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CASE REPORT

Myopericytoma as a Rare Tumor of The Oral Cavity: A Case Report

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Abstract

Myopericytoma (MPC) is a benign mesenchymal neoplasm consisting mainly of oval spindle-shaped myoid-like cells with perivascular growth. Apart from being a rare lesion in all anatomical regions of the body, it is scarce in the oral and maxillofacial regions and only a limited number of cases have been reported in the literature. In this case report; MPC in a 42-year-old female patient with a painless, enlarging lesion in the anterior mandible was presented. Intraoral examination of the lesion revealed a smooth, non-white, pedunculated, fibrotic, painless, and non-bleeding lesion in the lingual aspect of the anterior mandible. Histopathologic examination revealed a tumor consisting of spherical/nodular structures composed of spindle cells and hemangioperistomatous vascular structures in the form of a staghorn. These tumor cells showed no signs of malignancy. The immunohistochemical examination showed that the tumor cells were positive for CD34 and α-smooth muscle actin (α-SMA). α-SMA Protein was also focally positive in nodular structures formed by spindle cells and tumor stroma. S-100 protein was negative in the lesion. Oral MPC is an exceedingly rare tumor which diagnosis can be challenging, and to our knowledge, this is the second case of MPC that occurred in the mandibular gingiva. The aim of this report was to remind clinicians and pathologists that MPC can be encountered in the oral cavity.

Key words: Head and neck; Myopericytoma; Rare tumor; Surgery

Introduction

Myopericytoma (MPC) is a slow-growing, benign mesenchymal neoplasm composed mainly of cells with perivascular growth, with marked differentiation into spindle-shaped perivascular myoid cells called myopericytes.[1,2] The term myopericyte describes neoplastic pericytes that exhibit smooth muscle differentiation around vascular channels. Neoplasms arising from pericyte cells are referred to MPC.[3] This tumor was first described in 1966 as a slow-growing, well-demarcated nodule smaller than 2 cm affecting the skin and superficial soft tissues of the distal extremities.[2] MPC occurs more commonly in males and in the middle-aged adult group.[2,4] In the 2013 World Health Organization Classification of Soft Tissue and Bone Tumors, it was accepted as a type of pericytic (perivascular) neoplasm with myopericytoma and myofibroma.[5] MPC usually occurs as a well-circumscribed, slow-growing, painless, single nodule1,6 but can also be observed as multiple nodules.[6] Multinodular or deeply located tumors behave more aggressively than superficial ones.[4] MPC may occur in a single anatomic region or affect more than one anatomic region.[2] The etiology of MPC is not yet clear.[5] Reports of MPC in the oral cavity are infrequent, and there are only a limited number of cases in the literature. Three of these cases have been reported in the tongue[3,7,8], two in the buccal mucosa[9,10], two in the lips[4,11], and one in the alveolar mucosa[12]. Because of the low incidence of MPCs, there is limited information in the literature. The purpose of this case report was to discuss the MPC clinically and histologically; to highlight the lesion's similarities and differences with previous cases.

Case Report

A 42-year-old female patient was admitted to our clinic with an enlarged lingual lesion of the anterior mandible that she noticed 2 years ago Figure 1. In the anamnesis, the patient was found to have no systemic disease, did not smoke and had no infectious disease. The patient did not complain of pain but her tongue hitting the growing lesion. Radiographical examination revealed no abnormality or pathology in the mandibular and alveolar bone, or teeth Figure 2. No abnormal bone resorption was noted on the periapical

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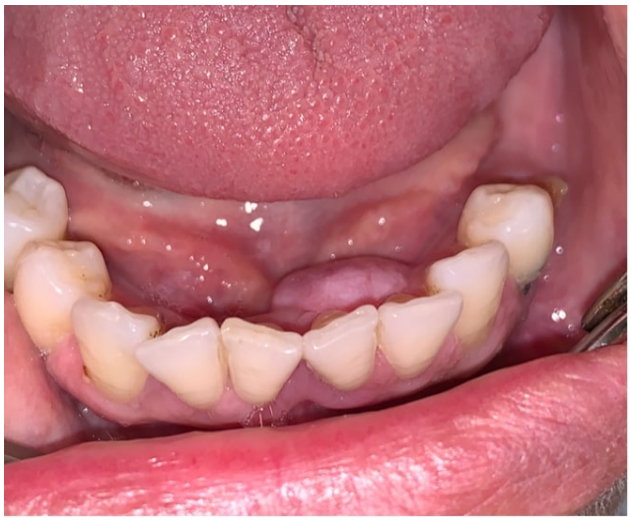

Figure 1. Clinical view of the gingival nodule on the lingual surface.

