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Case Report

Leiomyosarcoma of the buccal mucosa: a case report with immunohistochemistry findings

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Abstract: Leiomyosarcoma is a relatively uncommon malignant lesion that exhibits smooth muscle differentiation. Occurrence of this tumor in the oral cavity is exceedingly rare, reflecting the paucity of smooth muscle in this region. This article presents a rare case of leiomyosarcoma of the buccal mucosa, which was confirmed by immunohistochemical staining. (J. Oral Sci. 50, 215-218, 2008)

Keywords: buccal mucosa; leiomyosarcoma; immunohistochemistry.

Introduction

Leiomyosarcoma (LMS) is a malignant mesenchymal neoplasm exhibiting smooth muscle differentiation (1). It is more prevalent in older adults and arises most often in the uterus, gastrointestinal tract and retroperitoneal region (2), reflecting the preponderance of smooth muscle in these locations. LMS accounts for 6-7% of all soft tissue sarcomas (2), but its occurrence in the oral soft tissues or jaw bones is very unusual (3). This article presents a case report of LMS arising in the buccal mucosa, which was confirmed by immunohistochemical staining.

Case Report

A 27-year-old woman presented with a 5-month history of a painless nodular mass in the left buccal mucosa. Oral examination revealed that the lesion was nodular, non-tender and firm in consistency (Fig. 1). The lesion measured 1.5×1.5 cm, and the overlying mucosa was normal. The patient's medical history was noncontributory. No lymph nodes were palpable in the cervical region. Excisional biopsy was done under local anesthesia, and the formalin-fixed specimen was processed for histopathological examination. Microscopic examination showed interlacing fascicles of spindle-shaped cells with eosinophilic cytoplasm and vesicular nuclei. Scattered abnormal mitotic figures were also present. In some areas, the tumor cells showed marked cellular pleomorphism with irregularly shaped large, hyperchromatic, bizarre nuclei (Fig. 2). Scattered foamy histiocytes, inflammatory cells and vascular spaces were also evident. Immunohistochemical staining showed positivity for vimentin, smooth muscle actin (SMA) and muscle-specific actin (MSA) and negativity for S-100, cytokeratin and desmin (Fig. 3).

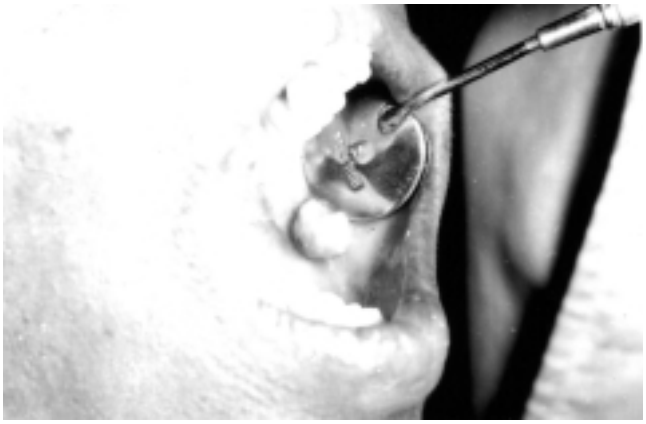

Fig. 1 Intra-oral photograph showing the nodular, non-tender mass in the buccal mucosa.

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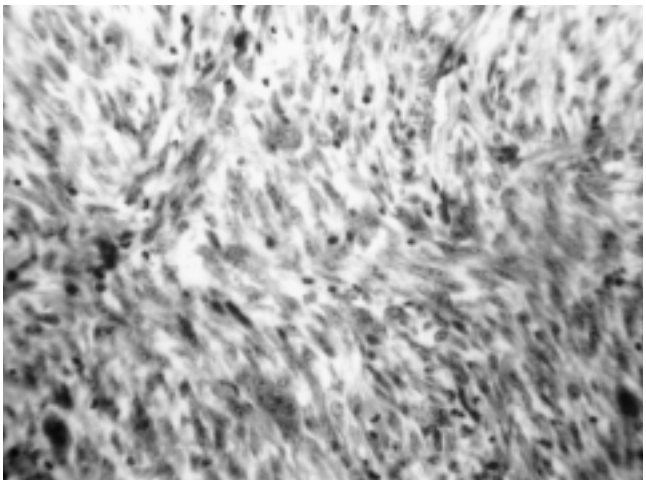
Therefore this case satisfied the immunohistochemical criteria for leiomyosarcoma Grade I. The patient remains disease-free after 18 months of follow up.

