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LEIOMYOSARCOMA OF THE MAXILLARY GINGIVA: A CASE REPORT

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ABSTRACT

An unusual case of leiomyosarcoma (LMS) of the maxillary gingiva is discussed here; this case presents a unique pattern of tumor growth and a long period between initial discovery and correct pathological diagnosis.

The tumor was incompletely resected twice by a private dentist over a period of 3 years, with a clinical diagnosis of epulis, no pathological examination was conducted during this period.

When it was finally removed, the tumor was very large (50 × 35 × 12 mm in size and 18 g in weight), consisting of an easily hemorrhagic mass originating in the gingival mucosa with the growth pattern of a polyp.

Following an extensive surgical excision and a unilateral radical neck dissection, the patient has been free of LMS for 8 years.

In light of this case, we strongly emphasize the importance of conducting a pathological examination, even though clinical examination seems to indicate a diagnosis of epulis or granulation. In this way, the presence of LMS can be ascertained in a timely manner with better prognosis for treatment and recovery.

Key words: Leiomyosarcoma, Diagnosis, Prognosis

INTRODUCTION

LMS is a common mesenchymal neoplasm that occurs most frequently in the uterus and gastrointestinal tract. The occurrence of primary LMS in the oral soft tissues or jawbones is very unusual.[1] Only 31 cases have been reported in the literature.[2] Among cases of oral soft tissue LMS, six arose in the cheek, five in the gingiva, five in the tongue and the remainder were located in the floor of the mouth, soft palate, hard palate, mandibular alveolar mucosa, posterior maxillary soft tissue and maxillary sinus. Among the jawbone lesion cases, six occurred in the mandible and four in the maxilla.

This paper discusses an unusual case of primary LMS of the oral cavity, which arose in the maxillary gingiva and had the growth pattern of a polyp.

REPORT OF CASE

A 17-year-old girl was referred to our hospital in October 1986 by a family dentist for further evaluation and treatment of a gingival mass involving the maxillary left premolar area. Three years before the development of this mass, the patient experienced gingival sensitivity in the same region. She saw her family dentist, who performed an excision of the mass and an extraction of the left second premolar tooth with a clinical diagnosis of epulis. No tissue was submitted for microscopic evaluation. Following the excision, the patient noted that the swelling

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had gradually recurred in the maxillary left premolar region, and often bled spontaneously. She saw the same dentist on 1986 and was treated by another excision of the mass. Following this second excision, the size of the swelling increased progressively and more rapidly than before. She saw the same dentist 6 months after the second excision. The dentist did not excise the mass again because it bled easily and was too large to handle, instead, he referred his patient to our hospital.

Oral examination showed an irregularly shaped mass occupying the whole palatal space with a mixed red and gray surface, having opposite teeth marks and measuring 5 × 3 cm (Fig. 1). The tumor had its stem at the site of the right second premolar gingiva. Most of tumor mass was expanding from palatal mucosa.

There was no lymph node enlargement. However, one palpable lymph node was detected in the right submandibular, cervical and supraclavicular regions, respectively.

Radiographs showed a poorly defined radiolucency in the area of the second premolar gingiva and the tumor mass seemed to extend into the maxillary sinus.

A ^{99m}Tc-MDP whole body bone and ^{67}Ga whole body scan revealed an area of increased radioactivity in the maxilla at the site of the tumor growth.

A bone marrow aspiration, a gastro-intestinal series and a chest CT scan failed to disclose any abnormalities.

All laboratory data were within normal limits except for serum iron (22 μg/dl), red blood cell count (3.42 × 10/μl), hemoglobin (7.3 g/dl) and CRP (10.3 mg/dl), which together suggested iron-deficiency anemia, probably caused by malnutrition and continuous bleeding from the tumor.

An incisional biopsy of the gingival mass was performed and microscopic examination revealed a stratified squamous epithelium covering a tumor composed of masses of scattered and elongated cells including blood vessels. More than five mitotic figures per 10 high power fields were seen in these cells. Other portions of the biopsy specimen showed areas of elongated cells arranged in a parallel fashion. These cells contained blunt tipped, "cigar-shaped" nuclei. The