Table 1. Immunohistochemical markers

Antibody	Host	Brand	Clone	Dilution	Control tissue
α-SMA	Mouse	Thermo	1A4	1:800	Smooth muscle
CD34	Mouse	Thermo	QBEnd/10	1:400	Tonsil
S-100	Mouse	Thermo	4C4.9	1:100	Melanoma

radiography of the area. Because clinical and two-dimensional radiographic examination of the lesion showed no evidence of malignancy, advanced imaging techniques were not required. No pathology was noted in the patient's submandibular, sublingual, and cervical lymph nodes. Clinical examination revealed a smooth, off-white, pedinculated, fibrotic, painless, and nonbleeding lesion approximately 10 mm in size in the lingual region of the anterior mandible. The lesion was excised and fixed in formalin (%10 buffered). Gross examination revealed cream-colored, semicircular, 10x7x3 mm mucosal tissue. In the histopathological examination, there was a non-encapsulated, well-circumscribed, benign lesion that developed nodularly in the oral mucosa covered with stratified squamous epithelium that was keratinized on the sections and atrophic in appearance in most areas Figure 3. It was observed that the tumor consisted of spherical/nodular structures formed by spindle cells and hemangioperistomatous branched vascular structures in the form of staghorn. It was noted that the tumor stroma contained cells with large-oval nuclei, most of which were pale vesicles. Immunohistochemical analysis revealed CD34 and α-SMA positivity in the vascular structures. α-SMA protein was also positive in nodular structures formed by spindle cells and tumor stroma Figure 4. S-100 protein was negative in the entire lesion. The immunohistochemical markers used are summarized in Table 1. The final diagnosis was established as MPC based on the histological and immunohistochemical analysis. No recurrence was detected during the one-year follow-up.

Discussion

MPC belongs to a family of diseases called as 'perivascular myoid cell neoplasms'. This family includes MPC, solitary myofibroma, infantile type myofibromatosis, glomangiopericytoma, angioleiomyoma, and glomangiomyoma.[7] Due to its nonspecific clinical features, it is often difficult to make an accurate diagnosis of MPC.[5] Because of the morphologic similarity between MPC and other perivascular myoid neoplasms, it can be difficult for oral pathologists to differentiate MPC.[7] MPCs present as slow-growing nodules that are occasionally be painful.[1] The tumors are rarely larger than 2 cm, and multiple lesions have been described as occurring

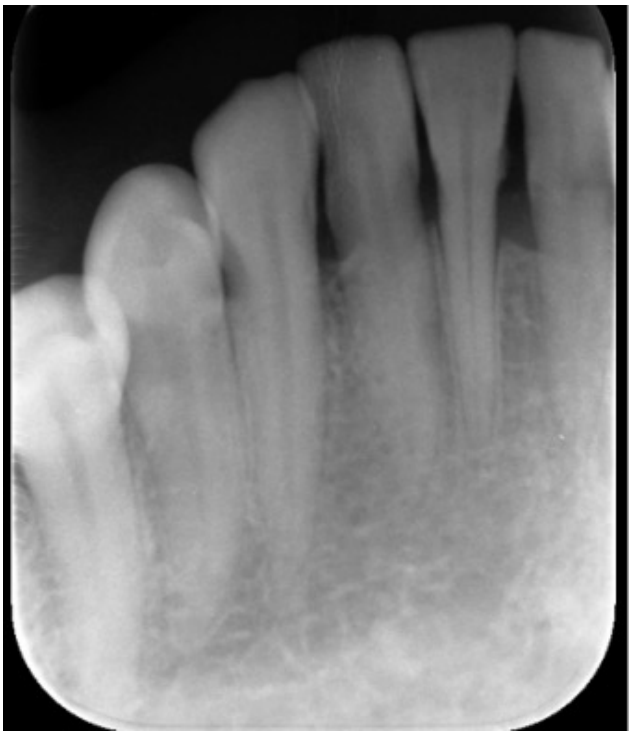

Figure 2. Radiologic examination revealed no abnormality or pathology in the mandibular bone, alveolar bone, or teeth.

