Clinical and pathologic analysis of myopericytoma in the oral and maxillofacial region

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Objective. The aim of this study was to analyze myopericytoma in the oral and maxillofacial region in terms of clinical appearance, diagnosis, treatment, and outcomes.

Study Design. Data on 5 new patients with myopericytoma in the oral and maxillofacial region treated at our department were collected and analyzed.

Results. There were 2 males and 3 females (age range 10−62 years; mean age 43.8 years). All of the 5 patients presented with masses showing benign biologic behavior. Imaging examinations with use of computed tomography or magnetic resonance imaging showed heterogeneous regions with internal contrast-enhancement or cystic change in 3 cases. All of the patients underwent surgery. Histologic examination showed a broad morphologic spectrum characterized by concentric and perivascular growth of ovoid, plump spindled, and/or round myoid tumor cells. Immunohistochemical examination showed positive staining for vimentin and smooth muscle actin, and negative for CD34 and desmin. During the follow-up period (8−56 months), there was no tumor recurrence.

Conclusions. Myopericytoma in the oral and maxillofacial region always exhibits benign biologic behavior and a heterogeneous region with internal contrast-enhancement or cystic change on imaging examinations. Surgery is the first choice of treatment and results in good clinical outcomes. (Oral Surg Oral Med Oral Pathol Oral Radiol 2019;128:393−399)

Myopericytoma, a benign tumor of perivascular/pericytic differentiation, typically arises in the subcutaneous and superficial soft tissues, mostly in the extremities.[1] Myopericytoma usually appears as a well-circumscribed nodular lesion and shows variable cellularity and a collagenous or sometimes myxoid stroma; it may also show bulging subendothelial proliferation of tumor cells within vessels walls.[2] A broad morphologic spectrum of concentric and perivascular growth of myoid tumor cells is considered to be a histologic characteristic of this tumor.[3] The etiology is not yet clear. In recent years, myopericytoma has been described as being associated with Epstein-Barr virus in patients with AIDS, but the mechanism underlying Epstein-Barr−associated myopericytoma remains unclear.[4-7] In the 2013 version of the *World Health Classification of Soft Tissue Tumors and Bone*, myopericytoma is classified as a type of pericytic (perivascular) neoplasm, along with myofibroma, angioleiomyoma, glomus tumor, and the so-called infantile hemangiopericytoma.[8]

In the oral and maxillofacial region, myopericytoma is rare, and only 10 cases have been reported in the English language literature (Table I).[9-18] Most of these tumors present as a slow-growing, painless masses in different parts of the oral cavity or the maxillofacial region. Surgical resection is reported to be the mainstay of treatment and offers a satisfactory prognosis. It is usually difficult to make an accurate differential diagnosis of myopericytoma because of its nonspecific clinical features, and histopathologically, some hybrid cases may be seen, with 1 case reported with different morphologic and immunohistochemical features of a continuous spectrum of pericytic tumors. Currently, the genetic features of myopericytoma largely remain unknown. In molecular genetic investigations, the presence of a translocation t(7;12)(p21-22;q13-15) was found in 5 cases with pericytic neoplasms,[19] but no other analyses of larger case series and better

Statement of Clinical Relevance

Myopericytoma is rare in the oral and maxillofacial region and presents characteristic pathologic features. A heterogeneous region with internal contrast-enhancement or cystic change on images might be a potential diagnosis characteristic. Surgery is recommended and results in good clinical outcomes.

The study was approved by our hospital ethics committee. Informed consent form was approved by the institutional review board of Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine.

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Table I. Clinicopathologic information of 10 patients from literature review

Published year	*Sex*	*Age (years)*	*Symptoms and features*	*Location*	*Size (cm × cm)*	*Duration of follow-up (months)*	*Treatment*	*Clinical outcomes*
2007	Female	45	A painless, slowly expanding mass in the submucosal space	Buccal mucosal	2.0 × 2.0	108	Radical resection	No recurrence
2007	Female	36	A painless, whitish-pink, non-descript firm nodule	Tongue	0.5 × 0.5	-	Radical resection	-
2008	Male	28	A slowly growing, painless, firm, ulcerated mass	Lip	1.5 × 1.5	36	Radical resection	No recurrence
2010	Female	43	A painless mass	Parotid	5.6 × 5.5	13	Radical resection	No recurrence
2012	Male	61	A reddish elevated mass	Buccal mucosal	1.0 × 1.0	6	Radical resection	No recurrence
2013	Female	42	Multiple slowly growing masses	Parotid	6.0 × 5.0 (the biggest one)	60	Radical resection of one mass	No recurrence for the excised tumor No obvious enlargement in the residual lesions
2013	Female	61	A slow-growing, painless, well-circumscribed firm nodule	Tongue	2.0 × 2.0	18	Radical resection	No recurrence
2014	Male	66	A painless, slow-growing, subcutaneous lump	Parotid	1.2 × 0.7	18	Radical resection	No recurrence
2015	Male	14	A slowly enlarging, painless, freely movable, round, well-circumscribed, firm, nontender mass	Lip	2.0 × 2.0	6	Radical resection	No recurrence
2018	Female	42	An enlarging, soft, fluctuant swelling with an associated bluish tint	Lip	3.0 × 3.0	6	Incisional biopsy	Healing of the mass at first; but relapse 6 months later; radical resection performed; no recurrence observed 3 months later

