

Primary Leiomyosarcoma in the Maxilla: A Case Report

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ABSTRACT

Primary leiomyosarcoma is a rare tumor in the head and neck. This report is to describe a case of maxillary leiomyosarcoma in a 36-year-old man, who was referred to Chamran Hospital (Shiraz, Iran) in September 2009. The patient was diagnosed with leiomyosarcoma, originated in the left premaxilla. Histogenetically, maxillary leiomyosarcoma arise from the medial muscle of blood vessels or from primitive mesenchyme in the maxilla (1). The lesion was treated with surgical resection; Midface Degloving procedure. The primary site received postoperative radiotherapy with external irradiation of 45 GY (25 treatments, 1/8 GY). The patient was monitored at follow-up visits in the next one year.

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Introduction

Leiomyosarcoma is a malignant neoplasm of smooth muscle origin and accounts for about 7% of all soft-tissue sarcomas [1]. It commonly occurs in the uterus, the gastrointestinal tract, or the retro peritoneum [2-3]. In rare occasions, leiomyosarcoma may occur in the oral cavity and leiomyosarcoma of the maxilla is extremely rare [1-2]. Diagnosis of these lesions, based on the routine histological diagnosis alone, can be challenging. Earlier reports were frequently submitted without any immunohistochemical assessments [2]. It is therefore not surprising that the experience of individual clinicians is not considerable and the procedures for the effective management of these rare lesions still remain unclear [2].

This report presents a rare case of maxillary leiomyosarcoma. To this end, a special emphasis has been given on the description, diagnosis and treatment of the disease.

Case Report

A 36-year-old man, suffering from left paranasal, premaxilla swelling and tooth pain, was referred to Chamran Hospital (Shiraz, Iran) in September 2009. Clinical examination of the patient face revealed the followings: a slight left paranasal swelling; a nose which was

slightly deviated to the right; and disappearing of the nasolabial fold (Figure 1).



Figure 1 Clinical examination of the patient showed that the lesion had caused swelling of the left side of the upper lip and lateral ala. It also revealed that the nasolabial fold was disappearing and the nose was slightly deviated.

The patient's general condition was not serious and the laboratory findings were not significant either. Oral examinations revealed a painful mass, extended from the alveolar process of the upper left second premolar to the upper right canine.

The mass was covered by normal mucosa. It was 35 mm in diameter, soft and tender. The upper left canine, the upper central and lateral incisors, and the first premolar adjacent to the mass were mobile.

Incisional biopsy revealed an extensive proliferation of neoplastic spindle cell sarcoma with smooth muscle differentiation. No regional lymph node enlargement was felt on palpation, and chest radiographs showed no abnormality.

Whole-body scanning with technetium-99m revealed an abnormal increase in the radiotracer activity in the mid left side of the maxilla both in the early and the final phases of scanning. No other abnormality was seen in the rest of the skeleton. The periapical radiograph of the anterior maxillary teeth revealed an ill-defined radiolucency of bone erosion and lamina dura resorption. Thus, the panoramic radiograph showed resorption of the maxilla extending up into the floor of the maxillary sinus (Figure 2).

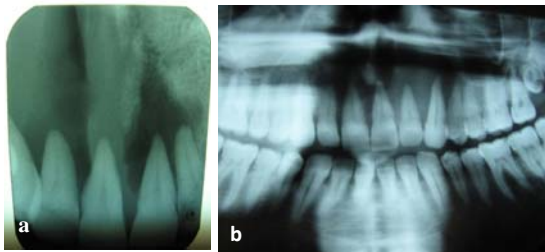


Figure 2a The periapical view of the anterior maxillary teeth revealed an ill-defined radiolucency of bone erosion and lamina dura resorption. **b** The panoramic view showed resorption of the maxilla extending into the bottom of the maxillary sinus

CT scan of the maxilla revealed a large mass in the left and midline of maxillary alveoli. It had caused bony destruction in the left maxillary alveoli and the anterior part of the hard palate in the left side. Bone erosion was also seen in the anterior medial wall of the left maxillary antrum (Figure 3). Distant metastasis work-up was negative. Microscopic examinations of the maxillary

lesion showed a neoplasm composed of spindle-shaped cells with a fascicular pattern. Most of the cells were blunt-ended. A moderate cellular pleomorphism was observed, nuclei were large, hyperchromatic and cigar-shaped. Mitotic figures could occasionally be identified (The average rate was 2 per 10 high-power fields). No necrosis was found (Figure 4 a). Immunohistochemistry (IHC) evaluation was carried out using the streptavidin-biotin method. The antibodies which were used included anti-vimentin, anti-desmin and anti-smooth muscle actin. All antibodies were from Dako (Dako NS, Glostrup, Denmark). Neoplastic cells expressed vimentin, desmin and smooth muscle actin and tumor cells were negative for s-100, CD 68, cytokeratin and HMB-45. These findings were consistent with the initial diagnosis of leiomyosarcoma (Figure 4-b).

