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Teaching Case

Large solitary fibrous tumor of the oral cavity—Report of a case



Denise Hélen Imaculada Pereira de Oliveira^{a,*}, Assis Filipe Medeiros Albuquerque^b, Matheus Dantas de Araújo Barreto^c, Cassiano Francisco Weege Nonaka^d, José Sandro Pereira da Silva^b, Adriano Rocha Germano^b, Lélia Maria Guedes Queiroz^a

- a Oral Pathology, Federal University of Rio Grande do Norte, Av. Senador Salgado Filho, 1787, Lagoa Nova, CEP 59056-000 Natal, RN, Brazil
- b Division of Oral and Maxillofacial Surgery, Federal University of Rio Grande do Norte, Av. Senador Salgado Filho, 1787, Lagoa Nova, CEP 59056-000 Natal, RN. Brazil
- ^c Federal University of Rio Grande do Norte, Av. Senador Salgado Filho, 1787, Lagoa Nova, CEP 59056-000 Natal, RN, Brazil
- d State University of Paraíba, Campina Grande, Av. Senador Salgado Filho, 1787, Lagoa Nova, CEP 59056-000 Natal, RN, Brazil

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ABSTRACT

The solitary fibrous tumor (SFT) is a rare soft tissue tumor with a substantially benign clinical behavior. The SFT of the oral cavity is a very uncommon entity. It is also of complicated diagnosis because of its extensive morphologic diversity and because of its similarity to many mesenchymal tumors. A 44-year-old man was referred for management of an asymptomatic lesion in the left buccal mucosa, which had been identified 10 years earlier. Intra-oral examination revealed a well-demarcated, fibroelastic, rounded exophytic mass located in the left buccal mucosa. The mass was covered with a non-ulcerated mucosa of normal color and measured approximately 4.0 cm in diameter. Histopathological examination showed proliferation of spindle-shaped cells arranged in fascicles and in a patternless pattern, highly vascularized, with focal staghorn vessels. Immunohistochemical analysis revealed diffuse positivity for CD34 and focal positivity for Bcl-2. Awareness of the morphological diversity of SFT coupled to a judicious use of appropriate immunohistochemical probes should prove valuable to accurately segregate SFT from other spindle cell neoplasms.

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Background

Solitary fibrous tumor (SFT) is a rare benign spindle cell mesenchymal neoplasm that was originally described by Klemperer and Rabin [1] in the pulmonary pleura [2,3]. The terminology of SFT has been somewhat unclear, typically based on the histologic features, location, and presumed histogenesis of the lesion (mesothelial or submesothelial). Because SFT of the head and neck region is rare, the behavior of the tumor in this location is not clearly understood [4].

The histological spectrum of SFT is broad, with appearances often varying from field-to-field within one tumor, thus contributing to diagnostic difficulties [5]. The objective of this paper is to present a new case of SFT of the oral cavity, to assess the clinical

E-mail addresses: denisehelen2011@hotmail.com, denise_helen_odonto@hotmail.com (D.H.I.P. de Oliveira).

and histological features and to correlate these characteristics with immunohistochemical findings.

Case report

A 44-year-old man was referred for management of an asymptomatic lesion in the left buccal mucosa, which had been identified 10 years earlier. Intra-oral examination revealed a welldemarcated exophytic mass covered with non-ulcerated mucosa of normal color, measuring approximately 4.0 cm in diameter (Fig. 1a). An excisional biopsy was performed based on the initial clinical diagnosis of benign soft tissue neoplasm. On gross examination, the lesion was firm and well-demarcated, with a whitish to reddish surface (Fig. 1b). Histopathological examination showed proliferation of spindle-shaped cells arranged in fascicles and in a patternless pattern, with predominance of the latter (Fig. 1c). The tumor cells exhibited a vesicular ovoid nucleus with inconspicuous nucleoli and scanty cytoplasm. No mitosis, cellular atypia, or necrosis was observed. The tumor was highly vascularized, with focal staghorn vessels (Fig. 1d), and well-demarcated by a thin fibrous capsule. Immunohistochemical analysis revealed diffuse positivity for CD34

^{*} Corresponding author at: Departamento de Odontologia, Área de CTBMF e Patologia Oral, Universidade Federal do Rio Grande do Norte, Av. Senador Salgado Filho, 1787, Lagoa Nova, CEP 59056-000 Natal, RN, Brazil. Tel.: +55 84 3215 4138; fax: +55 84 3215 4138.