Table II. Clinicopathologic summary of the 5 patients with myopericytoma in the oral and maxillofacial region

No.	*Sex*	*Age (years)*	*Symptoms and location*	*Size (cm × cm)*	*Report of preoperative fine-needle biopsy*	*Duration of follow-up (months)*	*Treatment*	*Clinical outcomes*
1	Female	46	Parotid swelling	3.7 × 3.5 2.5 × 1.1 2.5 × 1.3	-	56	Radical resection	No recurrence
2	Male	10	Tongue swelling and bleeding	3.8 × 3.1	Certain type of mesenchymal tumors	51		
3	Female	41	Buccal mass	1.6 × 1.5	Certain type of spindle cell type tumors	27		
4	Male	61	Tongue nodule	0.5 × 0.5	-	25		
5	Female	62	Submandibular swelling	3.5 × 2.5	Myopericytoma	8		

characterization of the biologic potential of these lesions have been reported. *BRAF* mutation is considered a novel genetic aberration in myopericytoma pathogenesis and myopericytoma-associated biomarkers by some researchers; however, there is some controversy regarding this opinion.[20,21] Thus, there is no accepted gold standard for diagnosing myopericytoma before pathologic examination.

From December 2013 to December 2017, 5 patients with myopericytoma in the oral and maxillofacial region were treated within our department. The patients' clinicopathologic information was collected, to analyze the clinical appearance, diagnosis, treatment, and outcomes of myopericytoma in the oral and maxillofacial region, with a focus on the diagnosis.

MATERIALS AND METHODS

The study was approved by the ethics committee of the Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, and was in compliance with the tenets of the Helsinki Declaration. Informed consent was obtained from the participants, and the study was approved by the institutional review board. Data on 5 patients who presented with myopericytoma in the oral and maxillofacial region and underwent surgery were collected in this study. These 5 cases were obtained from the Department of Oral and Maxillofacial−Head and Neck Oncology. The study patients' general information, clinical manifestations and symptoms, computed tomography (CT) or magnetic resonance imaging (MRI) images, pathologic examination results, treatments, and clinical outcomes were collected and analyzed (Table II). The pathologic diagnoses were confirmed by 2 experienced oral pathologists. All the enrolled patients were followed up on return visits, during which physical examination was routinely performed. CT or MRI examination was suggested once any suspicious recurrent mass was found.

RESULTS

General condition and tumor location

Five patients (2 males and 3 females; age range 10−62 years; mean age 43.8 years) were enrolled in the study. Two patients had the lesions in the tongue, the other 3 patients had the lesions in the buccal mucosa, submandibular region (Figure 1), and parotid region, respectively.

Symptom

Three patients presented with a single slow-growing, painless swelling or mass, 1 patient presented with multiple masses in the parotid region, and 1 patient presented with a single nodule in the tongue. None of the 5 patients had numbness or other signs of neurologic impairment or any other kind of tumor history.

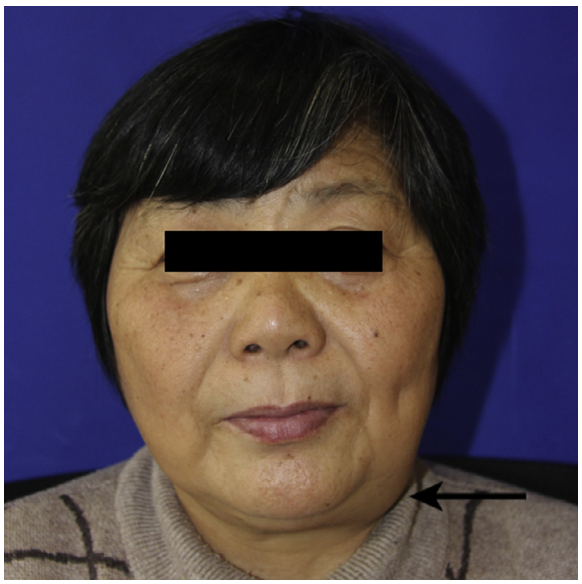

Fig. 1. Front view of patient (No. 5), presenting with a slow-growing, painless mass about 3.5 × 2.5 cm in her left submandibular region (arrow).

