



Contents lists available at ScienceDirect

# Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology

journal homepage: [www.elsevier.com/locate/jomsm](http://www.elsevier.com/locate/jomsm)

## Case Report

# Myofibroma of the tongue: A case report of a rapidly growing tumor and review of characteristics



Ramy R. Saleh<sup>a,\*</sup>, Stefan Rodic<sup>b</sup>, Umayya Musharrafieh<sup>c</sup>, Mark N. Jabbour<sup>d</sup>, Alain Sabri<sup>e</sup>

<sup>a</sup> Department of Internal Medicine, McGill University Health Centre/McGill University, Montreal, Canada

<sup>b</sup> Department of Biology, McGill University Health Centre/McGill University, Montreal, Canada

<sup>c</sup> Department of Family Medicine, American University of Beirut Medical Center, Beirut, Lebanon

<sup>d</sup> Department of Pathology, American University of Beirut Medical Center, Beirut, Lebanon

<sup>e</sup> Department of Otolaryngology American University of Beirut Medical Center, Beirut, Lebanon

## ARTICLE INFO

### Article history:

Received 20 November 2013

Received in revised form 20 May 2014

Accepted 12 June 2014

Available online 10 July 2014

### Keywords:

Myofibroma

Tongue

Oral cavity neoplasms

Head and neck

Cancer

## ABSTRACT

Solitary myofibroma of the tongue is an uncommon condition reported in 36 cases worldwide in the English literature. Herein, we present the first case of tongue myofibroma diagnosed at the American University of Beirut-Medical Center (Lebanon), found in a 50-year-old female with no evident risk factors or associated co-morbid conditions. The tumor was well-circumscribed, associated with pain, exhibited rapid growth and showed a distinct biphasic pattern upon staining. The unique clinical presentation and the histological findings of this rare tumor are presented and compared with previously reported cases. A review of the literature was used to evaluate this lesion's demographic characteristics using 16 cases, while nine detailed cases were used to determine a general pathological phenotype. We aimed to emphasize features that may express variability between lesions and revealed a possible bimodal distribution of tongue myofibroma occurring in infancy as well as middle-aged individuals. Physicians should include myofibroma in the differential diagnosis of tongue lesions regardless of age.

© 2014 Asian AOMS, ASOMP, JSOP, JSOMS, JSOM, and JAMI. Published by Elsevier Ltd. All rights reserved.<sup>☆</sup>

## 1. Introduction

Solitary myofibroma of the tongue is a rare spindle cell neoplasm, that is, benign in nature [1], accounting for 1% of all tumors in the oral cavity [2]. Myofibromas generally develop in infants under the age of two with a predilection to the head and neck, yet there are a few reported cases in the oral cavity [3]. The rare lesions that do occur in the oral soft tissues have a preponderance to the tongue followed by the buccal mucosa, palate, gingival, mandibular vestibule, and retromolar area, and least commonly the lip [5]. Throughout the literature, most oral spindle cell neoplasms have been reported either as single case reports [1] or as part of a small series [4,5]. The lack of detailed study concerning this tumor makes

diagnosis challenging to the pathologist, particularly due to the subtle histological patterns that overlap with other conditions. We present a case of solitary myofibroma of the tongue with unusually fast growth mimicking a malignancy. Furthermore, we review the available literature in an attempt to better study the clinical and pathological characteristics of this lesion.

## 2. Case report

A 50-year-old female presented to the Department of Family Medicine at the American University of Beirut-Medical Center (AUB-MC) for evaluation of a tongue protrusion that appeared acutely and grew rapidly on the dorsal side of the tongue over a 2-month period. There was some associated pain, but none of these such as dysphagia, difficulty in swallowing or difficulty in breathing was present. The patient did not have other similar lesions and she denied any history of such lesions in her direct family members. Additionally, the patient did not have any significant medical conditions in the past apart from dyslipidemia, for which she was receiving statin therapy. The patient denied any alcohol intake or smoking and there was no surgical or dental manipulation. Intra-oral examination revealed a firm and protruding

<sup>☆</sup> Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

\* Corresponding author at: Division of Internal Medicine, McGill University Health Centre, 670 Pine Avenue West, Montreal, Canada. Tel.: +1 514 934 1934x35800; fax: +1 514 843 2841.

