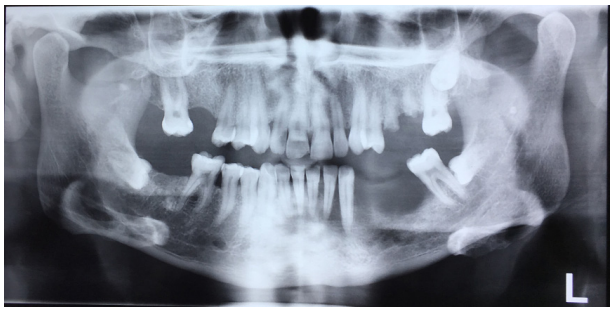


Fig 2. Panoramic radiograph revealed a radiolucent, ill- defined lesion extending from left central incisor to molar region.

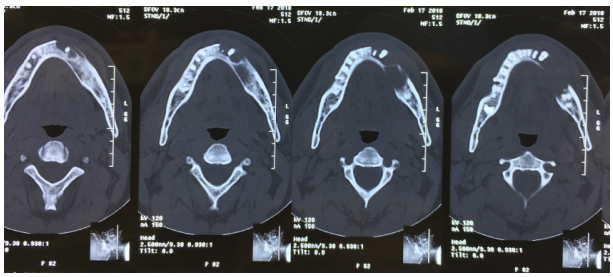


Fig 3. Ct scan shows destruction of the mandibular bone without any expansion.

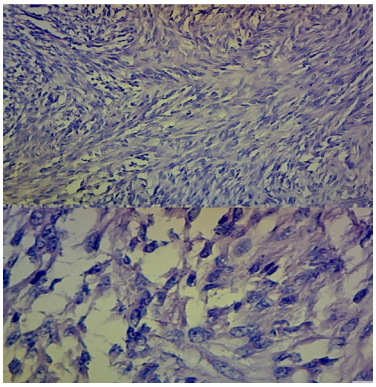


Fig 4. Microscopic view of atypical leiomyoma. Sections show a non-encapsulated soft tissue tumor that

consist of spindle shape cell that formed herring bone pattern and interlacing fascicles. The cells demonstrate cigar_shape morphology with acidophilic cytoplasm, nuclear atypia, eosinophilic nucleoli, high mitotic activity with occasional atypic-looking configuration.

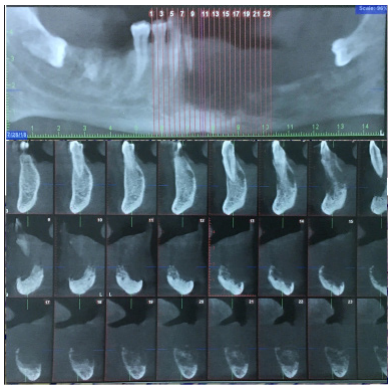
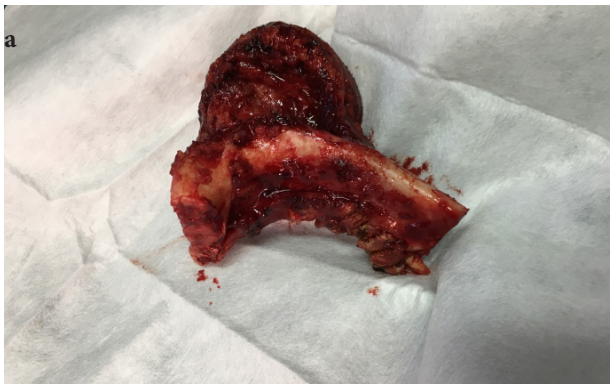


Fig 5. Cbct from mandibular bone shows recurrence has occurred in the left side.



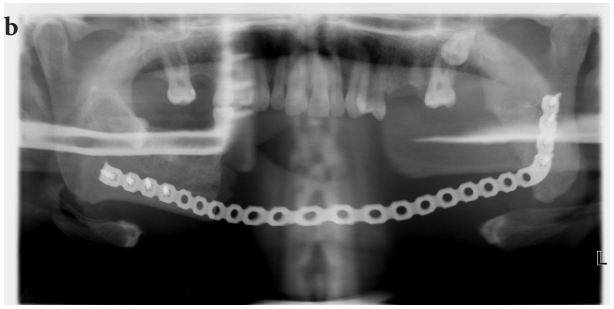


Fig 6. The tumor was resected (a) and reconstruction was done by reconstruction plate and submental flap (b,c).

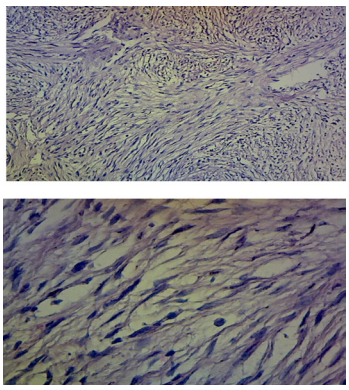
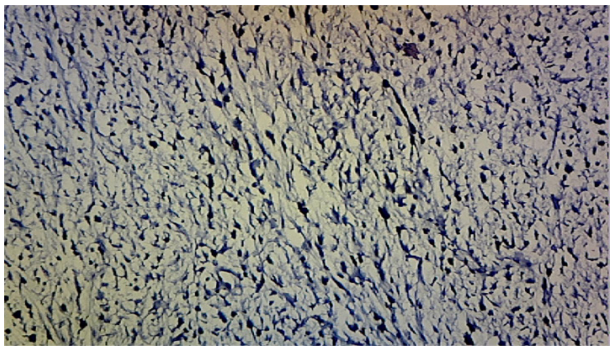
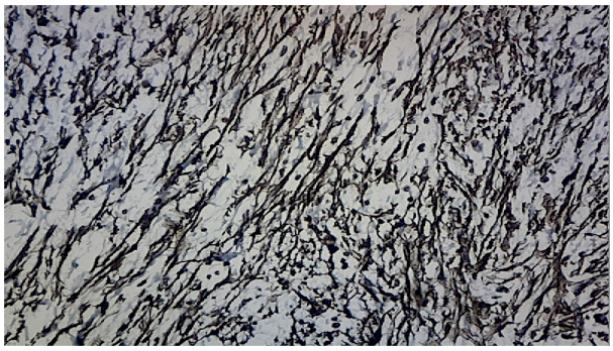


