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Case Report

Sporadic sclerotic fibroma of the tongue: A rare case

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ABSTRACT

Sclerotic fibroma (SF) is a distinct benign fibrous tumor that may manifest as a sporadic solitary mass or as skin nodules in patients with Cowden syndrome. Sporadic sclerotic fibroma is an extremely rare finding in the oral cavity. In this report, we describe the first case of sporadic SF presenting on the tongue. Histologically, the tumor was a well-demarcated hypocellular nodule. The collagen fibers were densely arranged in a vague, whorled pattern with clear clefts between the collagen bundles. The immunohistochemical staining was strongly positive for vimentin and weakly positive for bcl-2. Although rare, this report confirms that SF should be acknowledged as part of the differential diagnosis of soft tissue tumors of the oral cavity.

1. Introduction

Sclerotic fibroma (SF) is a rare benign fibrous tumor that was first described as a manifestation of Cowden syndrome [1]. Cowden syndrome is a rare, inherited disorder that affects the skin and mucous membranes [1,2,4–6]. The syndrome presents with multisystem abnormalities associated with benign and malignant tumors [1,4–6]. Sclerotic fibromas in Cowden syndrome may be found as nodules on the skin [2]. The nodules can function as cutaneous markers of the syndrome [2] that may help establish a diagnosis and lead to the discovery of associated malignant transformations.

However, there are also reports of SF presenting as sporadic, solitary lesions in patients without Cowden syndrome [3,7–12]. In a report by Rapini and Golitz [3], 11 lesions were described as sporadic solitary dermal fibromas that were histologically similar to those found in Cowden syndrome. In their study, these dermal fibromas were then suitably named "sclerotic fibromas" due to their unique histologic presentation. Histologically, these tumors have been described as sharply demarcated [2,3], hypocellular entities with sclerotic [3] collagen bundles presenting in a whorling pattern [1–3]. There are also clear clefts that separate the collagen bundles [2,3].

To our knowledge, there have been only ten reported cases of sporadic SF in the oral cavity [7–12]. Herein, we describe the first reported case of sporadic SF presenting on the tongue.

2. Case report

A 69-year-old Caucasian male was referred to the oral and maxillofacial surgery service for evaluation of a 1 cm mass on the right mid-dorsal tongue. The mass was asymptomatic and found during routine clinical examination by the patient's general dentist. The patient was not previously aware of the presence of the lesion or the duration of its presence. The patient reported that there had been no history of trauma to the site.

Examination revealed a palpable, non-tender nodule of the right mid-dorsal tongue. The overlying mucosa was not ulcerated or erythematous. The remainder of the intraoral exam showed normal mucosa with no evidence of papillomatous or papular lesions. The patient's medical history was significant for hypothyroidism, asthma, cardiac arrhythmia and hypercholesterolemia. The patient did not have any signs or symptoms consistent with Cowden syndrome, and he denied any family history of the stigmata found in Cowden syndrome.

An excisional biopsy was performed via infiltrations with a local anesthetic. The mass was completely excised and submitted to the

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Table 1
Antibodies used for immunohistochemistry study.

Antigen	Clone	Dilution	Manufacturer
Vimentin	V9	1:4000	Biogenex
Factor XIIIa	AC-1A1	1:40	Cell Marque
Bcl-2	124	RTU (ready to use)	Dako
CD34	QBend/10	RTU	Dako
α-SMA	1A4	1:1500	Dako
EMA	E29	1:1500	Cell Marque
Factor VIII	Polyclonal	1:2000	Dako
CD68	KPC	1:600	Cell Marque
S-100	Polyclonal	1:2000	Dako
STAT6 from Abcam	YE361, low pH target retrieval	1:200	Dako Flex polymer
CD99 from Dako	12E7, high pH target retrieval	RTU	Dako Flex polymer
β-Catenin from BD Bioscience	14/beta-catenin, low pH target retrieval	1:400	Dako Envision polymer

α-SMA, smooth muscle actin; EMA, epithelial membrane antigen; all antibodies used were monoclonal, except where indicated.

pathology service for gross and histologic evaluation. Primary closure was obtained in a layered fashion. The surgical site healed well and without complication. After an 18-month follow-up, there was no evidence of recurrence.

The tumor consisted of a non-encapsulated, well-demarcated hypocellular rounded eosinophilic nodule centered in the skeletal muscle of the tongue. The collagen fibers were densely hyalinized and showed a vague whorled pattern with numerous prominent clear clefts between the collagen bundles. The tumor cells, though paucicellular, were rather uniform with spindle to vaguely stellate shapes. Bland fusiform nuclei could be observed possessing finely dispersed chromatin with inconspicuous nucleoli. Muscle fibers were entrapped within the dense collagen network and contained foci of degeneration. Vascularity to the lesion was sparse, consisting of only scattered and small vessels.