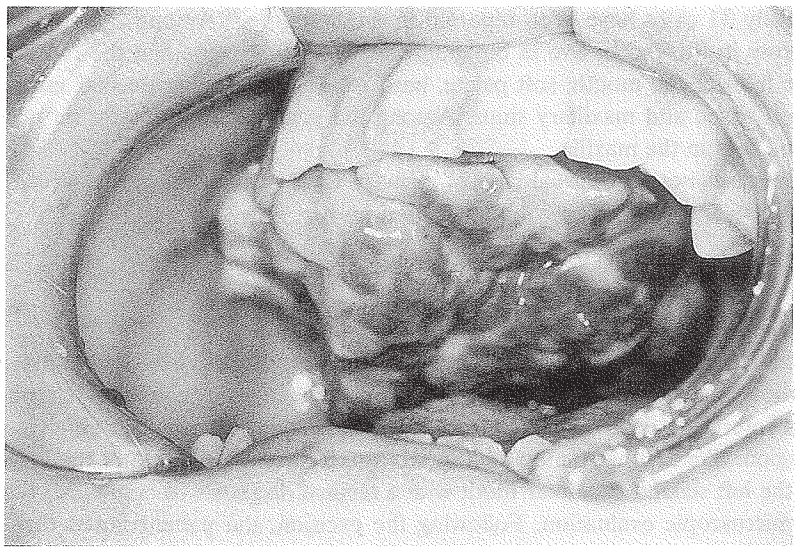

Fig. 1. Intraoral clinical photograph showing the tumor occupping palatal space on initial examination.

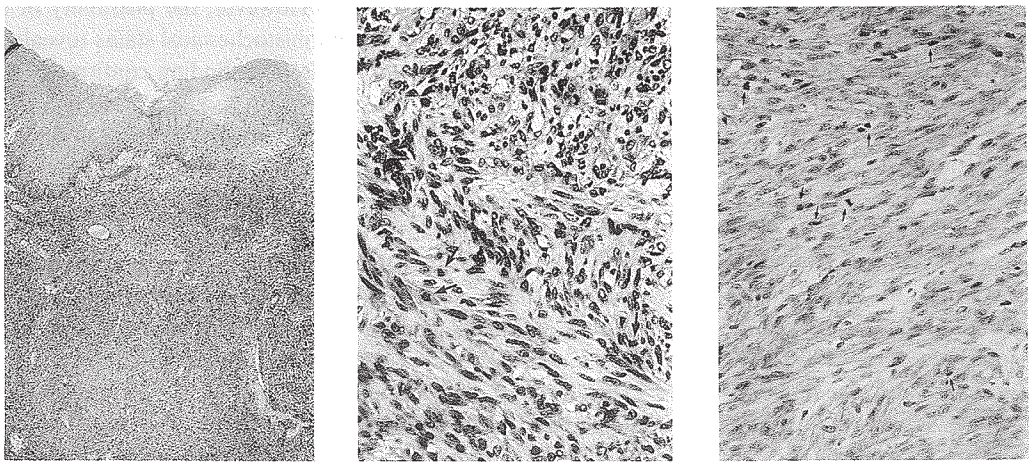

Fig. 2. (left) Low-power photomicrograph showing the interweaving arrangement of the spindle-shaped cells and the developed blood vessels.
(center) High-power photomicrograph displaying prominent cigar-shaped nuclei with perinuclear vacuoles. A number of atypical mitoses were seen (arrows).
(right) Positive cytoplasmic staining for desmin.

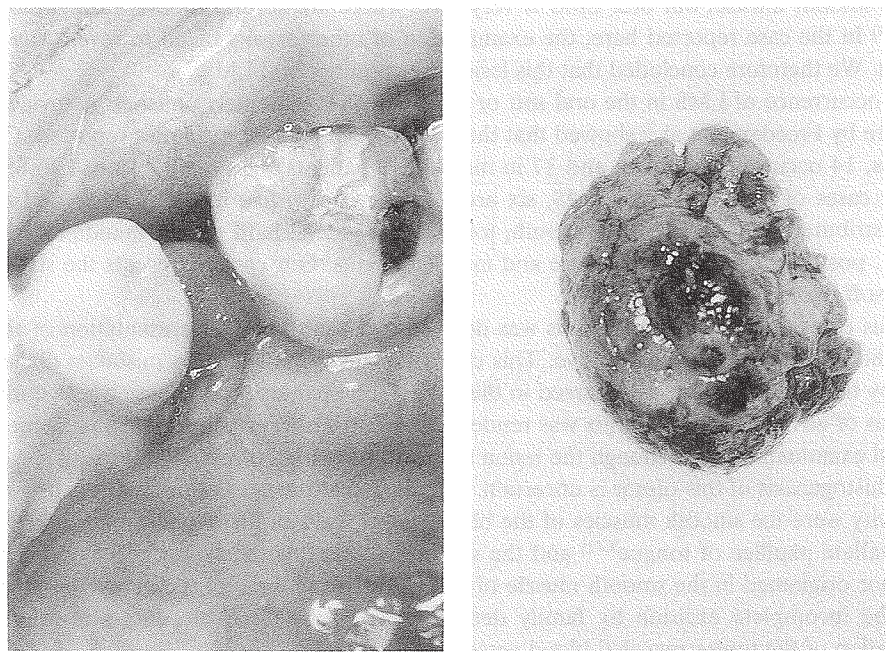

Fig. 3 (left) Intraoral clinical photograph of the originating site of the tumor after resection by suture of the stem.
(right) Photograph of the tumor mass resected from the stem. Macroscopically, most of the tumor was necrotic.