E-mail address: [ramy.saleh@mail.mcgill.ca](mailto:ramy.saleh@mail.mcgill.ca) (R.R. Saleh).

swelling over the dorsal aspect of the tongue, located submucosally but without overlying mucosal changes. There was no evidence of any lymphadenopathy. An MRI of the face and neck with gadolinium revealed a 2.1 cm × 1.5 cm × 2.7 cm homogeneously enhancing right tongue mass in its AP, transverse and cranio-caudal dimensions, respectively, crossing the lingual septum to the contralateral tongue by 9.5 mm (Fig. 1). The lesion was not found to invade or extend to the mandible, maxillary sinus, and floor of the mouth or the skin. Also, the mass did not involve the masticator spaces, pterygoid plates, base of the skull or the internal carotid artery.

Subsequently, the patient was referred to the Department of Otolaryngology-Head and Neck Surgery, where a biopsy was performed. Stained with hematoxylin and eosin (H&E), the tumor was well-circumscribed and characterized by a biphasic pattern with loose and dense areas. The loose areas contained eosinophilic cells with a plump monomorphic nuclear morphology that lacked significant atypia. The dense areas were cellular and composed of round or polygonal cells with mild nuclear pleomorphism. These cells had indistinct cell borders arranged in fascicles, forming a storiform pattern with a low mitotic index (Fig. 2f). Focal areas with a hemangiopericytoma-like vascular pattern were noted within these regions (Fig. 2a–d). Immunohistochemical analysis revealed cytoplasmic staining for both muscle-specific actin and smooth muscle actin (Fig. 2e), alongside corresponding negative staining for desmin, S-100 and CD34. Table 3 illustrates the specific antibodies used. This staining pattern was supportive of cellular differentiation toward a myofibroblastic phenotype (Fig. 2e).

The lesion was then excised completely with primary reconstruction and direct closure using local flap advancement, an intra-oral view of the tumor during surgery is shown in Fig. 3. Margins intra-operatively were deemed negative. After the operation, the patient had difficulty in swallowing with mild articulation problems that required speech therapy. However, she proceeded to full recovery within a few weeks of therapy. The case described herein was referred to as “Case X” when referenced below.

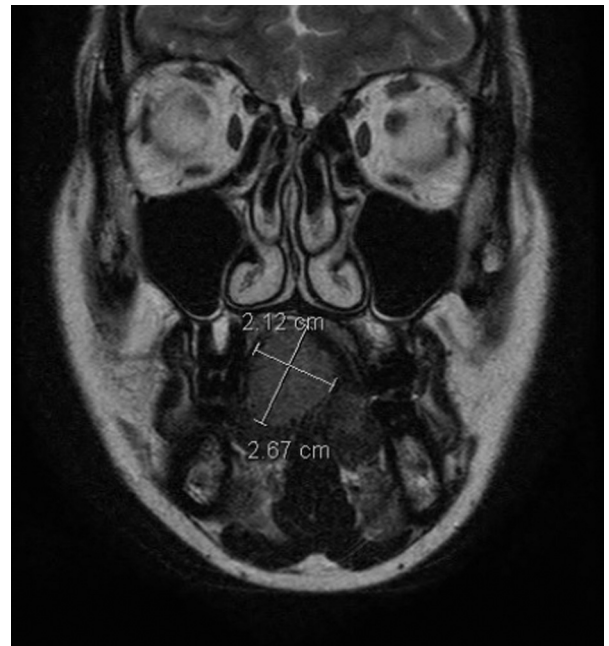


Fig. 1. MRI orbit, face and neck with gadolinium: coronal T2 weighted cut showing the tongue mass with a maximum diameter of 2.67 cm.