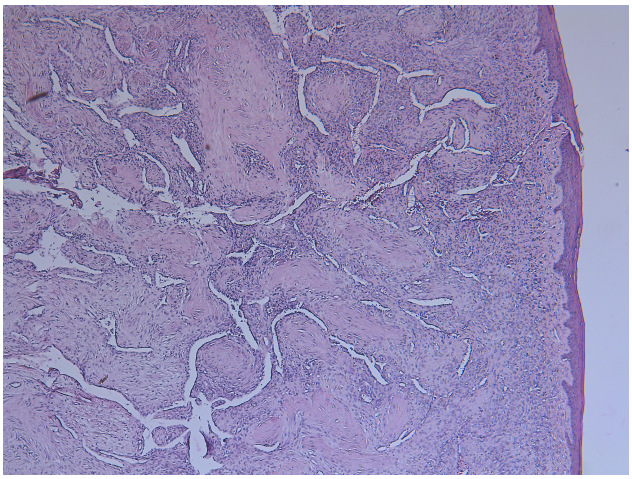

Figure 3. In histopathological examination, an unencapsulated, well-circumscribed benign lesion in the oral mucosa covered with stratified squamous epithelium was seen. (x40 magnification, Hematoxylin&Eosin stain)

metachronously in a single anatomic region.[3] The defining histologic feature of MPC is the characteristic concentric perivascular proliferation of myoid tumor cells.[13] Myopericites are reactive for CD34, α-SMA, and calponin[2], negative for desmin[12] which helps to distinguish them from other perivascular myoid neoplasms.[9,14] However, in some cases, it is unusually reactive for desmin.[2,12] In the histopathological analysis of this case, CD34 was positive in hemangioperistamotous-like vascular areas. In the literature, some cases histologically show CD34 positivity in MPCs[3,4], but there are also cases in which CD34 is negative.[5,7,10] In this context, CD34 was not revealed as a distinguishing feature for MPC. Moreover, in this case, α-SMA is focally positive in tumor stroma and numerous vessels Figure 5. Almedia et al. found perivascular smooth muscle cells surrounding small blood vessels with weak and focal expression in tumor cells.[15] In accordance with the literature,

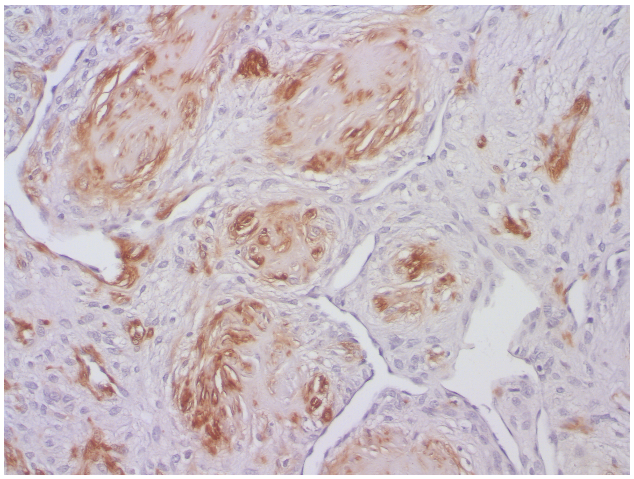

Figure 4. Immunohistochemical analysis revealed α-SMA protein was also positive in nodular structures formed by spindle cells and tumor stroma. (x200 magnification, DAB stain)

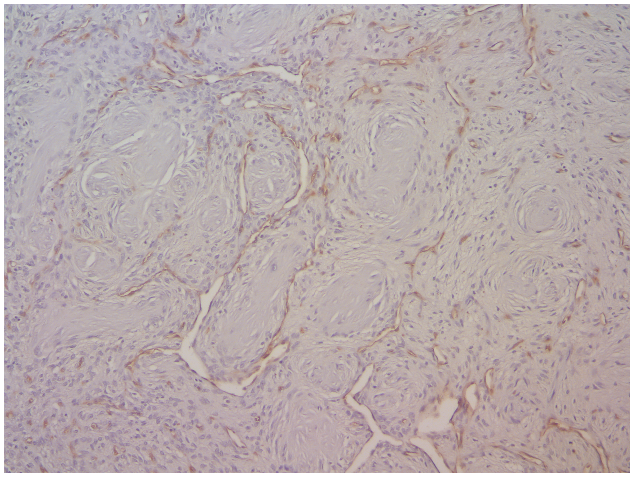

Figure 5. Immunohistochemical analysis showed positivity for CD34 vessels with the hemangiopericytoma-like pattern. (x100 magnification, DAB stain)