Immunohistochemical analysis of the tumor cells revealed strongly positive staining for vimentin, and weakly positive staining for bcl-2. Non-specific staining was observed in scattered dendritic spindle cells and histiocytes for Factor XIIIa. A lack of staining of the tumor cells indicated negative expression of CD34, SMA, EMA, Factor VIII, CD68, S-100, STAT6, CD99 and β-catenin (Table 1).

3. Discussion

The features of sclerotic fibromas were first depicted in a patient with Cowden syndrome (multiple hamartoma syndrome) [1]. Cowden syndrome is a rare multisystem disease that affects the skin and mucosal membranes [1,2,4–6]. It is inherited by an autosomal dominant trait [1,4]. The syndrome presents with oral and skin manifestations [1,2,4–6], as well as benign neoplasia of the internal organs with potential transformation to malignancy [1,5,6]. Malignant changes may develop in the thyroid [1,4–6], breast [1,4–6] and colon [1,5]. Abnormalities of the genitourinary system [1,5,6] and nervous system [1,6] may be found, along with a diverse involvement of other organ systems [1,4–6]. The cutaneous findings may include trichilemmomas [2], acral keratoses [1] and tumors of the skin, such as SF [2]. Intraoral manifestations may include papillomatous lesions [1,4–6], a high arched palate [1,5,6], a fissured tongue [1,4–6] and maxillary and mandibular hypoplasia [6]. The typical papillomatous lesions of the oral mucosa are mostly consistent with fibrous or epithelial hyperplasia [4–6]. Although Weary et al. [1] first described the SF lesion in a patient with Cowden syndrome intraorally, this is an extremely rare finding. The patient with Cowden syndrome described had the presentation on the tongue. SF in Cowden syndrome may present as nodules on the skin and may be a cutaneous indicator of Cowden syndrome [2].

Rapini and Golitz [3] first designated the term sclerotic fibroma after describing the histology of 11 cases of solitary skin lesions. These lesions were derived from patients who did not have Cowden syndrome, but had the same histologic presentation of the unique fibrous lesions found in Cowden syndrome [3]. The histological features described in the 11 cases showed sharp demarcations, hypocellularity and thick collagen bundles. These collagen bundles presented in an atypical pattern, were hyalinized, sclerotic and separated by clefts [3]. This description is consistent with nodules found in Cowden syndrome that are non-encapsulated, well-demarcated [2] and paucicellular with collagen fibers presenting in a whorled pattern [1,2].

In addition to the findings of solitary SF cutaneous lesions not associated with Cowden syndrome, such as those described by Rapini and Golitz [3], there have been reported cases of sporadic sclerotic fibromas in the oral cavity [7–12]. Ten cases of sporadic SF in the oral cavity have been reported [7–12]. This is the first reported case of sporadic SF with presentation on the tongue. Alawi and Freedman [7] first described sporadic SF in the oral cavity in a series of five cases. Four of the cases of this study were in the buccal mucosa and one presented on the lower lip. Lombardi et al. [8,9] reported two cases of sporadic SF. One SF occurred in the retromolar pad and the second in the lower lip. Ide et al. [10] reported a case of sporadic SF in the lower lip. Gonzalez-Vela et al. [11] described a case of sporadic SF on the buccal mucosa [11], and Lee et al. [12] also described a sporadic SF in the buccal mucosa. All of the solitary lesions shared the distinct histological features of SF [7–12].

In the current case, the entire lesion was available for examination, revealing the diagnostic H&E histological staining for features of SF. There was a sharply circumscribed hypocellular lesion with hyalinized collagen fibers, clefts and bland spindle cells characteristic of sclerotic fibroma. This distinct finding separates it from the other oral fibrous tumors usually considered in the differential diagnosis (Fig. 1). Immunohistochemical results revealed the tumor cells were positive for vimentin and bcl-2 (weak) and showed non-specific staining for Factor XIIIa (Fig. 2). The tumor cells were negative for CD34, SMA, EMA, Factor VIII, CD68, S-100 expression, STAT6, CD99 and β-catenin (Table 1). The positivity for vimentin resembles findings of other reported cases [7–12]. Factor XIIIa is reportedly found [7–10,13], and may be positive in a portion of cells [8,9] or in scattered mononuclear cells as well [7]. In this case, Factor XIIIa was considered non-specific with regard to the staining of scattered dendritic spindle cells and mononuclear cells for Factor XIIIa. Although this case did not present with either CD34 or CD99 positivity, in SFs, there are occasional positive findings for CD34 [7,9–13] and CD99 [11,12].

The differential diagnosis is inclusive of several types of histologic lesions. Non-fibrous and fibrous lesions will be categorized and discussed. Non-fibrous lesions to be considered include granular cell tumors, neurofibromas and schwannomas (neurilemmomas). Granular cell tumors are benign lesions often found on the tongue [14]. Histologically, they are composed of polygonal cells that are large with granular cytoplasm [14]. Neurofibromas