Scholars Journal of Dental Sciences (SJDS) **ISSN 2394-496X (Online)**
Sch. J. Dent. Sci., 2014; 1(1):28-30 **ISSN 2394-4951 (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjds.2014.v01i01.007

Review Article

Oral Solitary Fibrous Tumor: Short Review

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Abstract: Solitary Fibrous Tumors (SFTs) are uncommon spindle cell neoplasms that were first recognized as distinctive pleural lesions in 1931. SFTs were thought to originate exclusively in the pleura but later extra pleural lesions including those in the oral cavity were reported. The most common presentation of oral SFTs is in the form of an exophytic growth. Immunoreactivity of SFTs has been determined in the past using a panel of markers and one such marker expressed consistently by SFTs is CD 34 antigen.
Keywords: Solitary fibrous tumor, hemangiopericytoma, CD 34 immunomarker.

INTRODUCTION

Solitary fibrous tumors (SFT) are uncommon spindle cell neoplasms that were first recognized as a distinctive pleural lesion in 1931 by Klemperer and Rabin [1]. Various terms have been used previously to describe this lesion viz Solitary fibrous mesothelioma, benign fibrous mesothelioma and localized fibrous mesothelioma [1].

The etiopathogenisis of the tumors is obscure [3]. Two theories have been proposed. The first theory, which is widely accepted, states that there is multidirectional differentiation of the fibroblasts or of the pluripotent mesenchymal cells located in the connective tissue. Second theory suggests the existence of a specialized cell capable of differentiating into surface mesothelium [4-7].

SFTs occur in patients over a wide age range. Alawi F et al.; reported with a mean age at diagnosis of 56 years [8]; Esther M. O' Regan et al. reported median age of 51 years (range 37–83) [9]. It occurs more frequently around the third to the sixth decades of life without no gender predilection [10-12]. SFTs were initially thought to originate exclusively in the pleura but later extra pleural lesions including those in the oral cavity were reported [13, 14].

The first case of Oral SFT was reported by Suster *et al*. [15]. Various reports have described oral SFTs, on the buccal mucosa, mandibular gingiva, soft palate, palatine tonsil and tongue, in decreasing order of involvement [16]. Most common presentation of SFTs is in the form of an exophytic lesion and they can easily mimic the common oral reactive lesions such as pyogenic granuloma, peripheral giant cell granuloma or even the peripheral ossifying fibromas [17]. Size of the oral tumors has varied from 1cm-4cm with an average size of 2cm [16, 18]. The average time between the patient noticing the growth and reporting to the dentist has varied from 1 month to ten years [19].

SFTs most often have been slow growing tumors but the case reported by Ordonez *et al*. [19], was rapidly enlarging tumor though it was a benign lesion histologically. SFTs of the oral cavity will be asymptomatic, well circumscribed, mobile tumors. The overlying mucosa can be normal to slightly vascular [16, 18, 19]. Reports on conventional imaging modalities for oral SFTs and the radiographic findings are lacking. In a case report, CT scan of the lesion showed a mass within the buccal mucosa with well circumscribed borders. MRI revealed the internal density to be heterogeneous and scintigraphy using Ga [17, 20] disclosed no abnormal uptake [20].

Macroscopically, the tumors can either be encapsulated or unencapsulated and the color may be gray-white. The consistencies of the tumors can be firm to rubbery [2, 3, 6]. The H&E stained sections of SFTs commonly show haphazardly arranged spindle cells, often described as having a "pattern less pattern" with alternating hypercellular and hypocellular foci, few mitotic figures(4/10 high power fields), intimate interwining thick or thin collagen fibrils [16, 19, 21].

Two histopathologic forms of SFT have been described. The Fibrous form of SFT, shows

heterogeneous microscopic appearance, alternating presence of cellular and fibrous areas, hyalinized, thick walled vessels with open lumina, keloidal collagen and strong CD 34 reactivity. The cellular form of SFT shows monotonous microscopic appearance, moderate to high cellularity, little intervening fibrosis, thin walled branching vessels and focal positivity or absence of CD 34 reactivity. This form of SFT has been previously called as Hemangiopericytoma [22].

SFTs can be either benign or malignant and Malignant SFT of the tongue has been reported in literature [23]. Behavior of extra-pleural SFTs is unpredictable. Approximately 10-15% of the extra pleural SFTs show malignant behavior in the form of recurrence and/or metastasis. SFTs that show aggressive behavior clinically may histologically be benign where as histologically malignant lesions can have benign clinical course [23].

Immunoreactivity of SFTs has revealed that more than 90% of SFTs express CD34, CD99 and bcl-2 but negative for smooth muscle actin, cytokeratins, epithelial membrane antigen, S-100 protein, desmin and calretinin [24-30]. CD 34 reactivity is one of the fundamental criteria for diagnosis [18] and is consistently found in SFT. This feature may be a useful diagnostic tool especially in the context of a compatible histopathological background.6 Treatment of choice for SFTs is complete surgical excision [16, 17].

CONCLUSION

Although the initial impression can be that of an innocuous lesion such as a pyogenic granuloma, careful clinical and histopathologic evaluation is invaluable in recognizing this uncommon entity. Identification of this lesion becomes extremely imperative owing to its aggressive and unpredictable behavior like recurrences and metastasis.

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