S-100 protein was negative in this case as well.[15] According to the literature, preoperative Magnetic Resonance Imaging (MRI) and ultrasound examinations are inadequate or sometimes misleading.[16] Therefore, excisional biopsy and histological examination are essential for definitive diagnosis as performed in this case report. MPC is a slow-growing benign tumor, but in some cases, malignant transformation has also been reported, characterized histologically by pleomorphism, necrosis, and significant mitotic activity of tumor cells.[1,10] There are several lesions in the differential diagnosis of MPC which are glomus tumor, myofibroma, infantile hemangioendothelioma, and angioleiomyoma. Glomus tumor was not considered in the diagnosis because the tumor cells did not have an epithelioid pattern. In MPC, fibromatous areas are not seen as in myofibroma and it can be differentiated from angioleiomyoma by the absence of leiomyomatosis elements.[10] MPC can be distinguished from perivascular epithelioid cell neoplasms such as PEComa by histological features and negativity for HMB45.[10]

The etiology of MPC is not yet clear.[5] Laga et al. reported two cases of MPC significantly associated with trauma.[12] Since the localization was the lingual aspect of the anterior mandible in our case, the existing tumor may have been triggered by trauma to the tongue. Sadahira et al. also reported a patient with multiple periungual MPCs who had chronic obstructive pulmonary disease (COPD).[17] Considering that hypoxia-causing factor-1a induces vascular proliferation, the investigators hypothesized that COPD-induced hypoxia could cause multiple lesions.[17,18] In our case, the patient had no known history of systemic disease, and the lesion was solitary, so hypoxia was not considered an etiologic factor. Most cases of the MPC are benign, but a few malignant and/or recurrent cases have been reported in the literature.[1,10] Although the possibility of sarcoma is quite remote from the diagnosis of MPC, soft tissue sarcomas share similarities with myopericytoma not only in tumor cell morphology but also in microRNA expression pattern.[19] Clinicians and oral pathologists should be very careful in the differential diagnosis between sarcomas and MPCs. Terada reported a case of MPC in the oral cavity with low-grade malignancy with high atypia and mitotic activity in the tumor cells. They noted no recurrence in this case, but strict follow-up was required. The recommended follow-up period for MPC is six months.[10] The literature also reports that MPC occurs in patients with immunodeficiency.[2] In recent years, MPC has been associated with Epstein-Barr virus (EBV) in patients with acquired immunodeficiency syndrome (AIDS), but the mechanism underlying Epstein-Barr-associated MPC remains unclear.[5] When the lesion has occurred in a patient with an inadequate immune system, MPC should be distinguished from Kaposi's sarcoma. The former may be associated with EBV, whereas the latter is associated with human herpesvirus-8 (HHV-8).[4] The occurrence of MPCs with EBV in AIDS patients is a special phenomenon. EBV-associated smooth muscle tumors are a distinct class of neoplasms in which MPC can be included.[20] The recommended treatment for MPCs is local excision and careful follow-up of the area.[4] In our case, no recurrence or malignant transformation was observed after one year of follow-up.

Conclusion

This article presented a case of MPC, which is a very rare lesion in the oral cavity. Based on previously reported cases, it is noted that MPCs are difficult lesions to diagnose and can occur at various sites in the oral cavity. Most cases of MPCs are benign, but local recurrence and rare metastasis may occur, so strict follow-up has been recommended. Although MPC is a rare tumor in the oral cavity, it should be considered in the differential diagnosis, especially in lesions with a history of trauma and patients with immunodeficiency.

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Author Contributions

Study Idea / Hypothesis: E.P. Study Design: E.P. Data Collection: E.P., M.O., I.A.S. Literature Review:E.P, C.P. Analysis and / or Interpretation of Results: E.P., İ.A.S. Article Writing: E.P, I.A.S. Critical Review: C.S.P., E.P., I.A.S.

Conflict of Interest

Authors declare that they have no conflict of interest.

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