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AN INTRAORAL LEIOMYOSARCOMA MIMICKING DENTAL INFECTION: A CASE REPORT

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ABSTRACT

The aim of this study is to evaluate and discuss an oral leiomyosarcoma case which was misdiagnosed as an odontogenic infection. Leiomyosarcoma is a malignant smooth-muscle tumor that accounts for 5–10% of all soft tissue sarcomas. It is extremely rare in the oral cavity and has a low survival. The occurrence of oral leiomyosarcoma is 3-10% of all leiomyosarcomas arising in head and neck. The tumor is usually encountered as a slow growing, firm and non-ulcerated painless mass. In our case after the biopsy and histological examination, hemimaxillectomy and adjuvant radiotherapy was our treatment of choice. Our survival was 2 years. Due to the clinical appearance, tumor can appear misleadingly as a benign lesion or an infection. Early and correct diagnosis is crucial to avoid metastasis and follow-up examinations are essential on account of recurrence possibility.

Key words: Intraoral, Leiomyosarcoma, Malign, Tumor

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ÖZET

Bu olgu raporunun amacı odontojenik enfeksiyon olarak yanlış teşhis edilen leiomyosarkom vakasının değerlendirilmesi ve tartışılmasıdır. Leiomyosarkom malign bir düz kas tümörü olup, tüm yumuşak doku sarkomları arasında görülme oranı %5-10'dur. Oral kavitede çok nadir olarak izlenir ve düşük hayatta kalma oranına sahiptir. Oral leiomyosarkomlar, baş ve boyun bölgesinde görülün leiomyosarkomların %3-10'unu oluşturur. Sıklıkla yavaş büyüyen, sıkı ve ülsere olmayan şişlik şeklinde izlenir. Olgumuzda biyopsi ve histolojik inceleme sonrası tedavi seçeneği olarak hemimaksillektomi ve destekleyici radyoterapi uygulanmıştır. Hayatta kalma oranımız 2 yıldır. Klinik görünümü nedeni ile bu tümör benign lezyonlar ve enfeksiyonlar ile karışabilmektedir. Metastazın önlenmesi için erken ve doğru tanı çok önem taşımaktadır ve lezyonun olası tekrarı açısından hastalar yakın takibe alınmalıdır.

Anahtar Kelimeler: İntraoral, Leiomyosarkom, Malign, Tümor

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INTRODUCTION

Leiomyosarcoma (LMS) is a malignant mesenchymal smooth muscle neoplasm that most commonly occur in the uterine myometrium, the gastrointestinal tract and the retroperitoneum.[1-3] They are 7% of all soft tissue sarcomas.[4] The occurrence of this highly malignant tumor in the head and neck region is extremely rare, 3%-10% of all LMS's, probably because of the lack of smooth muscle tissue in this region.[1-5] LMS of the head and neck region is considered to originate from the tunica media of blood vessels, the ductus lingualis, the circumvallate papillae or pluripotential mesenchymal cells which undergo differentiation during the course of neoplastic transformation.[2, 4, 6-9] The cause of the LMS remains unknown however there are reports in the literature associating it with trauma, estrogenic stimulation, ionizing irradiation and Epstein-Barr virus.[10-12] LMS may occur as a primary tumor, metastasis or a radiation-associated tumor.[3] It is associated with aggressive clinical behavior and low survival.[13] The prognosis is usually poor because of high recurrence and metastatic rates.[10,14,15] The most common sites of metastasis are the lungs, bone, brain and the lymph nodes.[16] The recommended treatment for LMS is primarily wide surgical excision with histologically proven tumor-free margins based on the cases and literature.[17-19] The aim of this article is to report a case of misdiagnosed intraoral LMS, mimicking dental infection located in the right posterior maxilla managed with wide surgical excision and postoperative adjuvant radiotherapy. We emphasize the importance of early diagnosis and careful clinical and radiological evaluation of the suspicious lesions.

CASE REPORT

A 45-years-old woman was referred to our clinic for a painless, firmly adherent expansive mass at her right palatal and alveolar mucosa that had appeared 3 months before. An endodontist suspected dental infection and the patient had undergone endodontic treatment for her affected teeth. She had no clinical improvement in her condition after the endodontic treatment. The clinical examination disclosed that the pink-reddish lesion expands both labially and palatally (Figure 1). Periapical radiograph of the affected area revealed bone resorption, loss of trabecules and lamina dura (Figure 2). Her CT scan revealed a vertical bone resorption with irregular border from her right maxillary 1st premolar to right 1st molar region (Figure 3). No pathologic lymph nodes were found and patient had no history of malignancy or radiotherapy. The patient had undergone excisional biopsy from the lesion under local anesthesia. Lesion was excised including the overlying mucosa and the affected bone was removed with curettage. Excessive bleeding was not observed. The wound site was primarily closed with buccal flap.

