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Myopericytoma of the Oral Cavity

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# Abstract

## Background.

Myopericytoma is a rare mesenchymal neoplasm of pericytic cells demonstrating myoid differentiation. The lesion typically arises within the subcutaneous tissue of the extremities. We report a case that, to the best of our knowledge, is the first case of myopericytoma involving the soft tissue of the oral cavity.

## Methods.

A 36-year-old woman had a 5-mm sessile, whitish-pink, firm tongue nodule. The patient underwent excisional biopsy, and histopathologic examination as well as immunohistochemical analysis were performed.

## Results.

The differential diagnosis by histologic analysis included solitary fibrous tumor, myofibroma, glomus tumor, and myopericytoma. The results of immunohistochemical analysis, when combined with the histologic features, led to a diagnosis of myopericytoma.

## Conclusions.

Applying strict morphologic criteria and appropriately selective immunohistochemical markers will help to distinguish myopericytoma in the oral cavity.

# Introduction

Myopericytoma (MPC) is a benign neoplasm characterized by a perivascular proliferation of oval to spindle-shaped pericytic cells of myoid differentiation.**1** Histologically these cells are arranged in a concentric or whirling pattern around thin-walled vascular channels. Pericytes, first described by Zimmerman in a variety of tissues, have been thought to represent stem cells that may differentiate into either smooth muscle or endothelial cells.**2** The term myopericytes is used to describe neoplastic pericytes exhibiting smooth muscle differentiation around vascular channels.**3** The neoplasm arising from these cells is consequently termed myopericytoma.**4**

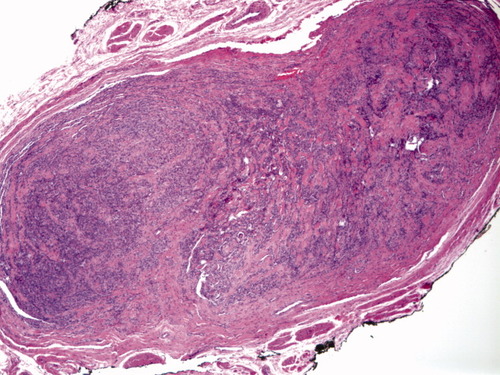
MPC is a slow-growing benign tumor, although a rare malignant transformation histologically characterized by high cellularity, significant mitotic activity, pleomorphism, and necrosis has also been reported.**1** Lesions generally arise within the subcutaneous tissue of the extremities.**5** This rare mesenchymal neoplasm has hitherto not been reported in the oral cavity. We report a case of myopericytoma affecting the connective tissue of the tongue.

# CASE REPORT

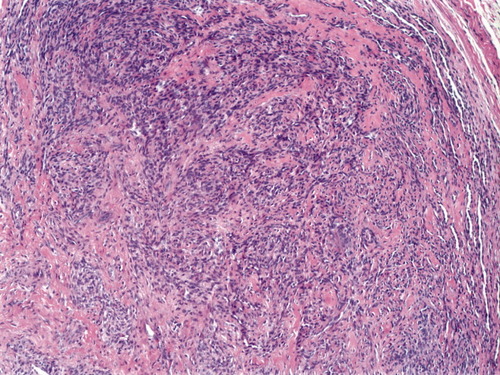
A 36-year-old white woman in otherwise unremarkable state of health was seen with a tongue nodule noticed about 6 months previously. She had no complaint of discomfort, pain, or bleeding from the lesion. On examination, a 5-mm sessile, whitish-pink, nondescript firm nodule was noted on the anterior left lateral tongue. An excisional biopsy of the nodule was performed for microscopic examination.

## Histopathologic Findings.

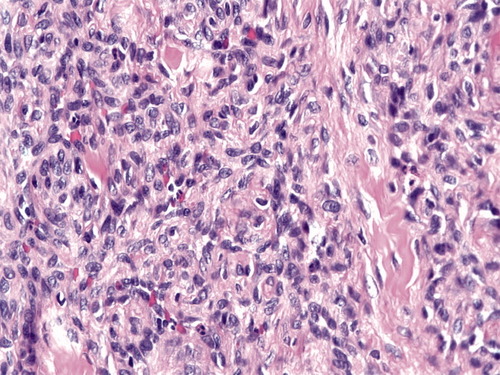
Histopathologic examination of routinely processed, hematoxylin–eosin stained sections revealed a circumscribed benign neoplasm that was, for the most part, unencapsulated within the deeper connective tissue of the tongue (Figure **1**). The cells were arranged in somewhat concentric whorls and swirls. Islands of such cells were separated by hyalinized collagen (Figure **2**). Higher magnification of the cellular areas revealed an arborizing vascular pattern consisting of endothelium-lined, single-walled vessels surrounded by plump nucleated cells with ill-defined cytoplasmic membranes and eosinophilc cytoplasm. The nuclei appeared bland, with some vacuolation. Mitotic figures and inflammatory cells were absent (Figure **3**). The periphery of the tumor exhibited a more prominently open vascular component (Figure **4**). On the basis of these morphological features, the differential diagnosis included solitary fibrous tumor, myofibroma, glomus tumor, and myopericytoma.