Physical examination

A clear-border nodule of 0.5 cm diameter was found in the tongue in 1 patient. In 3 patients with relatively larger single lesions, the sizes of the masses ranged from 1.6×1.5 cm^2 to 3.8×3.1 cm^2. In the fifth patient with multiple masses, 3 masses were found in the parotid region, ranging in size from 2.5×1.1 cm^2 to 3.7×3.5 cm^2; all the masses were well-circumscribed with good movability. No enlarged cervical lymph node (defined as > 2 cm in diameter) was found in any of the 5 patients.

Imaging examinations

CT or MRI was performed in 3 patients (Figure 2). On the images, solid masses with or without sharp borders could be seen. The density of the masses was heterogeneous, with internal contrast-enhancement or cystic change. No bone infiltration caused by the lesions was found in all images.

Laboratory examinations

No obvious abnormalities were found in routine laboratory examinations; serum HIV was negative in all of the 5 patients.

Diagnosis

Before treatment, fine-needle biopsy was performed in 3 patients. One patient was diagnosed with myopericytoma, whereas the other 2 patients had an unspecified type of mesenchymal or spindle cell tumor. Among the 5 patients, only 1 patient obtained the correct preoperative diagnosis; the other 4 patients obtained the correct diagnosis of myopericytoma on the basis of the results of histopathologic examination after surgery.

Pathologic features

Histologically, well-circumscribed, nodular or lobular neoplasms were present in all 5 patients. The tumors showed a broad morphologic spectrum. Characteristic concentric and perivascular accentuation of growing oval myoid tumor cells was found (Figure 3A); some areas were composed of elongated, spindle-shaped tumor cells arranged predominantly in perivascularly growing bundles, as in angioleiomyoma (Figure 3B); numerous gaping and branching, thin-walled vessels surrounded by myoid tumor cells were present in some cases (Figure 3C); mucoid changes could be detected in the mesenchyme (Figure 3D). Mitoses were very rarely identified in all cases. The results of immunohistochemical staining (Table III) showed that all patients were positive for vimentin and α-smooth muscle actin (SMA). CD34 and desmin were negative or only focally positive. There was low expression of Ki-67 ($< 8\%$).

Treatment and outcomes

After the patients signed the informed consent forms, all of them underwent surgical resection under general anesthesia. Extension resection was performed, as in other types of benign tumors. No radiotherapy or chemotherapy was administered. All 5 patients were followed up for 8 to 56 months (average 33.4 months). No recurrence was found, and all patients have remained alive and well.

DISCUSSION

Without data on a susceptible population and distinctive clinical features, the clinical diagnosis of myopericytoma is extremely difficult. In our study, myopericytoma arose in 5 patients, in a wide age range (10−62 years), in different sites of the oral and

