Primary Maxillary Leiomyosarcoma—A Case Report

Yu-Wei Chiu¹, Ho-Tai Wu¹, Wing-Yin Li², Man-Tin Lui¹, Shou-Yen Kao¹, Wen-Liang Lo¹

¹Section of Oral and Maxillofacial Surgery, Department of Stomatology,
²Department of Laboratory Medicine
Taipei Veterans General Hospital, Taipei, Taiwan, R.O.C.

Abstract

We present a rare case of primary oral leiomyosarcoma (LMS) occurring at maxilla pterygoid plate area. The patient had symptom of pain and swelling for more than 10 months over left pre-auricular area. The patient went to see the neurologist for help and was diagnosed as trigeminal neuralgia after checking computed tomography (CT) without any finding. Medication was prescribed at first but without any regressive change. Then, the magnetic resonance imaging (MRI) was arranged and a tumor over left maxillary pterygoid area extending to temporal space was found. The biopsy was arranged under general anesthesia and the result showed compatible with sarcoma. The patient was treated by radical surgery and reconstruction with free anterior lateral thigh flap. The pathology showed leiomyosarcoma and the immunohistochemical examination revealed positive cytologic features of HHF-35, smooth muscle actin and h-Caldesmon. The distant metastasis to thoracic and lumbar spines were found after 6 months of surgery and the patient is still under palliative chemo and radiotherapy.

Key words: Maxilla, Leiomyosarcoma.

Introduction

Soft tissue sarcoma of head and neck region are rare and only about 5% of all soft tissue sarcoma happened on this region¹. The leiomyosarcoma (LMS) is a malignant neoplasm of smooth muscle origin. The uterine wall and gastrointestinal tract is the relatively common site. LMS in oral region is very rare and only 0.064% of 7748 smooth muscle tumors happened in this region²³. Oral LMSs have been reported to occur over a wide age range from 10 months to 91 years and with male predilection⁴⁻⁷. In our acknowledgement, there have been about 65 cases reported in the English literature since 1950 and often as individual case studies. The diagnosis of LMS is difficult on routine histology along except immunohistochemistry stain. We present a case of LMS happened on maxillary pterygoid region extending to the temporal space.

Case report

The 58-year-old male has systemic disease
of hypertension under regular medical control and had habit of smoking more than 30 years. He has presented at our Oral and Maxillofacial section complaining swelling and pain especially when chewing, swallowing and brushing teeth at left peri-auricular and temporal region. The history could be traced back to 10 months ago when he first felt a swelling and mild pain on the left temporal area. He went to local medical clinic first and the antibiotic was prescribed. The symptom subsided temporarily. However, it recurred after few weeks. Then, he went to the Neurological department in our hospital for help and the brain computed tomography (CT) was checked. There was no significant finding of CT and the trigeminal neuralgia was initially diagnosed. The medication of Trileptal and Rivotril were prescribed. The symptom relieved temporarily and exaggerated later. The image study of head magnetic resonance imaging (MRI) was arranged and the result showed a 4.8 × 3.5 × 4.7 cm sized mass lesion with partial necrosis and heterogeneous contrast enhancement over deep part of left masticator space with left mandibular ramus destruction more on posterior part extending to condylar process and deep part of left parotid gland. The image impression is malignant peripheral nerve sheath tumor or other mesenchymal tumor. (Fig. 1).

He was referred to our department for further treatment. The extraoral examination revealed severe swelling over left facial area. The facial skin was intact with a 3 × 4 cm subdermal indurated mass without skin adhesion and no clinical palpable neck lymphadenopathy. The maximal mouth opening was 3.5 cm. The intraoral examination revealed intact and normal consistency of left buccal mucosa. He received incisional biopsy under general anesthesia and the pathology revealed compatible with sarcoma. The positron emission tomography (PET) was arranged and showed hypermetabolic mass located in the left maxilla with extension to level Ib cervical region. The patient was then received a wide excision of the tumor with hemimandibulectomy, partial maxillectomy and left side supraomohyoid neck lymph node dissection (Fig. 2). The surgical defect was repaired with free anterolateral thigh flap. The pathologic examination of the tumor from radical surgery was diagnosed as leiomyosarcoma, high grade composed of fascicules of spindle cell with pleomorphic nuclei and frequent mitoses (14/10 HPFs). Multinucleated tumor giant cells are also seen (Fig. 3). The spindle cells are diffusing reactive for HHF-35, smooth muscle actin and h-Caldesmon (Fig. 4). The tumor is unencapsulated and grew in infiltrative pattern with mandible and parotid gland invasion. The surgery resection margin is free. He was followed for 6 months after surgery but the distant metastasis to thoracic and lumbar spines were found. The patient is under palliative radiotherapy.

Discussion

Leiomyosarcoma (LMS) is relative common on retroperitoneal somatic soft tissue, superficial cutaneous origin and large blood vessels. LMS is rare in oral cavity due to general absence of smooth muscle in the region except in the blood vessels, circumvallate papillae of the tongue and the occasional primitive mesenchymal tissue. The tumor of intra-oral region are most commonly located on maxilla followed by the mandible, tongue, cheek and floor of mouth. In clinical symptom, the tumors most frequently presented as a painless mass. Only 6% cases of LMS were
Fig. 1. The face MRI revealed tumor invasion at deep part of left masticatory space, mandibular ramus and deep part of left parotid glad.
Fig. 2. The patient received hemimandibulectomy, partial maxillectomy and left side supraomohyoid neck lymph node dissection. The left parotid gland and facial nerve were all sacrificed due to tumor adhesion.

