Leiomyosarcoma of the Tongue—A Case Report

Yang TL1, Chiang CP2, Kok SH1 and Kuo YS1

1Oral & Maxillofacial Surgery,
2Oral Pathology, National Taiwan University Hospital

Abstract

Leiomyosarcoma (LMS) is a relatively uncommon mesenchymal tumor that exhibits smooth muscle differentiation. Only 3% to 10% of LMS arise in the head and neck. Most LMS involving the oral tissues primarily affect the maxillary sinus, the maxilla or mandible. LMS of the tongue, like the present case, is a rare lesion. Clinically, they generally appear as a painless, occasionally ulcerated, slowly enlarging, subcutaneous or submucosal mass. The tumors are usually firmly fixed to surrounding tissues. Histologically, the tumor was composed of spindle-shaped cells with elongated, "cigar-shaped" nuclei and eosinophilic cytoplasm. We report a case of LMS involving the tongue of a 57-year-old female. Tracing back to her history, she visited an ENT Department of a local hospital due to a reddish, nodular lesion on the right lateral border of tongue, and the diagnosis was mucocele. Then she came to another hospital for an incisional biopsy, and the pathological report was fibrosarcoma. Finally, she was referred to our department for help. Intra-oral examination showed a 3×2.5 cm exophytic, indurated lesion of the right lateral border of tongue. Therefore, she received right supraomohyoid neck dissection and right partial glossectomy on 2002-06-30. The surgical margins were tumor-free and no lymph node metastasis was observed. Now the patient was kept on follow-up and no signs of local recurrence could be detected.

Key words: LMS (Leiomyosarcoma), Tongue, Sarcoma.

Introduction

LMS accounts for 6% to 7% of soft tissues sarcomas. Their main site of occurrence is in the uterus, gastrointestinal tract, and retroperitoneum. Oral LMS are extremely rare. They primarily affect the maxillary sinus, the maxillary or mandibular bone.
chemical features that were useful in arriving at the final diagnosis.

**Report of case**

A 57-year-old woman came to our Department on 2002-08-10 with a reddish lesion on the right lateral border of tongue. It occurred about 9 months ago and caused mild discomfort. Intra-oral examination showed a 3×2.5 cm lesion with induration on the right ventral and lateral border of tongue (Fig. 1). No regional lymph nodes were palpable. A biopsy was performed and the diagnosis was LMS of the tongue.

RT supramohoyid neck dissection was performed on 2002-08-30 under general anesthesia and RT partial glossectomy with 2 cm safety margin (Fig. 2 & 3). Histopathologic examination showed that the surgical margins were tumor-free and in 19 lymph nodes were no metastasis. In addition, the tumor was composed of variable oriented fascicles of spindle-shaped cells with "cigar-shaped" nuclei and eosinophilic cytoplasm, containing occasional PAS-positive granules (Fig. 4).

In addition to these findings, immunohistochemical studies were also performed for anti-vimentin, desmin (Fig. 5-A), smooth-muscle actin (Fig. 5-B) and S-100 proteins. The cytoplasm of the tumor cells was positive for all of these intermediate filaments except S-100 protein. These findings were consistent with the diagnosis of LMS.

The patient had been follow-up for 3 years and, so far, no local recurrence could be observed. The cervical and submandibular regions remain free of palpable lymph nodes. (Fig. 6)

**Discussion**

LMS is a malignant mesenchymal tumor that is relatively common in the uterus and gastrointestinal tract. However, LMS of the oral cavity is a very rare tumor, and this has been attributed to the paucity of smooth muscle in this area. Only 3% to 10% of LMS arise in the head and neck, and in this area the nose and paranasal sinuses (19%), skin and subcutaneous tissues (16%), and cervical esophagus (12%) are the most common sites. The jaw bones have been the most prevalent location for this tumor accounting for 45% of all reported cases in the oral region.

LMS are tumors of adult life, with a peak incidence in the 5-7 decades, and are more common in women than men. Clinically, they generally appear as a painless, occasionally ulcerated, slowly enlarging, discrete, subcutaneous or submucosal mass. Grossly, the tumors are often deceptively circumscribed or pseudocapsulated, smooth and firm, and gray-white to pink-tan. Larger lesions may show focal necrosis and hemorrhage. The tumors are usually firmly fixed to surrounding tissues.