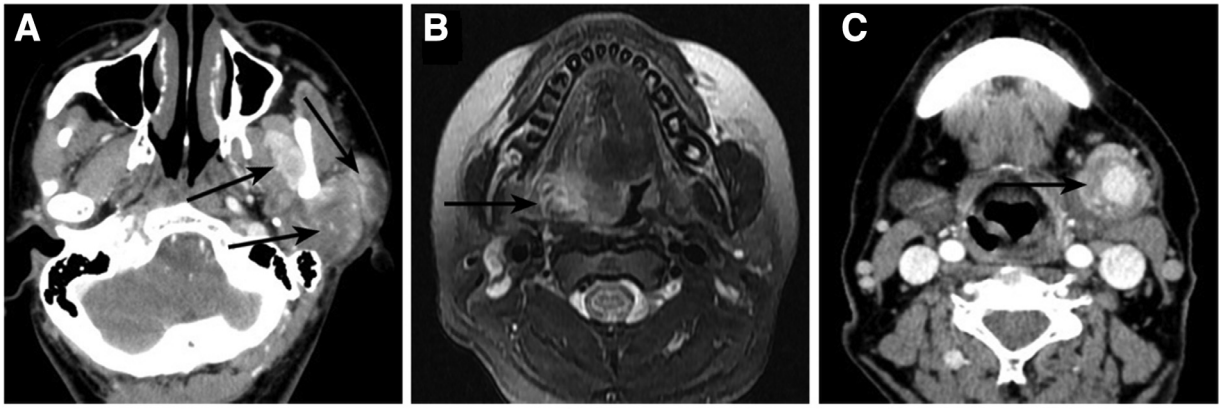

Fig. 2. Computed tomography (CT) or magnetic resonance imaging (MRI) scans from 3 patients. The masses were heterogeneous with internal contrast-enhancement or cystic change. **A**, Patient (No. 1) had 3 masses in the parotid and pterygomandibular space. **B**, Patient (No. 2) had a mass with ill-defined margins in his tongue base. **C**, Patient (No. 5) had a mass in her submandibular region.

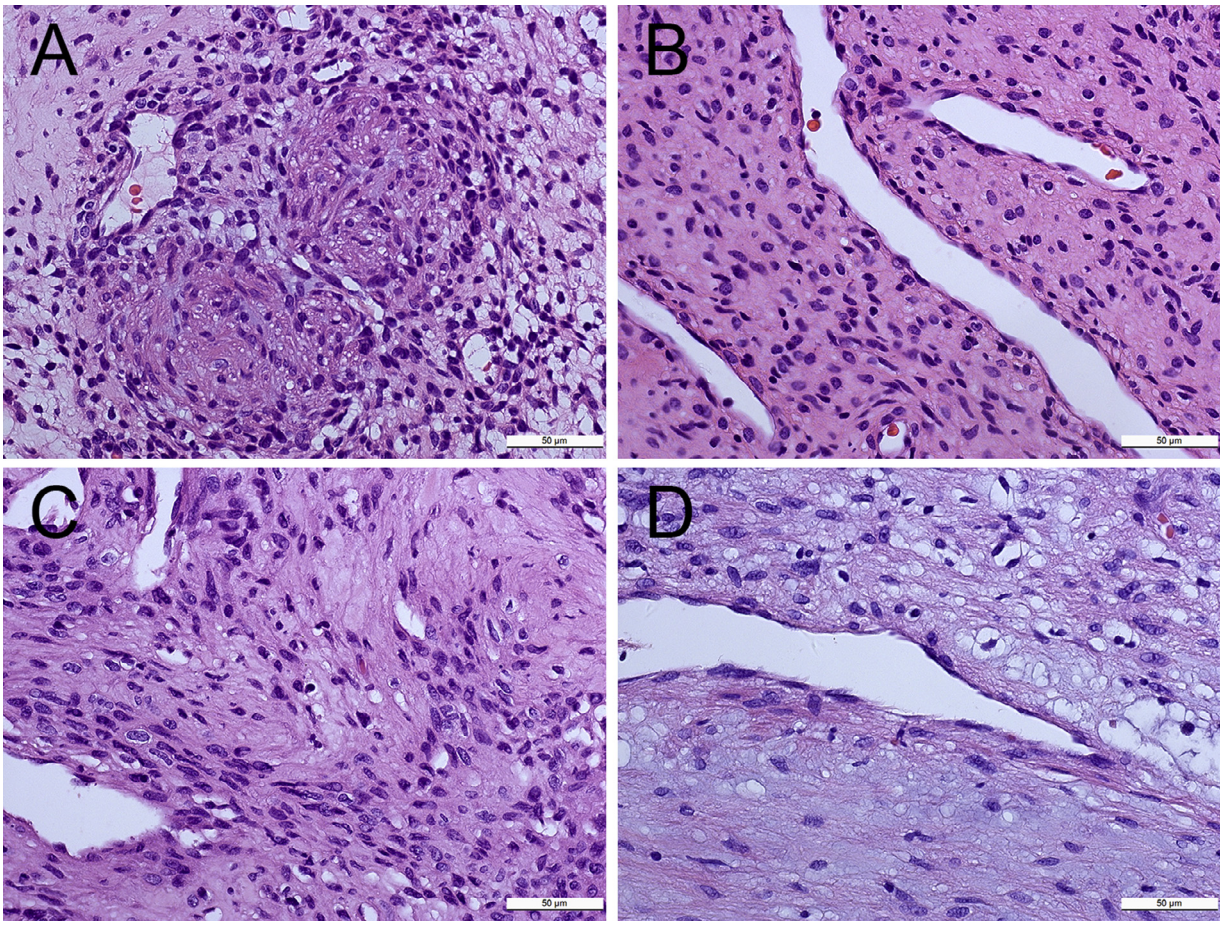

Fig. 3. Pathologic findings of myopericytoma in the oral and maxillofacial region. **A**, Morphologic diversity includes characteristic concentric perivascular accentuation of growing myoid tumor cells; **(B)** angioleiomyoma-like area; **(C)** numerous gaping and branching, thin-walled vessels surrounded by myoid tumor cells; and **(D)** mucoid changes in the mesenchyme (hematoxylin-eosin, original magnification × 40). *High-resolution versions of these slides for use with the Virtual Microscope are available as eSlides: VM05655, VM05656, VM05657, VM05658.*