### 3. Review methods

A PubMed and Medline search of the English literature was performed from 1960 to 2011 using the following key words: myofibroma, oral cavity neoplasms, tongue neoplasms, and infantile myofibroma. The total number of tongue myofibromas reported in the literature was 36 cases out of 431 non-odontogenic soft tissue benign and malignant spindle cell neoplasms. However, due to the lack of sufficient individual and clinical data, the series of cases reported by Chung and Enzinger [3], Jordan and Regezi [4],

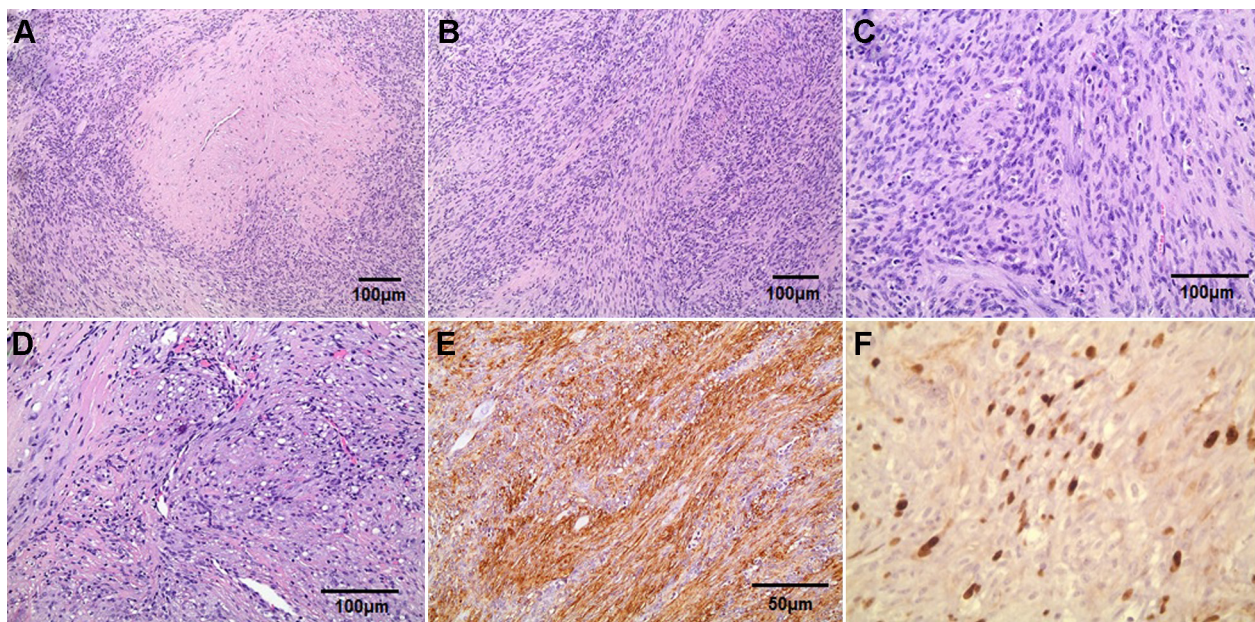


Fig. 2. Photomicrographs of Case X myofibroma: biphasic pattern of growth demonstrated and characterized by alternating loose and dense areas (a; 100× magnification), arranged in fascicles and forming a storiform pattern (b; 100× magnification). The cells within the dense areas are round to polygonal, slightly pleomorphic with a low mitotic index with occasionally scattered lymphocytes (c; 200× magnification). Note the hemangiopericytoma-like vasculature within the dense areas (d; 200× magnification). The cells show evidence of myofibroblastic differentiation by cytoplasmic staining for smooth muscle actin (SMA) (e; 400× magnification) and similar staining for muscle-specific actin (MSA) (image not included). Ki-67 staining of myofibroma demonstrated a proliferative index of less than 3% (f).

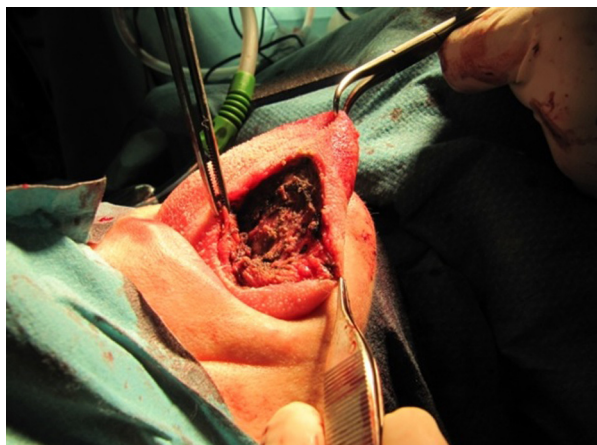


Fig. 3. Intra-oral view of the tumor during surgery.

and Foss and Ellis [5] were excluded from our gathered data. A total of 16 well-documented cases including our own were available for analysis and tabulation (Table 1). Furthermore, a total of nine cases were reported in extensive detail and were used to evaluate the histological and immunohistochemical characteristics of this lesion (Table 2).