Histological examination at x20 magnification (HEx20) revealed a mesenchimal neoplasm infiltrating into the bone trabecules with irregular borders. With x400 magnification (Hex400) leiomyosarcoma with fusiform cells was observed (Figures 4 and 5). The mitotic activity was clear. Immunohistochemical analysis was performed for the definitive diagnosis of LMS (Figure 6). The patient underwent hemimaxillectomy and adjuvant radiotherapy due to the invasive spread of tumor.



Figure 1. Intraoral view of the lesion after the extraction of tooth number 14



Figure 2. Periapical radiograph after root canal treatment

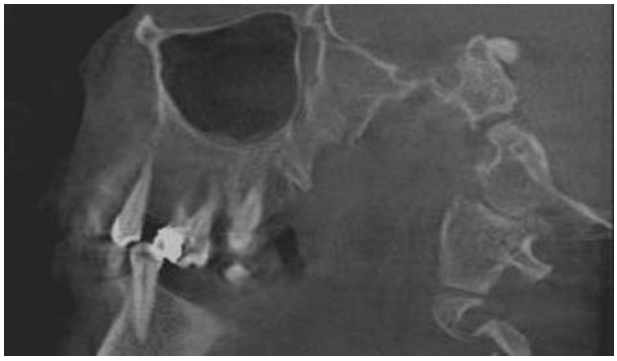


Figure 3. CT scan of the affected area

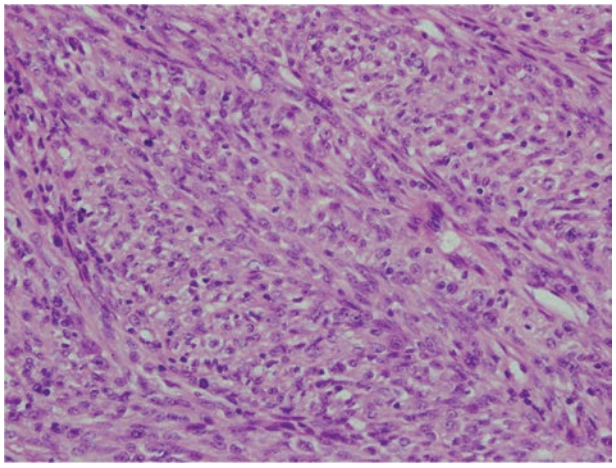


Figure 4. High-power view of the tumour demonstrating a rich cellularity and the spindle cell appearance of the cells (H&EX400).

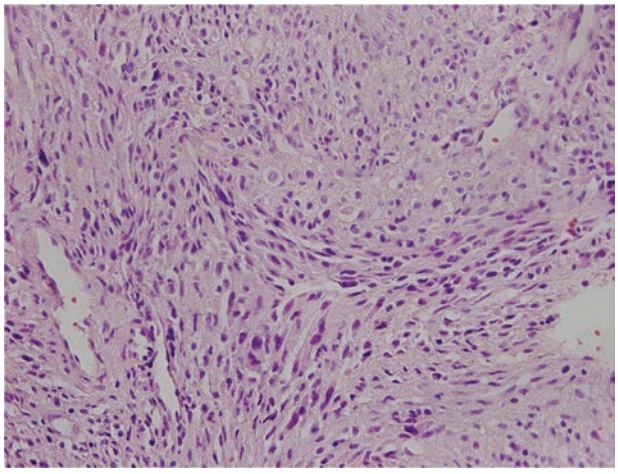


Figure 5. Illustrating the biphasic nature of the myofibroma with elongated spindle cells and more rounded cells with eosinophilic cytoplasm (H&EX400).