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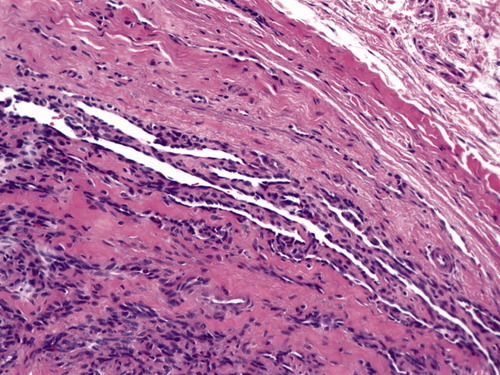
**Figure 1** The tumor is circumscribed but unencapsulated (hematoxylin-eosin stain, original magnification ×40).

[](https://onlinelibrary.wiley.com/cms/asset/93cdf83c-6855-4e12-9975-93d5ceaefa13/mfig002.jpg)

**Figure 2** The cells are arranged in concentric whorls. Islands of such cells are separated by collagen fibers (hematoxylin-eosin stain, original magnification ×100).

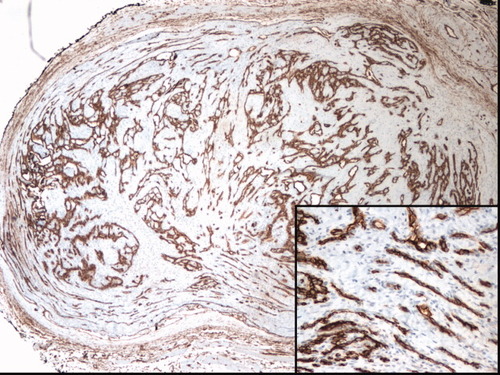
[](https://onlinelibrary.wiley.com/cms/asset/0eed4e65-6376-4d54-969a-af3cf541e009/mfig003.jpg)

**Figure 3** Individual cells possess bland nuclei within an eosinophilic cytoplasm with indistinct cytoplasmic membranes (hematoxylin-eosin stain, original magnification ×200).

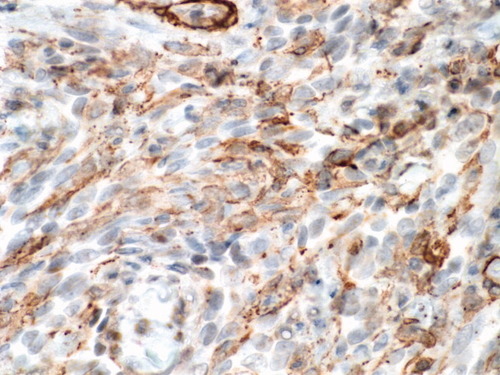
[](https://onlinelibrary.wiley.com/cms/asset/c92c6e18-6496-4cca-a7c5-31efc1a6e027/mfig004.jpg)

**Figure 4** The lesional periphery exhibits a more prominent vascular network (hematoxylin-eosin stain, original magnification ×200).

Immunohistochemical markers used included CD34, smooth muscle actin (SMA), and BCL2. CD34 antibody highlighted small vessel endothelial cells. This staining pattern revealed a well-distributed arborizing vascular network within the tumor (Figure **5**). The perivascular cells expressed reactivity against SMA (Figure **6**). Isolated staining with antibodies directed against BCL2 was seen, with less than 10% of tumor cells expressing this marker. After correlating the immunohistochemical reaction pattern with the histopathologic features of the tumor, a diagnosis of myopericytoma was rendered. As this circumscribed benign neoplasm was completely excised, no further treatment was necessary.

[](https://onlinelibrary.wiley.com/cms/asset/bab46e8f-18a3-4313-880c-21c298ce0911/mfig005.jpg)

**Figure 5** CD34 staining highlights the vascular channels (original magnification ×40), whereas the perivascular cells are nonreactive (inset, original magnification ×100).