#### 4. Discussion

Myofibromas were previously thought to occur mainly in the pediatric age group [16–18], yet half the reported cases involve adult patients. The age of patients with tongue myofibroma ranged broadly from newborns to older adults (55 years). Table 1 shows nine cases from birth to age 20, two cases from age 20 to 40 and five cases ages 40+. The small sample size reported with this specific lesion inhibits strong demographic conclusions to be drawn, however a high percentage of cases in the 40–60 age group could be suggestive of a bimodal occurrence pattern. Earlier publications described myofibroma to be more prevalent in males [3,16,18], while recently Vered et al. [15] showed that the subtype of myofibromas in the oral cavity is actually more common in females. Our data show an equal gender distribution of the lesion on the tongue in particular. Therefore, myofibromas of the tongue should be considered in any differential diagnosis of tongue masses, regardless of age or gender.

Clinically, these lesions are usually described as slowly growing soft tissue swellings that are painless, leave an intact mucosa and range in size from 0.5 cm to 5 cm [19]. The size range of these tumors pertaining to the tongue seems to follow this trend, between 0.4 cm and 4 cm (Table 1). Unusually, in the case of our patient the presentation was painful and growing very rapidly. Due to these unique characteristics, a malignant neoplasm was highly considered in our differential diagnosis. Our case is the only instance on the tongue that pain was reported in the literature; however, no definite perineural invasion was identified in the patient upon histological examination. Although believed to be rare in myofibromas, ulceration was found in one of eight cases that reported ulceration status [9]. Most cases of oral myofibroma are believed to be sporadic, but a family history of the disease was found in a recent case suggesting an autosomal dominant mode of inheritance [10]. Careful review of familial history may prove helpful in diagnosis and should be investigated in future cases to better understand this lesion's degree of heritability. Ultimately, tongue myofibroma is a benign tumor and the recurrence rate was found to be only 12.5% after surgical excision. This is in agreement with the literature, which reports recurrence rates of 7% after surgical intervention in all myofibromas with long-term follow-up [20].

The problem surrounding this benign lesion lies in its potential for misdiagnosis due to its similarity with an assortment of other neoplasms. A central goal of this review was to provide a generalized phenotype of tongue myofibroma and assess its level of variability, thereby allowing more accurate pathological assessment. Histological information was available in only nine studies, which is limiting to the study of variability [Case X, 1, 9–12, 15]. Despite such shortcomings, there was a consensus among the studies in describing this neoplasm. It was unanimously characterized as having an overall biphasic pattern with both fibrous and cellular areas. Mature spindle cells were mostly concentrated on the periphery, arranged in whorls/fascicles, possessing an elongated/tapered nuclei and had eosinophilic cytoplasm. The central areas of the lesion consisted of immature and undifferentiated round to polygonal-shaped cells as well as irregular branching blood vessels (hemangiopericytoma-like). Additional characteristics include indistinct cell borders [Case X, 15] and an abundant extracellular matrix [15], while all nine studies reported minimal nuclear atypia, a low mitotic index and an absence of necrosis. The case presented showed rapid growth highly uncharacteristic of myofibroma, despite exhibiting low mitotic activity and a proliferative index of less than 3% (Fig. 2f). These contradicting

Table 1  
Demographic and clinical data of 16 cases of solitary tongue myofibroma.