Discussion

Only 3-10% of LMS cases 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 commonly affected sites (4). In the head and neck region, smooth muscle is sparse and is found mainly in the walls of blood vessels, erector pili musculature of the skin, circumvallate papillae, and myoepithelial cells of salivary glands (5,6). LMS may also arise from pluripotential undifferentiated mesenchymal cells (7). Clinically, LMS generally appears as a painless, well



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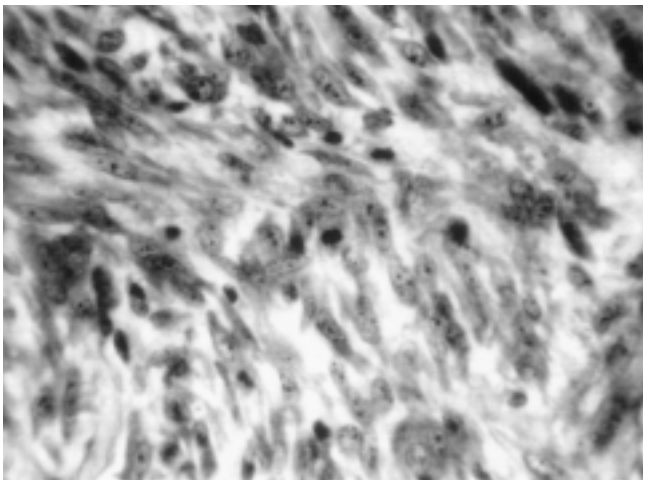
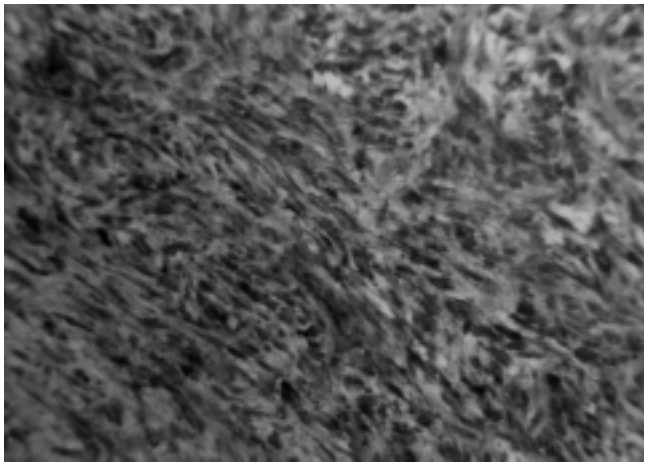


(2)

Fig. 2 Photomicrograph showing (1) interlacing fascicles and bundles of spindle cells (H-E staining, original magnification ×100), (2) spindle cells with cellular atypia and mitotic activity (H-E staining, original magnification ×400).



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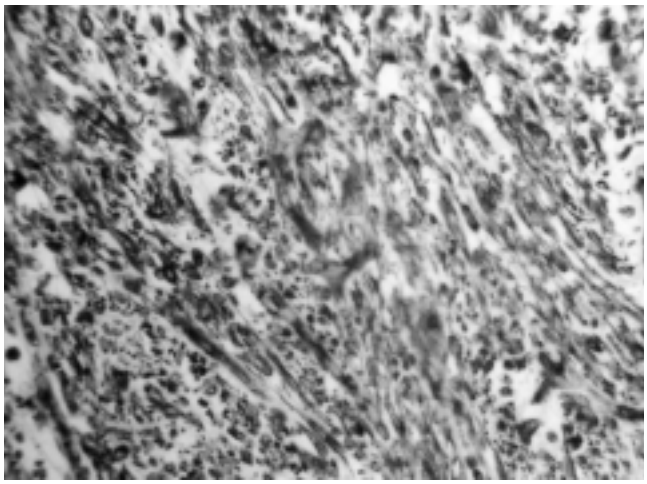

