Case Report

Myopericytoma of Oral Cavity: A Case Report and Review of Literature
Shivani Bansal, Rajiv S Desai, Subraj Shetty

Abstract
Myopericytoma is a recently proposed term to describe a group of tumors that originate from perivascular myoid cells and show a range of histological growth patterns. The tumor typically arises within subcutaneous tissues of extremities. Only a small number of cases describing myopericytoma in the oral cavity have been reported. Though rare, myopericytoma should be included in the differential diagnosis of well-circumscribed, vascular, slow growing lesions of the oral cavity. We hereby, present a rare case of myopericytoma on lower lip in a 40 year old woman. Literature pertaining to myopericytoma of oral cavity has been reviewed and described.

Keywords: Lip; Myopericytoma; Myofibroma; Neoplasm; Oral Cavity; Perivascular.

Introduction
Myopericytoma (MPC) is a benign tumor that is composed of oval-to-spindle shaped myoid appearing cells with striking tendency for concentric perivascular growth. It is believed that the lesional cells show apparent differentiation towards perivascular myoid cells or myopericytes with expression of myogenic markers. MPC forms a morphologic continuum with myofibroma, angioleiomyoma, and hemangiopericytoma.1,2 In the past, myopericytoma may have been diagnosed as myofibroma or hemangiopericytoma.1

The term “myopericytoma” was endorsed by the World health organisation (WHO) in 2002.1 MPC generally presents as a painless, slow growing subcutaneous nodule that can be present for years. There is a predilection for lesions to involve the distal extremities; however, tumors can also arise at other sites, including the proximal extremities and neck.1,3 Only a small number of cases describing myopericytoma in the oral cavity have been reported. It’s likely that a wider site distribution will be described with increased recognition of the tumor. We hereby, present a rare case of myopericytoma on lower lip in a 40 year old woman.

Case Report
A 40 year old woman reported at out-patient department with a slowly growing, pain-less mass on the lower lip of one month duration. Clinical examination revealed a 5 x 4mm bluish pink, sessile nodule of firm consistency, without discomfort, pain or bleeding from the lesion (Fig 1a). A provisional diagnosis of mucocele was made. An excisional biopsy of the nodule was performed under local anaesthesia and submitted for microscopic examination.

On gross examination the specimen was well circumscribed nodule measuring 5 x 4 x 3mm approximately. Histopathologic examination of haematoxylin and eosin stained tissue section revealed a well circumscribed, unencapsulated lesion consisting of endothelium lined, single-walled vessels surrounded by a concentric perivascular arrangement of neoplastic cells (Fig 1b). These cells were relatively monomorphic, oval to spindle shaped myoid cells with eosinophilic cytoplasm and bland nuclei (Fig 1c). The tumor cells showed positive immunohistochemical reactivity for smooth muscle actin (SMA) (Fig 1d) and were negative for CD34 (Fig 1e).

After correlating the immunohistochemical pattern with the histopathologic features of the lesion, a final diagnosis of myopericytoma was established. As this circumscribed benign neoplasm was completely excised, no further treatment was necessary. Follow up of six months duration showed no sign of recurrence.

Discussion
Myopericytoma is a rare mesenchymal neoplasm, recognized on the basis of both immunohistochemical and ultrastructural studies as a tumor that is derived from the
perivascular myoid cell. The tumor group of MPC has a spectrum of growth patterns that show overlap with those of myofibroma, angioleiomyoma, and hemangiopericytoma. Candidates for the progenitor cell of origin for the myopericyte include the myofibroblast or the pericyte, both of which exhibit properties of modified smooth muscle cells rather than endothelial cells.4

Figure 1: The clinical photograph showing bluish pink, sessile nodule on labial surface of lip (a). The photomicrograph showing stratified squamous epithelium with underlying circumscribed but unencapsulated tumor mass (Haematoxylin – Eosin stain, x50) (b), endothelium-lined, single walled vessels, with concentric arrangement of neoplastic cells (Haematoxylin – Eosin stain, x400) (c). The immunohistochemical stained photomicrograph showing SMA positive perivascular neoplastic cells (SMA x400) (d) and CD34 positive endothelium, whereas the perivascular neoplastic cells are nonreactive (CD34 x400) (e).

Pericyte is viewed as a pluripotential resting stem cell, capable of differentiating along smooth muscle, myofibroblast, pericyte, glomus cell, osseous, fibroblast and adipocyte cell lines. Differentiation of pericytes into myofibroblast and smooth muscle cells has been documented. This concept accounts for the spectrum of tumors and may explain the distinctive features of each variety.4

The term myopericytoma was proposed by Raquena et al5 as an alternate designation for solitary myofibroma, because of
purported myopericytic differentiation. Stout and Murray first described neoplasms showing a pericytic line of differentiation as hemangiopericytomatas, further characterized by Enzinger and Smith. The existence of hemangiopericytoma has been questioned because of number of neoplasms of different lines of differentiation are characterized by a hemangiopericytoma—like vascular pattern. Dictor et al described a case of hemangiopericytoma with a myofibromatous pattern, and coined the term myopericyte, comparable with the myofibroblast.

However, it was not until 1998 that the concept of neoplasms showing perivascular myoid differentiation was established by Granter et al. They adopted the term “myopericytoma” and described myopericytoma, myofibromatosis and glomangiopericytoma as three morphological appearances of MPC forming a spectrum of tumors in the MPC category. In the recent WHO classification of soft tissue tumors MPC and myofibroma are listed as separate entities and glomangiopericytoma appears as a subtype of MPC. MPC arises most commonly in mid adulthood and generally presents as a painless slow-growing subcutaneous nodule and is most common in the lower extremity, followed by the upper extremity, head, neck and trunk. Occasionally, multiple lesions arise in a given anatomic location in a field-like pattern. Tumors rarely exceed 2cm in size.

