Case Report

Solitary Fibrous Tumor of Retromolar Pad; a Rare Challenging Case

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KEY WORDS
Solitary fibrous tumor;
Retromolar;
CD34;
Immunohistochemistry;
Soft tissue tumor;

Received August 2015;
Received in Revised form January 2016;
Accepted February 2016;

ABSTRACT

Solitary fibrous tumor has a wide spectrum of histopathologic features and many tumors show similar microscopic features. This similarity poses diagnostic challenges to the pathologists and immunohistochemical analysis is required in many cases. Moreover, it is a rare entity in orofacial region which consequently would make its diagnosis more challenging in oral cavity. The knowledge of various microscopic patterns of this tumor contributes to a proper diagnosis and prevents unnecessary treatment. This study reports a case of solitary fibrous tumor in the retromolar pad area and discusses its various histological features and differential diagnoses.

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Cite this article as: Lotfi A., Mokhtari S., Moshref M., Shahla M., Atarbashi Moghadam S. Solitary Fibrous Tumor of Retromolar Pad; a Rare Challenging Case. *J Dent Shiraz Univ Med Sci.*, 2017 June; 18(2): 149-152.

Introduction

Solitary fibrous tumor (SFT), initially reported in the pulmonary pleura, is a rare benign tumor of mesenchymal derivation. [1] The head and neck area is an uncommon location for this tumor. [2] The majority of oral SFT cases present as a slow-growing, painless, well-circumscribed, mobile submucosal mass with variable size and duration. [3] They clinically may be confused with salivary gland tumors, lipoma, vascular malformation, or leiomyoma. [2] Because SFT of the head and neck region is rare, behavior of the tumor in this location is not clearly understood. [4] SFT has a wide spectrum of histopathologic features which poses diagnostic problems in many cases. [2] Therefore, the knowledge of various microscopic patterns of such tumor is essential to elude misdiagnosis and preclude unnecessary treatment. [5] Here, we report a case of solitary fibrous tumor in the retromolar pad area and discuss its different histological features and differential diagnoses.

Case report

A 49-year-old female was referred to an oral and maxillofacial pathology center for evaluation of a slowly growing, painless submucosal mass in the left retromolar pad region. The lesion was extended towards the floor of mouth and had three years duration (Figure 1).

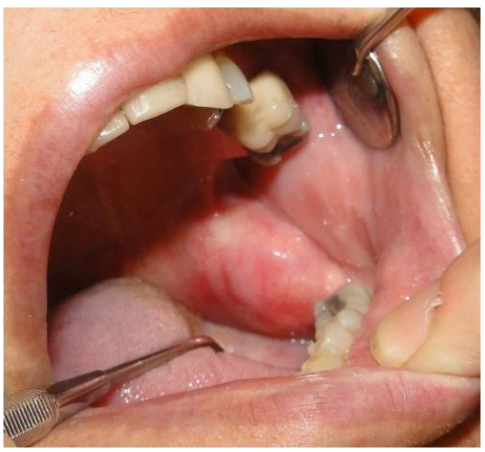

Figure 1: A painless submucosal mass in the left retromolar pad region.

The patient had no history of previous trauma or medical problems. The non-tender mass, measuring 3×3 cm, had firm consistency and the overlying mucosa was intact. The panoramic radiograph did not show any intrabony lesion in mandible. There was no cervi-

cal lymphadenopathy and the laboratory data was unremarkable. The lesion was completely excised under general anesthesia and the differential diagnosis including a salivary gland or soft tissue tumor such as pleomorphic adenoma, mucoepidermoid carcinoma or schwannoma was proposed. The mass was well circumscribed and was easily removed from surrounding tissue. Gross appearance of the cut surface was solid, white, and homogenous (Figure 2).

Figure 2: The gross specimen shows a circumscribed mass with white homogenous cut surface.