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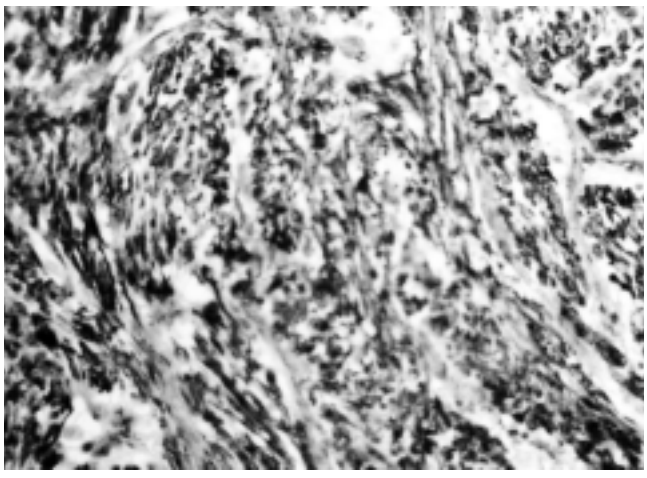

(3)

Fig. 3 Photomicrograph showing reactivity of tumor cells for (1) vimentin, (2) smooth muscle actin, and (3) muscle-specific actin.

circumscribed mass, firmly adherent to the surrounding tissues. Oral LMS has no distinct clinical feature that may aid its recognition. There is no sex predilection, and patients of all ages can be affected (age range 1-88 years) (7). Leiomyosarcoma is typically composed of elongated cells with abundant cytoplasm and a centrally located nucleus. Multinucleated giant cells are common. Microscopic criteria for the diagnosis of LMS are i) a pattern of interlacing bundles of smooth muscle cells, ii) a high mitotic rate, iii) pleomorphism, and iv) bizarre cell forms (8). Ghadially (9) reported that the characteristic ultrastructural features of smooth muscle tumors included i) folded or notched nuclei showing many invaginations, ii) thin intracytoplasmic myofibrils connected by focal dense bodies, iii) abundant micropinocytic vesicles, iv) an external lamina, and v) intercellular junction-like structures. These features are evident in well differentiated tumors. Masson's trichrome staining and immunohistochemical evaluation of muscle antigens can differentiate LMS from other sarcomas and is helpful for diagnosis (10,11). Immunopositivity for vimentin, SMA and MSA has been demonstrated in LMS (1,5,12,13). Although LMS may show immunopositivity for desmin, this feature is not consistent. The tumor tissue should be non-reactive for S-100 protein and cytokeratins (1,14).

LMS arising from the uterus, gastrointestinal tract, and retroperitoneum can metastasize to the lungs, bone and brain, but there are only two reports of metastasis to the oral cavity (1,15). Oral LMS tends to metastasize to the cervical nodes and lungs, and therefore when LMS is identified, it is necessary to determine whether the lesion is primary or a metastasis. The differential diagnosis of LMS can be difficult because of its similarity to other sarcomas composed of spindle cells, such as fibrosarcoma, malignant fibrous histiocytoma and neurogenic sarcoma. Sometimes, light microscopy with routine stains does not allow distinction of this sarcoma from others, especially when the cells are pleomorphic or if areas of nuclear palisading typical of schwannoma are observed. For this reason special stains, and above all immunohistochemistry, play a very important role in diagnosis (16). Early wide surgical excision with radical neck dissection for lymph node metastasis remains the mainstay of treatment (17). Requests for conservative surgical excision to preserve the facial appearance of young patients need to be considered, based on full disclosure of the known behavior of LMS and its nearly certain recurrence after anything less than aggressive initial surgery. Patients with LMS generally show little, if any, objective response to radiation or systemic chemotherapy (18,19). As a 50% rate of recurrence and metastasis and 40% mortality have been reported (14), meticulous and regular follow up is mandatory.

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