Immunohistochemical staining and electron microscopy are used for the final diagnosis of oral LMS. Histological criteria for diagnosis of LMS require: 1) a pattern of interlacing bundles of smooth muscle cells, 2) a high mitotic rate, 3) cellular pleomorphism, and 4) bizarre cell forms. Myofibrils are also present in some cases but are not required for the diagnosis. These microscopic findings are only seen in cases of well-differentiated LMS. Masson's trichome staining and immunohistochemical evaluation for muscle antigen separates of LMS from other sarcomas. Positive reactions for desmin, vimentin, and smooth muscle actin, nonreactive to S-100 protein and the cytokeratins have been demonstrated in LMS. The present case presented fulfilled these criteria and confirmed the diagnosis of LMS.

Unfortunately, the prognosis of this tumor cannot be predicted on either a clinical or histo-
Fig. 1. An exophytic, indurated lesion of the right ventral and lateral border of tongue. A. lateral border. B. ventral surface.

Fig. 2. R's supraomohyoid neck dissection. A. Neck incision line. B. Post-neck dissection view.
Fig. 3. Main tumor and neck specimen (5 x 2.5 cm)

Fig. 4. Microscopic view of specimen. The tumor was composed of variable oriented fascicles of spindle-shaped cells with "cigar-shaped" nuclei and eosinophilic cytoplasm, containing occasional PAS-positive granules. (H&E stain, original magnification, 400x)
Fig. 5. Microscopic view of specimen.
   A. The cytoplasm of the tumor cells was positive for Desmin(+) (Immunohistochemical stain, original magnification, 100x)
   B. The cytoplasm of the tumor cells was positive for smooth-muscle actin(+) (Immunohistochemical stain, original magnification, 100x)

Fig. 6. Three years after surgery. A. The right cervical and submandibular regions remain free of palpable lymph nodes. B. No evidence of local recurrence.
logical basis. Local recurrence and cervical and lung metastasis have been reported in nearly 40% of the cases. The high rate of recurrence shows that this tumor despite its well circumscribed appearance, has a tendency to infiltrate the surrounding tissues. Besides, LMS do not seem to benefit of radiochemotherapy. Therefore, wide surgical excision with histologically-proven tumor-free margins with regional lymph node dissection seems the most appropriate treatment that may guarantees prolonged survival. Follow-up for at least 5 years is strongly advised.

**Conclusion**

Intra-oral LMS is aggressive, and exceptionally rare. The diagnosis of LMS depends on accurate morphological and immunohistochemical characterization. The prognosis of intra-oral LMS is highly variable and seems to largely depend on the possibility of curative radical excision of the tumor. We reported a case of LMS of the tongue and the patient had undergone supramaxillary neck dissection and partial glossectomy. Postoperatively, her course was unremarkable. No signs of local recurrence were evident.

**References**

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舌之平滑肌肉瘤

楊子立、江俊斌、郭生興、郭雄飛
1台大醫院牙科部口腔及顱面外科
2口腔病理科

摘 要

平滑肌肉瘤(leiomyosarcoma)是由平滑肌分化衍生物來，是一種非常稀少的間葉性腫瘤，其中約只有3%~10%發生在頭頸部。在口腔癌組織當中，主要侵犯的是上頜骨和上、下頜骨，頜脂如本例發生在舌頭的病患是非常少見。在臨床上，平滑肌肉瘤是一無痛性，緩慢長大的皮下或粘膜下腫瘤，好發於50~70歲女性。腫瘤的顏色以灰白色和粉红色為主，通常會與鄰近組織緊密結合在一起。在顯微鏡下，此腫瘤是由多種不同的纖維細胞所構成，此細胞並含有雪茄狀的細胞核與嗜伊紅性的細胞質。本病例為54歲女性患者，因舌右側有一紅色病變而到某醫院耳鼻喉科求診，經診斷為粘液囊腫。病患又至另一醫院接受切片檢查，病理報告為纖維肉瘤；並於2002年8月10日至本院求診。口內發現在舌頭右側有一3×2.5 cm的外突性腫瘤，於是在同年8月30日接受廣泛腫瘤切除術及右側舌骨上頜部清創術，並於同年9月9日出院，術後病理報告為平滑肌肉瘤，並無頜部淋巴轉移，目前患者定期回診，觀察3年尚無復發現象。

關鍵語：平滑肌肉瘤，舌，肉瘤。

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Reprint requests to Dr. Ying-shiung Kuo, Oral & Maxillofacial Surgery, Department of Dentistry, National Taiwan University Hospital