Microscopic examination of the lesion showed a soft tissue tumor composed of a patternless proliferation of spindle cells within a background of abundant stromal hyalinization. Myxoid areas and scattered tumoral giant cells were also seen (Figure 3). According to the histopathologic features, the diagnosis of salivary gland tumors was excluded and the diagnosis of a spindle cell tumor was suggested. The immunohistochemical examination showed that spindle-shaped cells were strongly and diffusely positive for CD34 and BCL2 (Figure 4), and negative for desmin, ALK-1 (applied to rule out inflammatory myofibroblastic tumor) and S-100 protein. Therefore, histopathological and immunohistochemical findings were consistent with the diagnosis of solitary fibrous tumor. Clinical follow-up was performed at 3, 6 and 12 months after surgery and she has remained free of disease for 2 years postoperatively.

Discussion

The origin of SFT is uncertain. It is proposed that it arises from submesothelial connective tissue. [6] Submesothelial mesenchymal cells are primitive cells with spindle or dendritic shape. They demonstrate a broad distribution in various organs and tissues. [7-8] SFT is categorized as an intermediate fibroblastic tumor in WHO classification. [9] The most frequent abnormalities identified in SFT includes rearrangements of 12q13~q24 and 9q31q34. [10] Orofacial location of this tumor is rare. Fewer than 80 cases have been reported in the literature. The mean age of patients with oral SFT is 49 years (ranging from 20 to 83 years old), affecting mainly middle-aged and elderly patients. [5] No sex predilection is observed. [6] Based on the reported cases, buccal mucosa is the most common site of occurrence [4] followed by tongue, gingiva, palate, lip, buccal sulcus. Retromolar pad area is a very rare location of involvement. Histologic diagnosis of extrapleural SFT is difficult. [1, 11] This benign tumor has a wide spectrum of morphological features from mainly fibrous lesions containing alternating fibrous areas and hyalinized thick walled vessels to more cellular and less fibrous tumors with a ‘patternless pattern” and thin walled branching vessels. [3, 12] Chan [13] *et al.* described a set of diagnostic criteria for this entity. They proposed that SFTs are usually circumscribed, possessing alternating hypercellular foci and hypocellular sclerotic area, Bland-looking, short, spindly or ovoid cells with scanty and poorly defined cyto

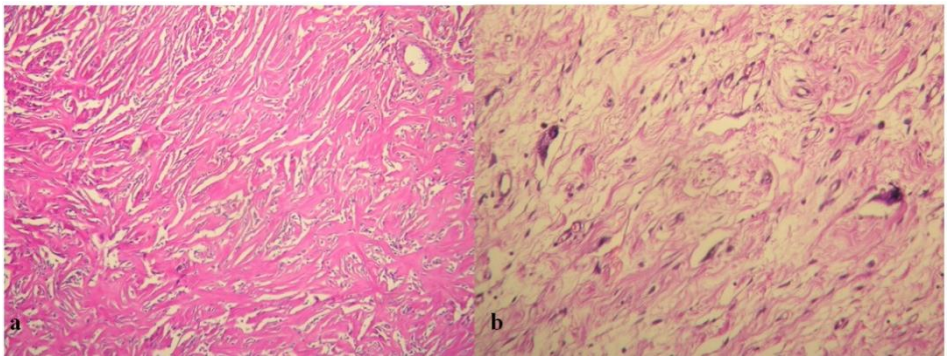

Figure 3a: Sections show a patternless proliferation of spindle cells within a background of dense fibrous and hyalinized matrix (400×). **b:** Myxoid areas and scattered giant cells are shown (400×).

