Solitary Fibrous Tumor of the Buccal Vestibule: Report of Two Cases

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Abstract Solitary fibrous tumor (SFT) is a rare benign tumor that occurs most frequently in the pleura. It is considered rare in the maxillofacial area. Two new cases of SFT of the buccal vestibule are reported. The previously reported cases of oral SFT are reviewed. The tumors were composed of spindle-shaped cells that were arranged haphazardly and were positive for CD-34, BCL-2, CD-99 and vimentin. Although rare, SFT should be included in the differential diagnosis of oral soft tissue tumors. The clinical presentation and imaging can provide the clinician a better tool for preoperative diagnosis.

Keywords Solitary fibrous tumor · Soft tissue neoplasm · Mandible · Oral cavity · CD34 · Immunohistochemistry

Introduction

Solitary fibrous tumor (SFT) is a rare benign, spindle-cell neoplasm that occurs most frequently in the pleura. It is thought to arise from submesothelial primitive mesenchymal cells, which are pluripotential and may differentiate towards mesothelial cells, fibroblastic cells or other cells [1].

Recently, SFT has been described in a variety of extrapleural sites, such the mediastinum, peritoneum, renal pelvis, periosteum, orbit, meninges and parenchymatous organs. The parenchymatous organs include lung, salivary gland, pancreas, liver, kidney, prostate, breast, adrenal and thyroid glands [2].

SFT of the oral and maxillofacial region is considered rare. 17 case reports of oral SFT have been previously reported [1, 3–13]. Most of them have been reported as single cases, and the diagnosis was established postoperatively, based on the histopathology report. The excellent reports by Alawi et al. [14] describing 16 cases of oral soft tissue SFT, and by O'Regan et al. [15] describing 21 cases of oral SFT, are based on the oral pathology department database, and they focus on the histopathologic and immunohistochemical features, and are lacking the clinical signs and symptoms or imaging modalities that can be helpful to the clinician in establishing the diagnosis, preoperatively.

The purpose of the present report is to describe the clinical and diagnostic features of two cases of sizeable oral SFT and review the literature.

Case Reports

Patient 1

A 50-year-old male presented with a chief complaint of a mass in the buccal vestibule of the right mandible of several month's duration. The lesion was asymptomatic and had increased in size. There was no previous trauma or contributory medical history. Physical examination revealed a well defined submucosal mass involving the vestibule of the right mandible. There was no cervical lymphadenopathy and the

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laboratory data were unremarkable. Axial CT scan, of the mandible revealed a soft tissue mass with no invasion to the periosteum or bone (Fig. 1). During the surgical procedure, a well demarcated mass was found just under the normal oral mucosa. A small peduncle connected the mass to the jaw bone. The surgical specimen was an oval, firm in consistency, soft tissue mass, 41 × 24 mm in size (Fig. 2).

Patient 2

A 43-year-old male presented with a mass in the right cheek of several month's duration (Fig. 3a, b). The lesion was asymptomatic and had increased in size. There was no previous trauma or contributory medical history. Physical examination revealed a well defined submucosal mass involving the right cheek. There was no cervical lymphadenopathy and the laboratory data were unremarkable. Axial CT scan, revealed a soft tissue mass (Fig. 4). During the surgical procedure, a well demarcated mass was found just under the normal oral mucosa. The surgical specimen was a pear-like, firm in consistency, soft tissue mass, 40 × 24 mm in size (Fig. 5).

Histopathology

The microscopy and immunohistochemistry of both cases were identical.

The tumors were composed of monomorphic spindle cells organised into interlacing fascicles in some areas, with more unstructured arrangement in others (hemangiopericytoma-like and stoniform). Immunohistochemically, the tumor cells were strongly and widely positive for vimentin, CD-34, CD-99, and diffusely positive for BCL-2 and less than 1% positive for K-67. There was no staining for epithelial membrane antigen, S-100, cytokeratin, smooth-muscle actin or desmin (Fig. 6a, b).

Fig. 1 Axial CT scan, of the mandible revealed a soft tissue mass located buccal to the body of the mandible, with no invasion to the periosteum or bone

Fig. 2 The surgical specimen was an oval, firm in consistency, soft tissue mass, 41 × 24 × 20 mm in size

Fig. 3 Solitary fibrous tumor of the right cheek. The mass can be seen and felt **a** extraorally and **b** intraorally

Fig. 4 Axial CT of the cheek revealed a soft tissue mass located in the right buccal space

Fig. 5 The surgical specimen was a pear-like, firm in consistency, soft tissue mass, 40 × 24 mm in size

The microscopical and immunohistochemical findings of both cases are compatible with the diagnosis of solitary fibrous tumor.

Discussion

The SFT is a rare, benign, soft tissue neoplasm that occurs most commonly in the pleura. The tumor has also been reported to occur in other serosal surfaces such as the pericardium and the peritoneum [2]. Recently, SFT has been reported in a wide variety of extrapleural sites [16]. Suster et al. [1] reported the occurrence of this tumor in non-serosal locations in 12 patients. The presentation was most often that of a subcutaneous nodule, and the locations involved included the supraclavicular region, the back, the perineum/buttocks/groin, the parotid/submandibular gland/neck, the cheek and the palate. Other reported locations

Fig. 6 Histopathology (a) and immunohistochemistry (b). **a** Solitary fibrous tumor showing variation in cellularity with foci of hyalinization and storiform pattern formation (H&E ×200). **b** Solitary fibrous tumor composed of ubiquitous fibroblast-like cells staining diffusely with CD-34 Immunostain (×200)

have included liver, lung, mediastinum, upper respiratory tract, thymus and orbit.