Case #	Age	Gender	Size (cm)	Duration	Follow-up	Author
1	6 y	M	0.9	N/A	6 m, NR	Kauffman and Stout [6]
2	13 y	M	1	N/A	14 m, R	Kauffman and Stout [6]
3	5 w	F	2.5	N/A	5 y, NR	Briselli et al. [7]
4	At birth	F	4	N/A	2 m, R	Alpers et al. [8]
5	41 y	F	2.5	3 m	18 m, NR	Speight et al. [9]
6	8 m	M	2	4 m	N/A	Speight et al. [9]
7	34 y	M	2	~2 m	N/A	Tajimi et al. [10]
8	55 y	F	0.4	1 m	N/A	Jones et al. [11]
9–12	11–56 y	F	1.5–2	1–6 m	1–6 m, NR	Lingen et al. [12]
13	5 m	F	2	N/A	3 y, NR	Magid et al. [13]
14	42 y	F	1	2 m	15 m, NR	Montgomery et al. [14]
15	14 y	F	0.8	2 m	2 y, NR	Montgomery et al. [14]
16	18 y	M	1.5	N/A	9 y, NR	Vered et al. [15]
17	23 y	M	1.5	7 m	2 y, NR	Brasileiro et al. [1]
18	51 y	F	2.7	2 m	4 m, NR	Present case
	Range: birth–55 Average: 22.8 y	Male: 43.8% Female: 56.2%	Range: 0.4–4 Average: 1.83		Recurrence rate: 12.5%	Statistics

Demographic and clinical patient data from 16 different cases of tongue myofibroma. In the follow-up column, 'R' indicates that a recurrence took place while 'NR' indicates an absence of any detectable recurrences.

**Table 2**  
Clinical, histological and immunohistochemical characteristics of nine detailed cases of solitary tongue myofibroma.

Characteristic	Positive cases reported	Negative cases reported
<b>Clinical</b>		
Family history	1/5 <sup>[10]</sup>	4/5 <sup>[Case X,1,12]</sup>
Presence of pain	1/6 <sup>[Case X]</sup>	5/6 <sup>[1,9,12]</sup>
Ulceration	1/7 <sup>[9]</sup>	6/7 <sup>[Case X,1,9–12,15]</sup>
<b>Histology</b>		
Biphasic pattern (fibrous/cellular)	9/9 <sup>[Case X,1,9–12,15]</sup>	0/9
Spindle cells in whorl/fascicle pattern	9/9 <sup>[Case X,1,9–12,15]</sup>	0/9
Eosinophilic cytoplasm of spindle cells	9/9 <sup>[Case X,1,9–12,15]</sup>	0/9
Elongated/tapered spindle cell nuclei	9/9 <sup>[Case X,1,9–12,15]</sup>	0/9
Low mitotic index	9/9 <sup>[Case X,1,9–12,15]</sup>	0/9
Immature rounded/polygonal cells	9/9 <sup>[Case X,1,9–12,15]</sup>	0/9
Irregular/branching blood vessels	9/9 <sup>[Case X,1,9–12,15]</sup>	0/9
Necrosis	9/9 <sup>[Case X,1,9–12,15]</sup>	0/9
Encapsulated tumor	2/8 <sup>[Case X,1]</sup>	6/8 <sup>[Case X,1,9–12,15]</sup>
<b>Antibody</b>		
<b>Immunohistochemical</b>		
α-Smooth muscle actin (SMA)	8/8 <sup>[Case X,1,9–12,15]</sup>	0/8
Muscle-specific actin	3/3 <sup>[Case X,11,15]</sup>	0/3
Vimentin	8/8 <sup>[Case X,1,9–12,15]</sup>	0/8
CD34	0/1	1/1 <sup>[Case X]</sup>
Desmin	0/9	9/9 <sup>[Case 1,1,9–12,15]</sup>
S-100	0/8	8/8 <sup>[Case 1,1,9–12,15]</sup>

A summary of the clinical, histological and immunohistochemical characteristics reported in nine studies along with corresponding citations in brackets. Only studies where the positivity status of a characteristic was specifically evaluated were included in the positive/negative column, otherwise the characteristic was considered unreported. A pathological characteristic was not assumed to be negative if it was unreported.

observations may be explained by a decrease in the growth fraction sometime prior to excision or the presence of a very small subset of highly proliferative cells accounting for most of the tumor growth. A characteristic which did vary was tumor encapsulation, most studies reported invasion into muscle/nerve bundles while only two cases involved a well-circumscribed tumor [Case X, 1]. This is contradictory to previous reports of most myofibromas being well-circumscribed [23]. Additionally, one case found the tumor to be evident within several dilated blood veins in the periphery of the neoplasm [9].