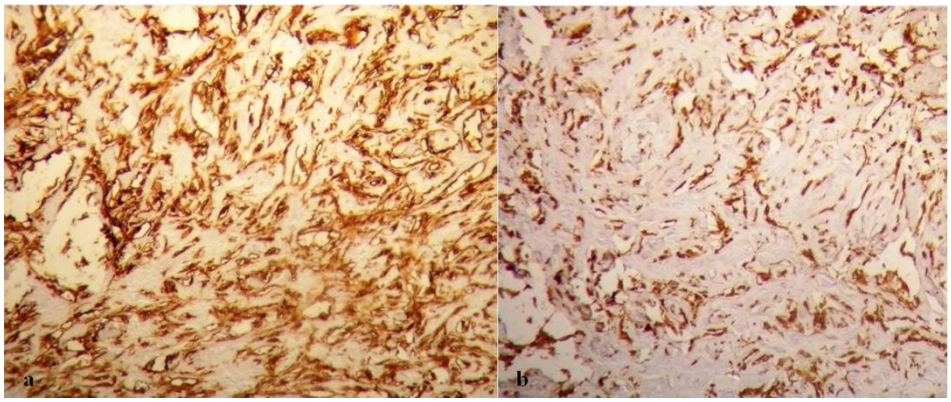

Figure 4a: Tumor cells are strongly positive for CD34 (100×). **b:** Tumor cells show strong positivity for BCL-2 (100×).

plasm, few mitotic activity, haphazard, storiform or fascicular arrangement of spindle cells and close interlacing thin or thick collagen fibrils. All these criteria were found in our case. Two well-defined histologic patterns are recognized in SFT. The more common pattern consists of small capillary-like vessels as well as staghorn vessels with mixed hyper- and hypocellular areas. The less common sclerotic pattern is characterized by dense bands of collagen and only subtle hypercellular areas. [14] Morphological characteristics that allow the detection of malignant variant include infiltrative borders, hypercellularity, nuclear atypia, necrosis and numerous mitoses (≥4 mitoses per 10 high power fields). [6]

The diagnosis of SFT is maintained by a characteristic immunohistochemical study. SFT shows reactivity to CD34, BCL-2, vimentin and CD99, but it is negative for keratin, EMA, S-100, desmin, smooth muscle actin and muscle-specific actin. [1, 4]

Numerous mast cells are present in the SFT and they originate from the multipotent CD34 cells and contribute to the tumor positivity for this endothelial marker. However, SFT is negative for other endothelial markers such as CD31 and factor VIII-related antigen. [15] Moreover, SFT has many T-lymphocytes that are strongly positive for Bcl-2 and CD3. [16] The mitotic activity (Ki-67 positivity) of tumor cells is low. [11] Malignant SFTs show a tendency to lose CD34 immunoreactivity and overexpress p53 and S-100 proteins. [3] The differential diagnosis of oral SFT includes other soft tissue tumors such as fibrous histiocytoma, neurofibroma, schwannoma, hemangiopericytoma, nodular fasciitis, fibromatosis, desmoplastic fibroma myofibroma, giant cell angiofibroma, leiomyosarcoma, fibrosarcoma, low-grade myxofibrosarcoma and myxoid spindle cell lipoma. [11, 15] Infrequently, oral SFT may have entrapment of adipose, skeletal muscle or salivary gland tissue and this make its histological diagnosis more difficult. [11] Some investigations show that cultured SFT cells from oral lesions present some unique cytogenetic finding such as 46, XX, inv(2) (p21q35), t(3;12) (q25;q15). Therefore, it has been suggested that the complicated pathogenetic nature of SFT is possibly tumor- or organ-related. [9]

Intraoral SFTs have an excellent prognosis due to their benign clinical behavior. [2] Complete surgical excision is commonly accepted as the best treatment. [5] Some of malignant SFT cases have been positive for c-kit. Therefore, tyrosine kinase inhibitors such as imatinib have been used in the treatment of SFT cases. [5] Recurrences and distant metastases are expected in malignant SFT. However, definite cases of microscopically benign SFTs may also recur or metastasize. Therefore, long term follow up of SFT patients is highly recommended. [3]

Acknowledgment

The authors are grateful to Dr. Abbas Mirafsharieh, assistant Professor of the pathology department in Modarres Hospital, Shahid Beheshti University of Medical Sciences of Tehran.