Reports describing MPC in the oral cavity are scarce. Our review of literature identified 8 cases of MPC in the oral cavity, including the present case. The lesion showed a wide age range of 28 to 67 years with no gender predilection. Out of the 8 reported cases, 4 (50%) were in the lip and 1 (25%) each was in tongue, buccal mucosa, floor of mouth and alveolar ridge respectively (Table 1). A case of MPC of the cheek was cited each by Mentzel et al and Xia Lili et al in their study respectively, with no description of the exact location of the lesion (intraoral or extraoral), hence not included in the present review.

<table>
<thead>
<tr>
<th>Sr. No</th>
<th>Author</th>
<th>Age</th>
<th>Sex</th>
<th>Anatomic Site</th>
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<td>Tongue</td>
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Table 1: Reported cases of oral myopericytoma (F – Female, M – Male, NR – No Recurrence)

MPC is now known to be the unifying designation for tumors derived from, or differentiating along myoepcytes. Microscopically, MPC presents as a well circumscribed, unencapsulated lesion composed of relatively monomorphic, oval-to-spindle shaped myoid appearing cells that show striking multilayered concentric growth around blood vessels. Ide et al in a clinicopathologic re-evaluation of 35 cases of perivascular myoid tumors of oral region found that myopericytoma showed a wide range of growth patterns; concentric perivascular whorls, haemangiopericytomatous areas, glomangiopericytoma type vessels and leiomyomatous foci. Intravascular growth was also seen in their cases. They suggested that MPC, glomus tumor, solitary myofibroma and angioleiomyoma exhibit histologic and immunohistochemical overlap with each other as seen in extra-oral sites.

Because of histological and immunohistochemical overlap differential diagnoses of MPC should include myofibroma (MF) / myofibromatosis (MFs), solitary fibrous tumor (SFT), glomus tumor (GT), perivascular epithelioid cell tumor (PECT), hemangiopericytoma (HPC) and angioleiomyoma (ALM).

MF/MFs displays distinctive biphasic pattern. The peripheral area of the tumor tends to be composed of bundles and sweeping fascicles of plump spindle cells with bland nuclei and abundant eosinophilic cytoplasm. Within the centre of the nodules, there are more cellular areas of primitive small, round to spindled cells with scanty cytoplasm,
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associated with thin-walled irregularly branching hemangiopericytoma-like blood vessels. Immunohistochemically, the smaller round cells and spindled myofibroblastic cells are reactive to SMA and the spindled myoblastic cells are more strongly reactive to panactin HHF35. Like MPC CD34 reactivity has not been recorded in MF/MFs. Therefore, the absence of a multinodular and biphasic pattern, including areas of hyalinised nodule would help to exclude myofibroma.

SFT is characterized by a pattern less proliferation of spindle cells admixed with variable amounts of collagen. Microscopically, they show a wide range of morphological features, from predominantly fibrous lesions containing large collagenized areas and thick/hyalinised-walled vessels to a more cellular and less fibrous neoplasm with numerous thin-walled staghorn branching vessels, and round-to-oval monomorphic tumor cell nuclei. SFT show strong diffuse immunoreactivity against CD34 and CD99. In contrast, CD34 reactivity in a MPC is limited to small vessel endothelial cells, with only isolated reactivity by perivascular lesional cells. SMA highlights the perivascular myoid cells in a MPC.

GT is a perivascular tumor of highly differentiated pericytes. They are composed of cuboidal cells with distinct cell borders and pale cytoplasm, and a round, centrally located nucleus. Although tumor cells may proliferate around vessels, they do not show striking concentric perivascular growth pattern. Spindle cell morphology usually is not seen in GT. The differentiation of a GT from a MPC is based upon the cytomorphological differences of the resident cell population, as these cells consistently express SMA reactivity.

Peripheral epitheloid cell tumor is positive for melanocytic markers, but negative for HMB45 and MITF. HPC is characterized by staghorn-shaped spaces with an intervening proliferation of cells; which is not seen in MPC. HPC is immunoreactive to CD34 & CD99. In contrast to MPC, HPC rarely shows SMA immunoreactivity.

ALM is a well defined tumor entity composed of mature smooth muscle cells and components of blood vessels. Despite overlapping morphologic features, AL usually lack the characteristic perivascular growth of neoplastic cells typically seen in MPC. Desmin is diffusely positive in AL, whereas most of MPC are negative for desmin.

Prognosis of MCP of the oral cavity after adequate excision seems to be good. There has been no report of recurrence of MPC in the oral cavity, but the follow up period for most cases was either short or not mentioned. All the reported cases were of benign myopericytoma. No malignant variant has been reported till date in the oral cavity. Malignant myopericytoma is associated with an infiltrating growth pattern, cytological atypia and mitotic activity.

Conclusion
MPC is a rare entity in the oral cavity. As MPC shares morphologic features with other perivascular myoid neoplasm, it should be included in the differential diagnosis of well-circumscribed, vascular, slow growing lesions of the oral cavity. It is possible that cytogenetic and/or molecular genetic studies may suggest methods for classifying these types of lesions in the future.

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References
4. Dray MS, McCarthy SW, Palmer AA, Bonar SF, Stalley PD, Marjoniemi V.

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