microscopic features of the biopsy suggested a diagnosis of LMS. However, the possibility of a myofibroblastic tumor could not be conclusively ruled out. Immunohistochemical stains revealed positive cytoplasmic staining for desmin in some of the tumor cells. This finding confirmed the diagnosis of LMS (Fig. 2).

Presurgical treatment consisted of the suture of the tumor stem and arterial infusion of 20 mg of adriamycin or day 1 and 20 mg of cis-platinum (CDDP) on days 2—6. One week later, the tumor became necrotic and was cut from the stem. No bleeding was evident. The weight and the size of the tumor were 18 g and 50 × 35 × 12 mm, respectively (Fig. 3).

After expenencing a general recovery, the patient was treated with a right partial maxillectomy from the lateral incisor to the maxillary tuberculum and a right radical neck dissection. Residual tumor was present in the right second premolar gingiva, but no invasion into the maxillary sinus was noted. Postsurgical microscopic examination of the resected specimen revealed features similar to the biopsy, with the exception of a decreased mitosis figure (3 per 10 high power fields). None of the lymph nodes contained any signs of metastatic tumor.

The surgical wound healed uneventfully. The patient was able to wear a maxillary removable partial denture 2 months after the operation. The patient has been followed for 8 years with no signs of tumor recurrence.

DISCUSSION

LMS occurs most frequently in the uterus and the gastrointestinal tract which are both abundant in smooth muscle, but oral LMS is very rare. There are several reports of metastatic oral LMS.[3,4] In the case reported here, the examination of other organs failed to reveal any abnormalities. We therefore concluded that this lesion was primary oral LMS.

The occurrence of LMS in the oral soft or hard tissues is extremely unusual. A review of the literature by Freedman et al.[2] showed that there have been only 31 examples reported. Of these 31 cases, 14 occurred in females and 17 in males. The patients' ages ranged from 1 to 88 years. Among cases of oral soft tissue LMS, six arose in the cheek, five in the gingiva, and the rest were distributed in the floor of the mouth, tongue, soft palate, hard palate, mandibular alveolar mucosa, posterior maxillary soft tissue and maxillary sinus. Our case represents the sixth report of oral soft tissue LMS in the gingiva.

In this case, the definitive diagnosis was delayed because the initial presentation of the neoplasm did not appear to be aggressive. This characteristic might lead to a mistaken diagnosis of epulis by the family dentist, as occurred in this case. Some papers report cases where a mistaken diagnosis of advanced periodontitis was rendered.[5,6,7,8] It is always important to perform an histological examination, even though the lesion seems to be benign or inflammatory.

The histogenesis of this tumor is uncertain. We think that the sites of the tumor's origin in the oral cavity were the smooth muscles of the blood vessel walls in the maxillary gingiva,[9,10,11] the circumvallate papillae of tongue[9,12] and the undifferentiated mesenchyme.[13,14] We believe that the tumor originated in the smooth muscle of the blood vessel walls of the maxillary gingiva because the incomplete excision by family dentist caused substantial bleeding and microscopic examination of the tumor revealed blood vessel development.

Clinically this tumor grew in a polyp pattern with a narrow stem in the gingival mucosa. This growth pattern was very unusual for this kind of tumor. No other reports describe a tumor growth similar to this case.

Immunohistochemical studies of smooth and skeletal muscle tumors confirm positive identification using the cytoskeletal intermediate filament desmin.[15,16] Although smooth and skeletal

muscle tumors may stain with desmin, the light microscopic features of our case did not suggest the diagnosis of rhabdomyosarcoma.

This case was treated by combination of chemotherapy and surgery. The effect of chemotherapy was uncertain, although postsurgical examination of the residual tumor revealed a decreased mitotic rate. Generally wide surgical excision remains the treatment of choice in the oral LMS,[17,18] with irradiation and chemotherapy as adjunctive treatment. Recently, however, there are some reports[19,20] that chemotherapy was effective in controling some cases of soft tissue LMS.

The 5-year survival rate determined in 13 cases with adequate follow-up was as follows: 23% free of disease, 8% alive with disease and 69% who died of disease. Our case has been followed for 8 years with no sign of tumor recurrence.

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