Early experience suggested that these extrapleural SFT tended to follow a benign clinical course, but it is now appreciated that they too may recur, or metastasize, sometime many years after total removal [17]. The reason for this ambivalent behavior, on the one hand a benign appearance and on the other hand recurrence and metastasis, is unclear. It can not be explained on the histopathology alone.

In the maxillofacial region SFT are uncommon. Only 17 cases have been reported previously. These cases and the present cases are summarized in Table 1 [1, 3–13]. The mean age of the patients with oral SFT was 48.68 years (range 20–83), and the average tumor size was 25.42 mm (range 10–48 mm). The mean age reported by Alawi et al. [14] and by O'Rega et al. [15] was 53.7 (range 19–94) and 53.4 (range 37–83) and mean size of tumor was 24 mm (range 7–75) and 19.8 mm (range 7–54) respectively. No sex predilection was found.

Table 1 Reported cases of oral solitary fibrous tumor

Report (name, year, reference)	Age (years)	Sex	Location	Size (mm)
1. Suster et al. (1995) [1]	50	F	Soft palate	40
2. Suster et al. (1995)	83	M	Cheek	15
3. Shirozu et al. (1995) [3]	57	F	Cheek	25
4. Fornelli et al. (1996) [4]	55	M	Cheek	20
5. Piatelli et al. (1998) [5]	66	M	Tongue	10
6. Kurihara et al. (1999) [6]	34	F	Cheek	15
7. Peretz-Ordóñez et al. (1999) [7]	39	F	Mandible	10
8. Peretz-Ordóñez et al. (1999)	70	M	Buccal mucosa	40
9. Iwai et al. (1999) [8]	27	M	Cheek	20
10. Lukinmaa et al. (2000) [9]	56	M	Cheek	20
11. Lukinmaa et al. (2000)	45	F	Cheek	15
12. Lukinmaa et al. (2000)	70	M	Cheek	10
13. Harada et al. (2001) [10]	32	F	Gingiva	32
14. Shin et al. (2001) [11]	46	M	Buccal space	27
15. Vargas et al. (2002) [12]	65	F	Tongue	48
16. Vargas et al. (2002)	20	F	Cheek	30
17. Sakamoto et al. (2005) [13]	80	F	Tongue	25
18. Present case 1	50	M	Buccal vestibule	41
19. Present case 2	43	M	Buccal vestibule	40

Our cases are in the reported range in terms of age of patients and are considered large (>40 mm) in terms of tumor size.

The origin of SFT is obscure. Most reviewers currently believe that it arises from submesothelial mesenchymal cells and is not specific to the serosa [2].

The diagnosis is supported by a characteristic immunohistochemical profile. SFT stain strongly for CD34 and vimentin but are usually negative for epithelial, vascular, neural crest and muscle markers [18].

Recent studies indicate that a certain specific population of CD-34 positive cells are the possible histogenetic origin of SFT [1]. These cells are primitive mesenchymal cells that are spindled or dendritic in shape and show a wide distribution in various organs and tissues, including the oral cavity [19]. Therefore, the occurrence of SFT in the oral cavity is not surprising. However the exact biological role of CD-34 positive cells in the oral mucosa is still unknown. A possibility exists that the etiopathogenesis of the tumor is from stem cells that are present in the tissue (nest) at the tumor site.

SFT has no specific characteristic features from clinical or imaging aspects, therefore in most reported cases, usually as single cases, the diagnosis of SFT was not set up preoperatively, but based only at the histopathologic report. The present cases with the preoperative clinical presentation, imaging and surgical specimen are able to provide better tools to the clinician to set up a proper preoperative diagnosis.

The microscopic differential diagnosis of SFT involving the oral cavity includes a variety of lesions: hemangiopericytoma,

solitary myofibroma, fibrous histiocytoma, desmoid tumor, spindle cell lipoma, myofibroma, peripheral nerve sheath tumor, leiomyosarcoma and fibrosarcoma [14, 15].

Histologically the SFT will usually appear as a spindle cell tumor with cellular areas and paucicellular/fibrotic areas, showing a histologic pattern with fascicles and sometimes a “patternless” pattern with hemangiopericytoma-like vascular clefts/spaces. Typically there will be little or no mitotic activity and only mild nuclear atypia and mild pleomorphism with no necrotic areas. It does not show immunochemical staining or histologic patterns of mesothelioma and appears more like non-specific fibroblastic lesions.

The solitary fibrous tumor is usually regarded as benign but may be locally “aggressive”. Its behavior cannot be fully predicted from the histology. Several patients with mediastinal lesions died of the disease and pulmonary metastases have been reported. Features suggestive of aggressive behavior include high cellularity, nuclear crowding, atypia, >4 mitoses/10 HPF and tumor necrosis [20].

Although rare, SFT should be included in the differential diagnosis of oral soft tissue tumors.

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