Findings such as multicentricity and or vascular invasion could be misinterpreted as evidence of malignancy [3]. Despite the significant degree of overlap in the morphological features between benign myofibromas and myofibrosarcomas, histological differences between the two entities can clearly be perceived. Unlike myofibrosarcomas, the presence of uniformly high cellularity, herring-bone fascicular growth and nuclear atypia was not perceived. As in our case, the biphasic pattern and bundled arrangement of spindle cells help expose the lesion's benign nature from solitary fibrous tumors [23]. Soft tissue leiomyoma and leiomyosarcoma can be ruled out due to the absence of undifferentiated

**Table 3**  
Specific antibodies and staining products: antibodies used during the study of Case X.

Primary Ab	Company	Clone	Dilution
SMA	BioGenex	1A4	1:75
MSA	BioGenex	HHF35	1:100
Desmin	BioGenex	33	1:60
S-100	BioGenex	15E2E2	1:200
CD34	Novocastra	QBEND/10	RTU
Ki-67	Dako	MIB-1	1:100

Ab, antibody; SMA, smooth muscle actin; MSA, muscle-specific actin; RTU, ready to use.

round cells and hemangiopericytoma-like vasculature [21]. Glomangiopericytomas are also devoid of these smaller primitive cells [24]. Immunohistochemical evaluation is particularly useful in differential diagnosis, for example, desmin expression has rarely been reported in myofibroma [21]. A main distinguishing feature of hemangiopericytoma is the rare or focal staining for smooth muscle actin [25], unlike the strong staining reported for tongue myofibroma. The lack of immunoreactivity to CD34 [22] and to S-100 [4] supports the diagnosis of myofibroma rather than fibrous tumors or a neoplasm with a neurogenic origin respectively. Table 2 illustrates the variety of antibodies used in nine different studies of which none had contradictory findings. From this small sample, we postulate that tongue myofibromas are generally positive for α-smooth muscle actin, muscle-specific actin and vimentin while simultaneously being negative for S-100 and desmin [Case X, 1, 9–12, 15].

In summary, our report adds a unique presentation of tongue myofibroma to the literature while characterizing the neoplasm using information from previous studies. Case X was exceptional in its acute appearance, presence of pain, and rapid growth over a 2-month interval. This phenotype matches the one found in a minority of myofibromas of the gingiva or mandible, which exhibited rapid growth and/or were associated with pain [26]. We highlighted a potential biphasic distribution of this lesion and physicians need to be aware of the occurrence of this tumor when assessing a solitary tongue mass in older age groups. So far, tongue myofibroma has proven to be benign in nature and requires only local excision and a conservative approach, yet its potential for misdiagnosis highlights the need for careful pathological evaluation.

## Funding

The study was not funded by a grant or an external party.

## Competing interest

None declared.