[](https://onlinelibrary.wiley.com/cms/asset/154ba51f-d979-40f4-a427-b328232bd7a0/mfig006.jpg)

**Figure 6** The perivascular cells express reactivity to smooth muscle actin (original magnification ×200).

# DISCUSSION

Benign tumors characterized histologically by a perivascular proliferation of oval to spindle-shaped cells exhibiting differentiation toward perivascular myoid cells or pericytes were first described in 1998 as myopericytomas.**6** These neoplasms arise over a wide age range and most frequently affect the subcutaneous tissue of the distal extremities. Lesions affecting the subcutaneous tissue of the proximal extremities and head and neck as well as the trunk have recently been described.**7** Myopericytomas have not been described in the mouth, and the present case appears to be the first documentation of this lesion in the oral mucosa.

MPCs present as slow-growing nodules may occasionally be painful.**8** Tumors rarely exceed 2 cm in size**8** and multiple lesions arising metachronously in a single anatomic site have been described.**1**,**7**,**8**

Histologically, the myopericytoma presents as a well circumscribed, unencapsulated tumor. It is composed of numerous thin-walled blood vessels surrounded by plump ovoid or spindle-shaped myoid tumor cells. This arrangement may appear multilayered and concentric.**6**-**8** The cellular whorls may be separated by a myxoid stroma**8**,**9** or lesions that may be uniformly cellular.**8** Lesional blood vessels vary in number and size from almost inconspicuous and surrounded by solid nests of cells to prominent and gaping. The latter feature may be especially prominent at the periphery of the lesion resulting in a hemangiopericytomatous appearance.**7**,**8** Focally, the perivascular cells may appear glomoid with well-defined epithelioid cytoplasmic membranes and large, round, centrally located nuclei.**7** Because of overlapping morphologic features, a myopericytoma should be differentiated from a solitary fibrous tumor, glomus tumor, and myofibroma.

The solitary fibrous tumor presents as a well-demarcated but unencapsulated soft tissue mass.**8**,**10** It is characterized by a patternless proliferation of spindle cells admixed with variable amounts of collagen.**8**,**11** Alternating hypocellular and hypercellular areas may be separated by thick bands of keloidal collagen and branching “stag-horn” like blood vessels imparting a hemangiopericytoma-like appearance.**8**,**10**,**12** Storiform, herring bone, or a wavy neural arrangement of tumor cells has also been described.**12** Variants demonstrating a perivascular distribution of lesional cells may resemble a myopericytoma. Solitary fibrous tumors show strong diffuse immunoreactivity against CD34**8**,**10**-**12** and CD99.**8**,**10**,**11** Reactivity against BCL2 is less intense and variable.**8**,**10** In contrast, CD34 reactivity in a myopericytoma is limited to small vessel endothelial cells (Figure **5**), with only isolated reactivity by perivascular lesional cells.**8** SMA highlights the perivascular myoid cells in a myopericytoma.**6**-**8** Only 20% to 35% of cases of solitary fibrous tumor are variably positive for SMA expression.**8**

The glomus tumor is a perivascular tumor of highly differentiated pericytes. The perivascular tumor cells are round with a distinct well-defined cell border, pale cytoplasm, and a round, centrally located nucleus. Although the cells are perivascular, a concentric pattern is uncommon.**1**,**8** As these cells consistently express SMA reactivity, the differentiation of a glomus tumor from a myopericytoma is based upon the cytomorphological differences of the resident cell population.

A significant consideration within the differential diagnosis is the solitary myofibroma. This benign neoplasm of myofibroblastic differentiation is commonly seen in the head and neck region, including the oral cavity.**13** At low-power observation, the myofibroma exhibits a characteristic multinodular architecture with a zonal or biphasic appearance. The whorled nodules are often hyalinized with a pseudochondroid change.**7**,**8** Zones of smaller, round cells and spindled cells with pale eosinophilic cytoplasm and elongated, tapering nuclei are also seen. The spindle cells may be arranged in a fascicular pattern. The vascularity may range from compressed crescent-shaped vessels to a prominent hemangiopericytoma-like vascular pattern.**8**,**13** Immunohistochemically, the smaller round cells and the spindled myofibroblastic cells are reactive to SMA**8**,**13** and the spindled myofibroblastic cells are more strongly reactive to pan-actin HHF35.**8** CD34 reactivity has not been recorded in the solitary adult myofibroma but, as in the myopericytoma, CD34 will highlight small vessel endothelium. Therefore, the absence of a multinodular and biphasic pattern, including areas of hyalinized nodules, would help exclude a myofibroma. The whorled, perivascular arrangement of SMA positive myoid cells would be indicative of a myopericytoma.

The myopericytoma shares morphologic features with the solitary fibrous tumor, glomus tumor, and myofibroma. The myopericytoma is a distinct perivascular myoid neoplasm that is characterized by a benign clinical course. Application of strict morphologic criteria and appropriately selective immunohistochemical markers will help in distinguishing the myopericytoma from its mimics in the oral cavity.

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