Under general anesthesia, partial maxillectomy was performed via the Midfacial Degloving procedure (Figure 5). Maxillary resection included from the right first premolar to the left first molar, including the maxillary sinus, thus. Intraoperative frozen-sections diagnosis showed the resection margins free of any sign of tumor. Postoperative recovery was incomplete, since achieving a complete resection with wide margins in the head and neck region is not always possible. In order to reduce the probability of micrometastases, the primary site received postoperative external irradiation of 45 GY (25 treatments, 1/8 GY). The patient was then referred to a prosthodontist to restore edentulous area of the maxilla. Partial dentures were constructed after three month (Figure 6).

Discussion

Primary leiomyosarcoma is a rare neoplasm that can occur at any anatomic site [1]. Soft tissue sarcomas

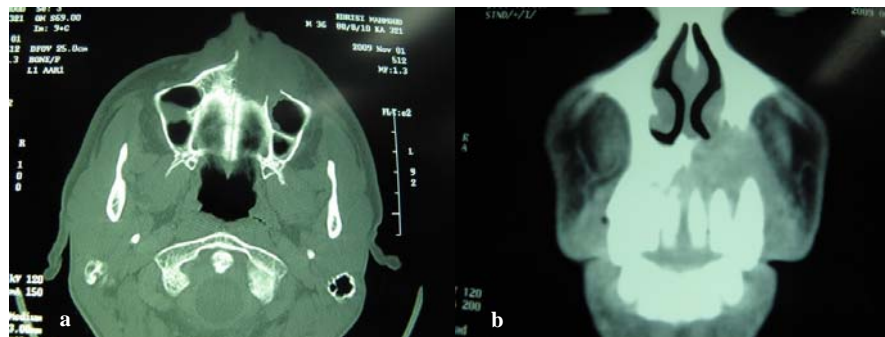


Figure 3a A view of Preoperative coronal CT of midface **b** A view of preoperative axial CT of midface, The process of bone destruction of the left maxillary alveolar and the anterior part of the hard palate with the left maxillary sinus extension

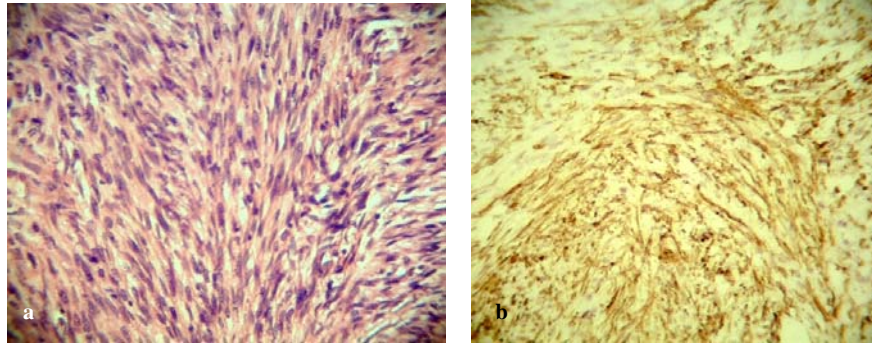


Figure 4 Microscopic examinations: **a** interlacing fascicles of blunt-ended spindle cells with cellular atypia and mitotic activity. **b** IHC staining showing reactivity of tumor cells for smooth muscle actin.

account for less than 1% of all tumors of the head and neck region [4-5]. Primary leiomyosarcoma (LMS) accounts for approximately 7% of all soft-tissue sarcomas. Primary leiomyosarcoma (LMS) of the head and neck region is extremely rare [4-5]. LMS is rare in the orofacial region and is the fourth most common sarcoma in the maxilla [2, 5].