Fig. 3. The tumor composed of fascicles of spindle cell with pleomorphic hyperchromatic nuclei, (H&E, 100X).
Fig. 4. Alpha-smooth muscle actin (a) and HHF-35 (b) immunoreactivity were evident in most tumor cells (200X).
painful and tender\(^9\). The lack of any distinguishing clinical features and the rarity of these lesions often result in being mistaken for other common lesions in oral cavity. In our case, he was diagnosed as trigeminal neuralgia at first, and the proper treatment time had been postponed.

The correct diagnosis is made only following definitive histological examination. The clinically pathologic feature of primary oral LMS appears microscopically as spindle cell neoplastic proliferations. The tumor cells are arranged in an interlacing fascicular pattern and contain oval to elongated, blunt-ended (cigar-shaped) nuclei. The nuclei may show hyperchromatism and varying degrees of atypia and pleomorphism. Based on the morphology alone, several different tumors may be difficult to differentiate. These include spindle cell carcinoma, angiosarcoma, malignant peripheral nerve sheath tumor, malignant melanoma, rhabdomyosarcoma, anaplastic lymphoma and solitary fibrous tumor. Immunohistochemical (IHC) and molecular markers may be useful to assess the biologic behavior of smooth muscle tumors. The review of IHC marker showed react positively with antibodies targeted against smooth muscle antigenic epitopes of mesenchymal origin, such as smooth muscle actin, muscle specific actin (HHF 35), vimentin and CD 146. A negative reaction is the application of antibodies target against epithelial antigenic epitopes, such as cytokeratins and EMA. A negative reaction is also negative with the use of antibodies against S-100 protein (neurogenic sarcomas), HMB-45 (melanoma) and angiosarcoma antigens (CD31, CD34) and factor VIII–related antigen. The presence of desmin may be either positive or negative\(^{10,11}\).

The treatment option is not complicated. The complete surgical excision is the most commonly treatment option and offer best outcome due to totally remove the tumor. However, surgery may be difficult when tumor involving infratemporal fossa, pterygoid plates, maxillary sinus or mandibular condyle. These cases may account for poor prognosis and high mortality rate detected in current review\(^4,11-13\). Local recurrence rate for primary oral LMS is 34%\(^6\). It is lower than soft tissue origin (50%)\(^{14}\), retroperitoneum, and blood vessels (50%)\(^{15}\). The frequency of distant metastasis is 35% and occurred most frequently in the lung\(^6\). Regional lymph nodes metastasis is frequently more than 15%\(^{13}\). The metastatic rate was significant more often originating from floor of the mouth and maxilla/mandible than buccal mucosa and tongue. The larger tumor size may increase metastatic incidence, but no statistically significance\(^8\).

Adjuvant radiotherapy can be used postoperatively, but this may not influence recurrence free or overall survival rate\(^{12,14,16,17}\). The chemotherapy appears ineffective and should be reserved for palliative treatment of patient with distant metastasis or inoperable tumor\(^{12,14,18}\). The 5-year survival was 55% – 62% and tumor with bony involvement and metastasis were associated with poor prognosis. The resection on microscopically tumor-free margins is an important factor for long-term survival in the cases of primary LMS in oral cavity\(^{11}\). Other correlated poor prognostic factors might include male gender and old age, but there were no statistically significant differences\(^6,11\).

In summary, primary oral LMS is a rare tumor in the oral cavity and is often ignored due to lack of clinical symptom and signs. The histological examination and following immunohistochemical confirmation must be used for diagnosis. If the tumor is resectable, surgical
excision seems to be the preferred method of treatment. Local recurrence and metastasis were not uncommon. Bony involvement and metastasis were associated with poor prognosis. Chemotherapy and radiotherapy can be used for metastasis or palliative treatment. No significant evidence showed the prognosis related to post-operative chemotherapy or radiotherapy.

Reference


原發性上顎平滑肌肉瘤—病例報告

邱昱瑋1 史和泰1 李永賢2 雷文天1 高壽延1 羅文良1

1台北榮民總醫院口腔醫學部，口腔顱面外科
2台北榮民總醫院病理檢驗部

摘 要

我們提出一個罕見的病例，發生於上顎翼板之原發性上顎平滑肌肉瘤。病患十個月前在顳區出現疼痛和腫脹之症狀，病患首先至神經內科就診，電腦斷層攝影顯示無任何異狀，因此被診斷為三叉神經痛。病患起初接受藥物治療，但症狀並無任何緩解。之後，經過神經內科醫師再安排核磁共振檢查，發現在上顒骨翼板延伸至顳窩區有腫瘤生成。病患被轉診至本科，經過全身麻醉下之切片檢查後證實為肉瘤。病人其後接受廣泛性切除手術及前大腿外側游離皮瓣重建。手術標本免疫組織學染色顯示HHF-35, smooth muscle actin及h-Caldesmon皆為陽性反應，病理報告為平滑肌肉瘤。病患於手術後6個月發現遠處轉移至胸椎及腰椎，目前正在接受緩和性放射線治療。

關鍵詞: 上顒, 平滑肌肉瘤。