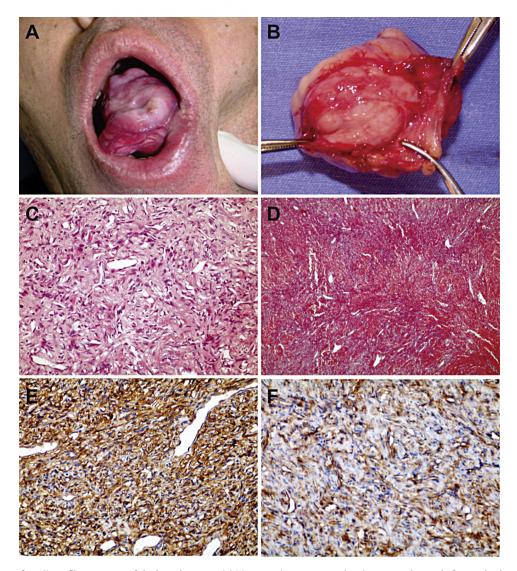


Fig. 1. Composite figure of a solitary fibrous tumor of the buccal mucosa. (a) Macroscopic appearance showing a non-ulcerated of normal color; (b) well-encapsulated measuring approximately 4.0 cm in diameter; (c) microscopic appearance of SFT showing proliferation of spindle-shaped cells arranged in fascicles and in a patternless pattern (H&E); (d) highly vascularized, with focal staghorn vessels (H&E); (e) consistent immunoreactivity for CD34 in the spindle cell component; (h) moderate immunoreactivity for bcl-2.

(Fig. 1e) and focal positivity for Bcl-2 (Fig. 1f). There was no immunoreactivity to α -smooth muscle actin, desmin, or S-100 protein. The definitive diagnosis of SFT was issued. The patient remains under careful monitoring (Fig. 2).

Discussion

Originally described by Kemperer and Rabin [1] as a "primary neoplasm of the pleura", early reports uniformly regarded SFT as a tumor of mesothelial origin [6]. This view was gradually modified with the observation that SFT can affect many other sites of the body [5]. Nowadays, it has been suggested that SFT arises from pluripotential mesenchymal cells located in the connective tissue [7], and the World Health Organization (WHO) currently classifies it as a probable fibroblastic/myofibroblastic tumor [2].

Clinically, SFT of oral cavity presents as a well circumscribed submucosal mass, staining normal, asymptomatic and slow-growing, and can often be confused with injury as mucocele, salivary gland tumors, lipoma, vascular malformations, leiomyoma, among other [8]. Our case is distinguished by its exceptionally large dimensions.

Microscopically, typical SFTs exhibit a patternless architecture characterized by a combination of hypercellular and hypocellular areas separated from each other by thick bands of collagen and branching hemangiopericytoma-like vessels. The round to spindle-shaped tumor cells have scanty cytoplasm with indistinct borders and dispersed chromatin within vesicular nuclei [2]. Mitoses are generally scarce, rarely exceeding > 3/10 high-power fields. Mature adipocytes and giant multinucleated stromal cells have been reported in some cases of SFT [2,7,9].

The diagnosis of SFT may be difficult in extrapleural locations due to its wide histological spectrum [10]. Moreover, diagnosis of SFT in a small biopsy specimen is known to be difficult, because of extreme intratumor variability and the close similarity of isolated parts of individual soft tissue tumors [11].

Establishing a final diagnosis requires both conventional microscopic and immunohistochemical analyses. In 1997, Chan [5] published a list of diagnostic criteria for benign and malignant forms. Criteria for the benign form have been subdivided into essential and secondary. Essential diagnostic features include: circumscription; alternating hypercellular foci and hypocellular sclerotic foci; spindly or ovoid cells with scanty and poorly-defined

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