Based on the previous studies, Oral LMS does not display any significant gender predilection and has been reported to occur over a wide range of age, with most patients in their third or sixth or seventh decade of life [2]. Jaw bones appear to be the preferred site for Oral LMS; approximately 70% of these tumors arise in the maxilla and mandible [6]. Other less frequent intraoral sites include the tongue, the soft and the hard palate, the floor of the mouth, the buccal mucosa, the gingiva and the upper lip [6]. In a different study, Chidzonga et al. reviewed the case notes of 88 patients, suffering from sarcomas of the oral and maxillofacial region. It was reported that LMS occurred mainly in the maxilla, with a five to one ratio of males to females [5]. LMS may occur in sites with scanty smooth muscle, such as the oral and maxillofacial region. In such a case, it arises from the tunica media of the blood vessels or from

pluripotential undifferentiated mesenchymal cells [6]. The definitive diagnosis of LMS in the jaw can be a challenging task because it shares some similarities with other sarcomas which are composed of spindle cells, such as fibrosarcoma, malignant fibrous histiocytoma, neurogenic sarcoma, rhabdomyosarcoma and malignant schwannoma [1, 6-7]. A definitive diagnosis of LMS is generally established through the use of a light microscope, but histochemical and immunohistochemical findings as well as the findings obtained through the use of an electron microscope are widely accepted as useful in confirming the diagnosis [1, 6-7]. In the present case, the diagnosis was made through biopsy. Immunoreactivity for smooth muscle actin, vimentin and desmin was positive but it was negative for cytokeratin, HMB-45, CD 68 and s-100. These findings as well as some others were in line with the previous findings about Oral LMS. Generally, LMS does not have definite signs and symptoms and pain and tenderness are not common [2, 8]. Commonly, it presents as a slowly enlarging, discrete, firm, non-ulcerated painless mass [2, 8]. Wide local field resection is the treatment of choice for Oral LMS. A review of literature on Oral LMS reveals that treatments have included wide-field surgical resection.

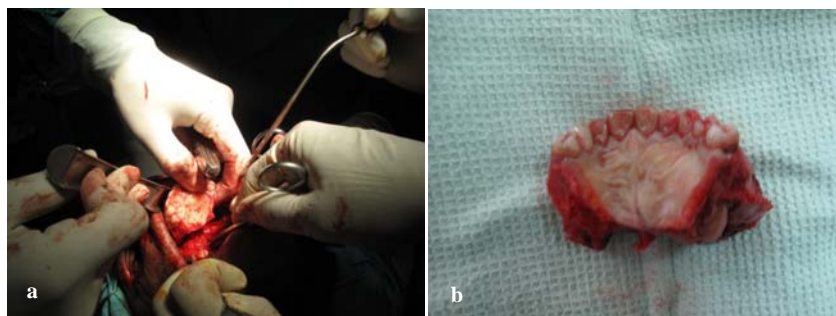


Figure 5a Partial maxillectomy was performed via a midfacial degloving procedure **b** The maxilla was resected

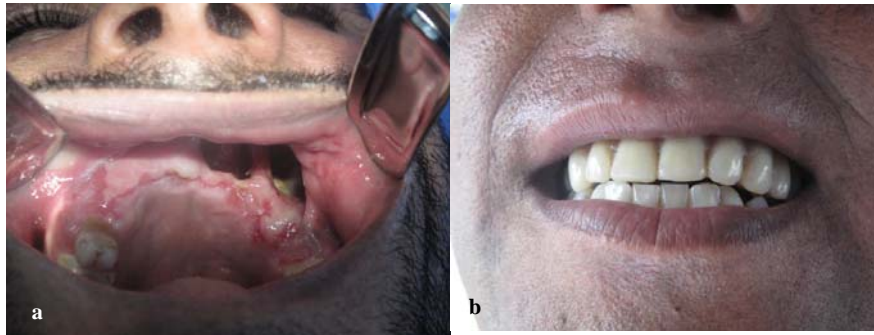


Figure 6 a Intra oral view of the patient eight weeks after the surgery b Extraoral view of the patient wearing partial dentures three month after the surgery

A few cases, however, have received radiotherapy, and chemotherapy has been used only as a follow-up treatment in patients with recurrence [1, 2, 4]. The prognosis of LMS is not usually good, and this is due high risk of recurrence and also a high metastatic rate [1, 9]. The reported local recurrence rate for primary Oral LMS is 34% [2]. The incidence rate of distant metastasis in primary oral LMS is 35% [2]. The lung has been reported as the most frequent site of metastasis [2].

The followings can be stated in summary: primary Oral LMS is a rare tumor in the oral cavity and is often mistaken for other more common neoplasms which arise in the mouth [10]. The findings of this study were consistent with those obtained through histological examinations. They are further confirmed with the following immunohistochemical results. Surgical excisions seem to be the preferred method of treatment. Local recurrence and metastasis are not uncommon, and the site of the mentioned tumor can be a predictor of metastasis [10]. Bone involvement and metastasis are associated with poor prognosis [10]. Finally, it should be mentioned that further studies and new and improved methods of identification can affect the management of LMS in the oral cavity in the future.

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