Conflict of interest

None declared.

References

[1] Alonso-Rodríguez E, González-Otero T, Castro-Calvo A, Ruiz-Bravo E, Burgueño M. Parotid gland solitary fi-

brous tumor with mandibular bone destruction and aggressive behavior. J Clin Exp Dent. 2014; 6: e299-e302.

[2] Cristofaro MG, Allegra E, Giudice M. Two new localizations of solitary fibrous tumor in the Italian population: parotid gland and oral cavity-review of the literature. J Oral Maxillofac Surg. 2012; 70: 2360-2367.

[3] Li XM, Yu JQ, Xu GH. Solitary fibrous tumor of the soft palate: A report of two cases. Oncol Lett. 2014; 7: 1975-1977.

[4] Yamada H, Hamada Y, Fujihara H, Fukami K, Mishima K, Nakaoka K, et al. Solitary fibrous tumor of the buccal space resected in combination with coronoidectomy. Oral Surg Oral Med Oral Pathol Oral Radiol. 2012; 114: e9-e14.

[5] Satomi T, Hasegawa O, Abukawa H, Kohno M, Enomoto A, Chikazu D, et al. Exceptionally large solitary fibrous tumor arising from the cheek: an immunohistochemical and ultrastructural study with a review of the literature. Med Mol Morphol. 2014; 47: 108-116.

[6] Manor E, Sion-Vardy N, Woldenberg Y, Bodner L. Solitary fibrous tumor of the buccal vestibule: report of two cases. J Maxillofac Oral Surg. 2012; 11: 323-327.

[7] Veltrini VC, Etges A, Magalhães MH, de Araújo NS, de Araújo VC. Solitary fibrous tumor of the oral mucosa--morphological and immunohistochemical profile in the differential diagnosis with hemangiopericytoma. Oral Oncol. 2003; 39: 420-426.

[8] Fletcher CD. The evolving classification of soft tissue tumours: an update based on the new WHO classification. Histopathology. 2006; 48: 3-12.

[9] Swelam WM, Cheng J, Ida-Yonemochi H, Maruyama S, Saku T. Oral solitary fibrous tumor: a cytogenetic analysis of tumor cells in culture with literature review. Cancer Genet Cytogenet. 2009 15; 194: 75-81.

[10] Tapia JL, Goodloe S 3rd, Margarone JE 3rd, Markiewicz MR, Aguirre A. Solitary fibrous tumor with entrapment of minor salivary gland tissue: an unusual presentation that requires exclusion of pleomorphic adenoma. Head Neck Pathol. 2011; 5: 314-320.

[11] Gengler C, Guillou L. Solitary fibrous tumour and haemangiopericytoma: evolution of a concept. Histopathology. 2006; 48: 63-74.

[12] Chan JK. Solitary fibrous tumour--everywhere, and a diagnosis in vogue. Histopathology. 1997; 31: 568-576.

[13] Cox DP, Daniels T, Jordan RC. Solitary fibrous tumor of the head and neck. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2010; 110: 79-84.

[14] O'Regan EM, Vanguri V, Allen CM, Eversole LR, Wright JM, Woo SB. Solitary fibrous tumor of the oral cavity: clinicopathologic and immunohistochemical study of 21 cases. Head Neck Pathol. 2009; 3: 106-115.

[15] Fusconi M, Ciofalo A, Greco A, Pulice G, Macci M, Mariotti M. et al. Solitary fibrous tumor of the oral cavity: case report and pathologic consideration. J Oral Maxillofac Surg. 2008; 66: 530-534.

[16] Kumagai M, Suzuki H, Takahashi E, Matsuura K, Furukawa M, Suzuki H et al. A case of solitary fibrous tumor of the parotid gland: review of the literatures. Tohoku J Exp Med. 2002; 198: 41-46.