## References

- [1] Brasileiro BF, Martins-Filho PR, Piva MR, da Silva LC, Nonaka CF, Miguel MC. Myofibroma of the oral cavity. A rare spindle cell neoplasm. *Med Oral Patol Oral Cir Bucal* 2010;15:596–600.
- [2] Pandey M, Thomas G, Mathew A, Abraham EK, Somanathan T, Ramadas K, et al. Sarcoma of the oral and maxillofacial soft tissue in adults. *Eur J Surg Oncol* 2000;26:145–8.
- [3] Chung EB, Enzinger FM. Infantile myofibromatosis. *Cancer* 1981;48:1807–18.
- [4] Jordan RC, Regezi JA. Oral spindle cell neoplasms: a review of 307 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2003;95:717–24.
- [5] Foss RD, Ellis GL. Myofibromas and myofibromatosis of the oral region: a clinicopathologic analysis of 79 cases. *Oral Surg Oral Med Oral Radiol Endod* 2000;89:57–65.
- [6] Kauffman SL, Stout AP. Hemangiopericytoma in children. *Cancer* 1960;13:695–710.
- [7] Briselli MF, Soule EH, Gilchrist GS. Congenital fibromatosis. Report of 18 cases of solitary and 4 cases of multiple tumors. *Mayo Clin Proc* 1980;55:554–62.
- [8] Alpers CE, Rosenau W, Finkbeiner WE, De Lorimier AA, Kronish D. Congenital (infantile) hemangiopericytoma of the tongue and sublingual region. *Am J Clin Pathol* 1984;81:377–82.
- [9] Speight PM, Dayan D, Fletcher CD. Adult and infantile myofibromatosis: a report of three cases affecting the oral cavity. *J Oral Pathol Med* 1991;20:380–4.
- [10] Tajimi N, Shiraishi T, Ohba S, Fujita S, Asahina I. Myofibroma of the tongue: a case suggesting autosomal dominant inheritance. *Oral Maxillofac Surg* 2013:e1–5.
- [11] Jones AC, Freedman PD, Kerpel SM. Oral myofibromas: a report of 13 cases and review of the literature. *J Oral Maxillofac Surg* 1994;52:870–5.
- [12] Lingen MW, Mostofi RS, Solt DB. Myofibromas of the oral cavity. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1995;80:297–302.
- [13] Magid MS, Campbell WG, Ngadiman S, Godwin TA, Ward R. Infantile myofibromatosis with hemangiopericytoma-like features of the tongue: a case study including ultrastructure. *Pediatr Pathol Lab Med* 1997;17:303–13.
- [14] Montgomery E, Speight PM, Fisher C. Myofibromas presenting in the oral cavity: a series of 9 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2000;89:343–8.

- [15] Vered M, Allon I, Buchner A, Dayan D. Clinico-pathologic correlations of myofibroblastic tumors of the oral cavity. II. Myofibroma and myofibromatosis of the oral soft tissues. *J Oral Pathol Med* 2007;36:304–14.
- [16] Myofibroma and myofibromatosis. Weiss SW, Goldblum JR, editors. Enzinger and Weiss's soft tissue tumors. Mosby: St. Louis; 2001. p. 357–63.
- [17] Wiswell TE, Sakas EL, Stephenson SR, Lesica JJ, Reddoch SR. Infantile myofibromatosis. *Pediatrics* 1987;76:981–4.
- [18] Rubin BO, Bridge JA. Myofibroma/myofibromatosis. In: Fletcher CD, Unni KK, Mertens F, editors. WHO classification of tumors. Pathology and genetics. Tumours of soft tissue and bone. Lyon: IARC Press; 2002. p. 59–614.
- [19] Scheper MA, Difabio VE, Sauk JJ, Nikitakis NG. Myofibromatosis: a case report with a unique clinical presentation. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2005;99:325–30.
- [20] Daimaru Y, Hashimoto H, Enjoji M. Myofibromatosis in adults (adult counterpart of infantile myofibromatosis). *Am J Surg Pathol* 1989;13:859–65.
- [21] Fletcher CD, Achu P, Van Noorden S, McKee PH. Infantile myofibromatosis: a light histochemical and immunohistochemical study suggesting true smooth muscle differentiation. *Histopathology* 1987;11:248–578.
- [22] Guillo L, Fletcher JA, Fletcher CD, Mandahl N. Extrapleural solitary fibrous tumour and hemangiopericytoma. In: Fletcher CD, Unni KK, Mertens F, editors. WHO classification of tumors. Pathology and genetics. Tumors of soft tissue and bone. Lyon: IRAC Press; 2002. p. 86–8.
- [23] Azevedo RS, Pires FR, Coletta RD, Almeida OP, Kowalski LP, Lopes MA. Oral myofibromas: report of two cases and review of clinical and histopathologic differential diagnosis. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2008;105:e35–40.
- [24] Granter SR, Badizadegan K, Fletcher C. Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: a spectrum of tumors showing perivascular myoid differentiation. *Am J Surg Pathol* 1998;22:513–25.
- [25] Veltrini VC, Etges A, Magalhaes MHCG, Araujo NS, Araujo VC. Solitary fibrous tumor of the oral mucosa-morphological and immunohistochemical profile in the differential. *Oral Oncol* 2003;39:420–6.
- [26] Aiki M, Yoshimura H, Ohba S, Kimura S, Imamura Y, Sano K. Rapid growing myofibroma of the gingiva: report of a case and review of the literature. *J Oral Maxillofac Surg* 2014;72:99–105.