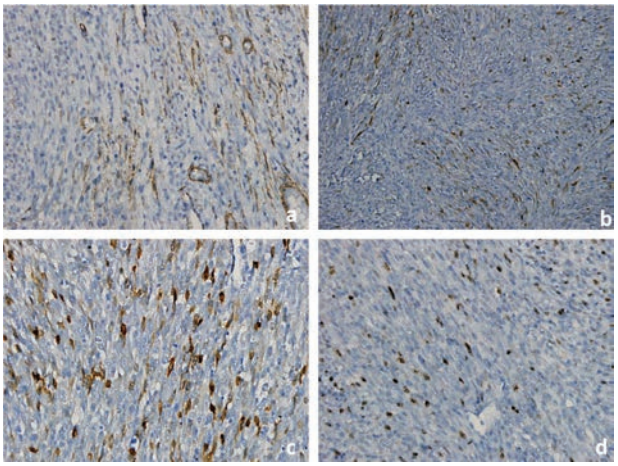


Figure 6(a). Demonstrating α-smooth muscle actin focal positivity within the tumour (immunoperoxidase-haematoxylinX400) (b). Demonstrating desmin focal positivity within the tumour (immunoperoxidase-haematoxylinX400) (c). Demonstrating S100 focal positivity within the tumour (immunoperoxidase-haematoxylinX400) (d). Demonstrating Ki-67 positivity(>%10) within the tumour (immunoperoxidase-haematoxylinX400)

The final diagnosis was primer LMS. In our case, there is no evidence of recurrence or metastasis at 12 months follow-up examination. However the patient died after 24 months.

DISCUSSION

LMS's of the head and neck region are quite rare with a ratio of 3-10% in total LMS's. Within this area the nose and paranasal sinuses (19%), skin and subcutaneous tissues (16%) and cervical esophagus (12%) are the most commonly affected sites.[20] LMS's occurring in the oral cavity is exceptionally rare. Maxilla and mandible are the most common involved sites (65%).[21] Larynx, hypopharynx, tongue, floor of the mouth, gingiva, soft palate, para pharyngeal space, lips and thyroid are the other intraoral sites of occurrence.

In a review Nagpal et al.[22] reported that among 63 patients reported in English literature among the past 20 years, mandible is the most frequent site with 20 cases. It was followed by maxilla with 15 cases. Our case has maxillary gingival, alveolar and palatal involvement. No involved lymph nodes or metastasis were detected.

LMS's occur over a wide age from 10 months to 91 years, with a mean age of 45 years.[21] Oral LMS does not display any significant gender predilection.[15]

LMS mostly presents as a well-circumscribed slow growing, discreet non-ulcerated painless mass that is firmly adherent

to the surrounding soft tissues, similar to our case.[10, 21] Pain and tenderness are unlikely features.[17] These lesions are easily misdiagnosed or go unnoticed due to the lack of definitive symptoms and the rarity of the disease. Also in our case this lesion was misdiagnosed as a dental infection and endodontic treatment was performed to the involved tooth, which was a waste of time for the patient. Clinical information, a thorough physical examination, imaging studies and a careful histopathological examination are extremely important for the correct diagnosis of LMS.[9] The presented case showed a painless pink-reddish mass mimicking dental infection at the maxilla expanding labially and palatally.

The treatment of LMS is wide and complete surgical excision to assure low local recurrence and longer survival.[2] Some authors also recommend adjuvant radiotherapy for high-grade sarcomas, large tumors and close or positive surgical margins.[23-26] Maxillary LMS's seem to have an unfavorable prognosis because of the greater difficulty in resecting maxillary lesions.[14,15,27,28] In our case a radical surgery was performed with hemimaxillectomy and adjuvant radiotherapy that may lead to a favorable prognosis.

Histologically LMS's are composed of bundles and fascicles of spindle-shaped cells with elongated, "cigar-shaped" nuclei and abundant eosinophylic cytoplasm.[28] The differential diagnosis of LMS may be challenging therefore the diagnosis should be supported by immunohistochemistry. Our diagnosis is also supported with immunuhistochemistry in our case.

The prognosis of LMS is usually poor. Ethunandan et al.[21] reported a local recurrence rate of 34% and metastasis rate of 35%. LMS arising from uterus, gastrointestinal tract and retroperitoneum can metastasize to the lungs, bone, brain and oral cavity and oral LMS tends to metastasize to the cervical nodes and lungs.[29,30] In another study Lo Muzio et al.[31] considered the prognosis as uncertain; reporting a local recurrence rate of 20.6% and distant metastasis rate of 20%. When LMS is diagnosed it is important to determine whether the lesion is primary or metastasis.[20] 2 to 5 years survival rate has been reported.[17] The survival of our case was 2 years. As the course of this disease is unpredictable and a high rate of recurrence, metastasis and mortality has been reported, long-term follow up of these patients is extremely important.[15,32]

Patel et al.[33] reported that there was no specific radiological finding for LMS; computed tomography and magnetic resonance imaging only reveal a soft tissue mass. Therefore clinical and radiological evaluation should be performed with caution for these lesions as there is always a risk of misdiagnose.

As a conclusion all the dental practitioners should be awake and aware for LMS's and other malignancies, which are easily misdiagnosed or go unnoticed; as time can be life saving for these patients.

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