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

A R T I C L E   I N F O

Article history:
Received 29 August 2013
Received in revised form 1 September 2014
Accepted 29 September 2014

Keywords:
Solitary fibrous tumor
Oral cavity
Differential diagnosis
Immunohistochemistry
CD34

A B S T R A C T

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.

© 2014 Elsevier GmbH. All rights reserved.

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 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 well-demarcated 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.
(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 mucocle, 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: circumscriptin; alternating hypercellular foci and hypocellular sclerotic foci; spindly or ovoid cells with scanty and poorly-defined
cytoplasm; low (<4/10 high-power fields) mitotic activity; haphazard, storiform, or fascicular arrangement of spindle cells; intimate intertwining of thin or thick collagen fibrils with spindle cells and CD34 positivity. The secondary criteria are seen in some but not all cases, for example, hemangiopericytoma-like vascular pattern; dilated, thin-walled vessel with perivascular sclerosis; isolated mononuclear or multinucleated large cells with hyper-chromatic nuclei [5].

Tumor cells in SFT are characteristically immunoreactive for CD34 and variably positive for CD99 and Bcl-2 [12,13]. Anti-CD34 antibody, which recognizes a 110-kDa transmembrane cell surface glycoprotein found on myeloid progenitor cells, is the most consistent and reliable immunohistochemical marker for SFT [14–16]. On the other hand, neoplastic cells are negative for cytokeratins, S-100 protein, epithelial membrane antigen, smooth muscle actin, and Factor VIII [13,17]. The immunohistochemical profile of the present case is in agreement with those reports.

Given the wide range of morphology, SFT can potentially be mistaken for other benign and malignant soft tissue tumors [7,12,14,16]. The differential diagnosis of benign SFT involving the oral cavity includes a variety of lesions, especially fibrous histiocytoma, schwannoma, and, most importantly, hemangiopericytoma [14,17].

The extent to which a given histopathologic feature is present can help determine the most appropriate diagnosis. For example, although SFT can show a storiform pattern similar to that of fibrous histiocytoma, the storiform pattern in SFT is not as widespread as that in fibrous histiocytoma. Similarly, the presence of sclerosis favors the diagnosis of SFT, because it is less frequently seen

in fibrous histiocytoma than in SFT [17]. The fibrous histiocytoma is generally negative for CD34. However, a positive reaction to CD34 antigen has been found in more than 18% of the cutaneous fibrous histiocytomas (dermatofibromas) [18]. For these cases, the differential diagnosis has to be based on morphology because dense collagenization and variable cellularity are present in SFT but absent in fibrous histiocytoma [19]. Schwannomas can exhibit variable cellularity, but they are almost always positive for S-100 protein, whereas SFTs are negative for this immunohistochemical marker [3,20].

The major diagnostic dilemma lies mainly in distinguishing SFT from hemangiopericytoma due to their overlapping histological features [3,21]. The diagnosis of hemangiopericytoma should be considered with extreme caution. Both tumors share the “staghorn-like” vascular pattern and cellular morphology [12,15]. In addition, SFT and hemangiopericytoma have been reported to show similar immunohistochemical and ultrastructural profiles. SFT and hemangiopericytoma display immunoreactivity for CD34, Bcl-2 and CD99 antigens. Ultrastructural studies have demonstrated pericytic, fibroblastic, and myofibroblastic differentiation in both tumors [3,16]. These findings have led pathologists to consider hemangiopericytoma and SFT as a spectrum of the same entity [22].

Even when strict morphologic criteria are adopted and appropriate immunostains employed, the diagnosis of SFT remains difficult, especially at extrapoleric sites, due to extreme intratumoral variability and close similarity of single parts of individual tumors with prognostically different benign and malignant soft tissue neoplasms [11]. Overall, there are no histopathologic or immunophenotypical features that separate SFTs arising in the head and neck from those arising at other sites.

A malignant variant of SFT has been described. This tumor usually shows increased cellularity, at least focally moderate to marked cytological atypia, areas of necrosis, numerous mitoses (>4 mitoses/10 high-power fields) and/or infiltrative margins [2,19,23]. Malignant SFT exhibits histologic features resembling malignant FH, malignant hemangiopericytoma, or fibrosarcoma. Angiosarcoma and Kaposi’s sarcoma have to be considered in the differential diagnosis because these neoplasms are CD34-positive. However, these tumors express other endothelial markers that are usually negative in SFT [7,19,24].

Because of the rarity of this tumor little is known about its clinical behavior. The prognosis is based on tumor location, size and histological features considered as malignancy parameters [11]. Two case series of intraoral SFTs, with extensive follow-up information, showed no signs of local recurrence and/or malignant transformation after treatment [12,14]. In contrast, malignant and metastatic SFTs have been reported to occur in extraperic sites. Vallat-Decouvelae et al. [23] and Gold et al. [25] found local recurrence in 4.3% and 6.7% and metastases in 5.4% and 5.3%, respectively. Sites of distant metastases were lung, liver, bones, mesentery, mediastinum and retro-peritoneum [7].

The treatment of choice is usually surgical excision with wide, tumor-free margins, without requiring additional treatment since those located in the head and neck rarely recur and metastases are not known. Therefore, a close long-term follow-up has to be recommended even after radical excision [15]. On the whole, SFTs have a benign clinical course, but the clinical behavior is unpredictable, and the relationship between morphology and clinical behavior is poor.

Conflict of interest

The authors declare that they have no conflicts of interest.
References