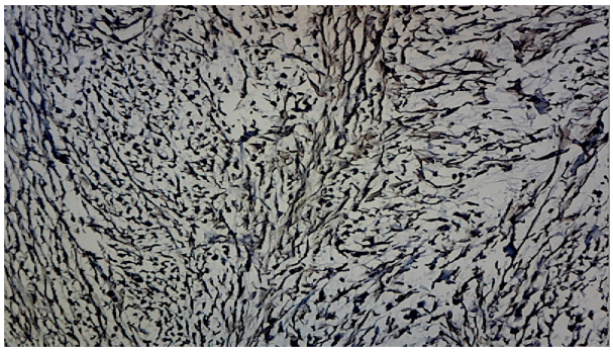
Fig 7. Microscopic examination revealed a malignant mesenchymal tumor that was composed of spindle cell proliferation forming rough bundles and fascicles with interlacing pattern. Scattered abnormal mitotic figures were also present.



KI67: Up to 50% of neoplastic cells are positive (a).



SMA: Strongly positive (b).



Vimentin: Strongly positive (c).

Fig 8. Immono histo chemistry staining.

Discussion

LMS is an uncommon malignant mesenchymal neoplasm originating from smooth muscle. It occurs frequently in the gastrointestinal tract and uterus but oral LMS is very rare. The rarity of this neoplasm in the oral cavity can be attributed to the paucity of smooth muscle at this site. Only 3–7% of LMS cases occur in head and neck region [8]. However, when present in the head and neck region, they are usually occurred in cheek, mandible, gingiva, maxilla, floor of the mouth, tongue, soft and hard palate mucosa [9,10]. The most frequent regions in the intraoral locations of these tumors are as follows: buccal mucosa and mandible [11,12]. In our case it occurred in the mandible and including the current case, only 9 cases have been reported in the mandible. Mandibular canal may also be the site of origin in some cases of mandibular involvement [13].

The possibility of a metastatic LMS to the oral cavity from other primary sites should always be considered [14]. There are several reports of metastatic oral LMS [15,16]. In the case reported here, the examination of other organs failed to reveal any abnormality. We therefore concluded that this lesion was primary oral LMS. Oral LMS tends to metastasize to cervical nodes, lung and liver, unlike the LMS in other soft tissues, which rarely has nodal involvement [17]. There is no age and sex predilection [11,18]. Youngest reported case is a 1 year old and the oldest is 88 years. Higher incidence is supposed to occur among the middle age or elderly [19,20].

The cause of LMS is still uncertain, although association with trauma, estrogen therapy, ionizing radiation, and Epstein–Barr virus has been documented in the literatures [1]. The incidence of sarcomas in patients operated for uterin leiomyomas is 0.23% [21]. eiomyosarcoma arising in leiomyoma in oral region have not been reported in the literature. Clinically, LMS often presents as a rapidly growing, painless, well circumscribed mass, adhered firmly to the surrounding tissues which sometimes may be ulcerated. in our case the patient had pain probably because location of the tumor nearby mental foramen [22]. Due to nonspecific clinical presentation, diagnosis of LMS is based primarily on pathologic criteria. Difficulty in the microscopic diagnosis of LMS, especially in its differentiation from leiomyoma, has been widely recognized [17]. Immunohistochemistry or electron microscopy must be carried out to achieve more specific differential diagnosis [23]. The histological criteria include the presence of pleomorphism, bizarre cell forms, pattern of interlacing bundles of smooth muscle cells, and high mitotic rate [24].

Although histologic features such as atypia, cellularity and necrosis are correlated with malignancy, the number of mitoses per high-power field (HPF) seems to represent the most reliable criterion of malignancy [14]. Immunohistochemical study of LMS was consistently positive for alpha smooth muscle actin, vimentin, desmin, and negative for S-100 protein and cytokeratins [25]. These histopathological features along with immunohistochemical profiles aid in differentiating LMS from other similar spindle cell malignancies like malignant fibrous histiocytoma, fibrosarcoma, etc., When LMS is identified, it is necessary to determine whether the lesion is primary or secondary. As a general rule, the likelihood of distant metastasis is related to histological grade and tumor size; the larger and higher grade lesions, the higher risk of metastasis [26]. Early wide surgical excision with radical neck dissection remains the mainstay of treatment. Adjuvant radiotherapy and chemotherapy have also been beneficial for treatment. Overall the prognosis of LMS is poor and hence early diagnosis is the key to the management [27]. Because it has a high rate of recurrence and metastasis, long term regular follow up is necessary [28].

Conclusion

This case showed transformation of the atypical leiomyoma to leiomyosarcoma so clinicians should be aware of this occurrence to prevent misdiagnosis althoughthis conclusion will need more study in the future.

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Conflict of Interest

There is no conflict of interest to declare.

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