maxillofacial region, with 1 patient having multiple masses. No specific concomitant symptom was reported. CT or MRI images were consistent in the 3 patients who received imaging examinations, such as heterogeneous regions with internal contrast-enhancement or cystic change. However, these CT or MRI findings could not be considered characteristic or pathognomonic/diagnostic of the tumor.

Of the 5 study patients, 3 underwent preoperative fine-needle biopsy, and only 1 patient (No.5) obtained a precise preoperative diagnosis. In the other 2 patients (No. 2 and No. 3), no diagnosis of a specific pathologic type was given; only an unspecified spindle cell-type or mesenchymal tumor, possibly a sarcoma, was identified. Although the possibility of a sarcoma was quite distant from a diagnosis of myopericytoma, sarcoma does occasionally show similarities to myopericytoma, not only in the morphology of tumor cells but also in the microRNA expression pattern.[22,23] Clinicians and pathologists should be vigilant while making the differential diagnosis of myopericytoma from sarcoma.

Fine-needle biopsy may sometimes be problematic in the accurate identification of myopericytoma because of the limited tissue obtained. Because all of our study cases presented well-circumscribed nodules or masses, as described above, and no evidence of malignancy had been found, we chose not

Table III. Summary of immunohistochemical results

Patient No.	*Vimentin*	*SMA*	*CD34*	*Desmin*	*Ki-67*
1	+	Partial +	–	–	< 5%
2	+	Partial +	–	–	2%–3%
3	+	+	–	Partial +	1%
4	+	+	Partial +	–	< 5%
5	+	+	–	–	1%–8%

+, positive; –, negative; *SMA*, α-smooth muscle actin.

to perform an incisional biopsy to avoid causing pain and disrupting the integrity of the tumor border. The final definitive diagnosis depends on the results of the histopathologic examination of the excised specimens.

Myopericytoma shows a broad morphologic spectrum, as demonstrated in previous studies and this study, and it is characterized by the presence of numerous thin-walled branching blood vessels concentrically surrounded by ovoid, plump spindled, and/or round myoid tumor cells. Sometimes, it has morphologic features that overlap with those of myofibroma, angioleiomyoma, and glomus tumor.[24] Therefore, immunohistochemical staining is necessary and critical for the differential diagnosis of myopericytomas, even though the clinicopathologic characteristics of a glomus tumor are well defined.[25] In our study, the expression of SMA and vimentin was positive in all of the 5 patients, whereas desmin was negative in 4 patients and focally positive in 1 patient. CD34 was mostly negative as well. Although H-caldesmon is considered another indicator of myopericytoma,[1,18,26] it was not detected in our study. These immunohistochemical features are helpful in the differential diagnosis from other perivascular myoid tumors: SMA is negative in myofibromas, whereas desmin staining is invariably observed in angioleiomyomas.[24]

All the patients in our study had benign clinical courses, and the Ki67 index was consistently low (4 patients < 5%; 1 patient < 8%). Moreover, hardly any mitoses were detected. Malignant myopericytoma is extremely rare, but it does exist and shows aggressive biologic behavior with metastases.[27] It is usually described as a noncapsulated tumor, with ill-defined cell borders, zones of necrosis, and frequent mitosis microscopically.[28] From our literature review, only 1 patient had a pathologic diagnosis of myopericytoma with low-grade malignancy in the oral and maxillofacial region.[13]

CONCLUSIONS

Here, we have reported five cases of myopericytoma in the oral and maxillofacial region, presenting with benign biologic behavior and characteristic thin-walled branching blood vessels concentrically surrounded by ovoid, plump spindled, and/or round myoid tumor cells. We propose that a heterogeneous region with internal contrast-enhancement or cystic change, as seen on CT or MRI images, could be a potential clinical feature of oral and maxillofacial myopericytoma. Surgery is the first choice of treatment and results in good clinical outcomes.

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