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Photo vignette

Oral focal mucinosis: review of the literature and two case reports

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Abstract

Oral focal mucinosis is a rare condition, clinically characterized by an asymptomatic swelling, without distinct, specific features, which occurs predominantly in adults of the female gender. Its clinical aspect leads to various differential diagnoses, and final diagnosis is only possible by means of histopathological exam, in which a well-delimited myxomatous area containing mucinous material is observed. In the present study, a review of the English-language literature about the lesion, was conducted, covering the period from 1974 to March 2015. We report two new cases, thereby contributing to the knowledge and differential diagnosis of this entity.

Keywords: oral focal mucinosis; myxoid lesions; oral cavity

Introduction

Oral Focal Mucinosis (OFM) was described for the first time in 1974, in a report of 8 cases, when it was suggested that this is the oral lesion corresponding to cutaneous focal mucinosis [1]. The lesion is clinically characterized by an asymptomatic swelling and histologically, as a well-delimited myxomatous area containing mucinous material, circumscribed by collagen fibers [2]. It is a rare condition occurring predominantly in adults and has a predilection for the female gender. There is a certain degree of difficulty with its diagnosis, because OFM has no distinct specific features [3].

In view of the scarcity of cases of OFM, the intention of the present study was to review the literature, and add two new cases, thereby contributing to the differential diagnosis of this entity.

Case synopsis

Case 1

The patient, a 35-year-old woman, complained of a painless swelling in the gingiva, without bleeding, noted four months previously. On intraoral clinical exam, a nodule was observed in the vestibular gingiva of the right mandibular second molar, with a sessile base, firm on palpation, without change in color and texture of the mucosa, measuring approximately 1.0 cm in diameter (Figure 1). With the hypothesis of irritation fibroma, lipoma or peripheral ossifying fibroma an excisional biopsy was performed and the material was sent for histopathological examination.

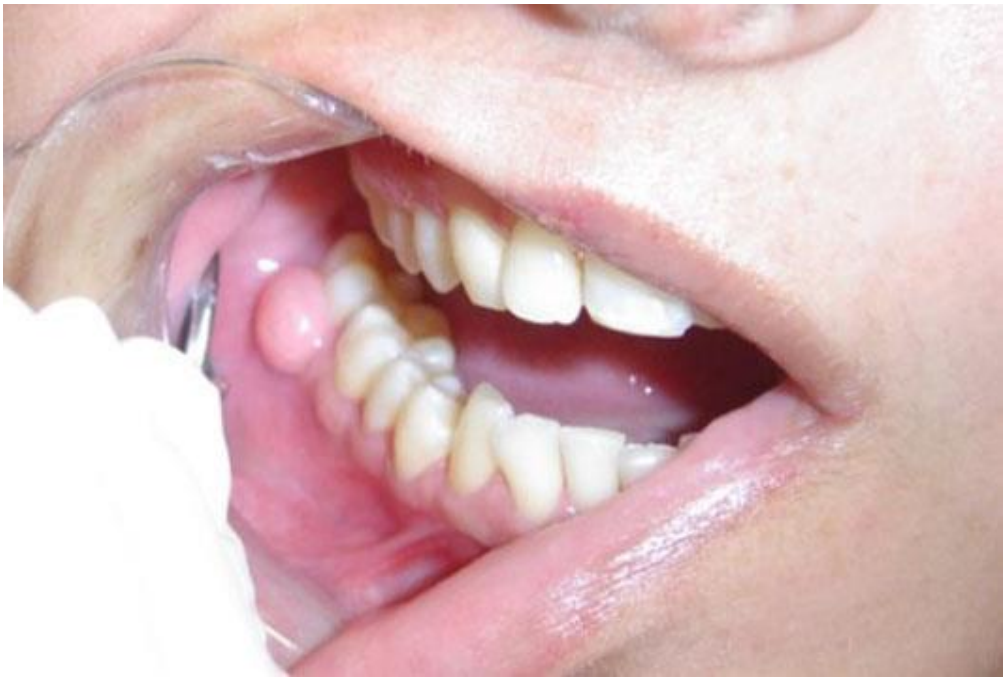

Figure 1: Clinical image - Case 1. The nodular lesion is noted in the buccal gingiva of the right mandibular second molar, without change in mucosal color and texture.

Microscopic examination revealed fragments of mucosa with an atrophic, parakeratinized, squamous epithelium and a well delimited, but not encapsulated area of loose myxomatous connective tissue in the lamina propria, surrounded by fibrous connective tissue (Figure 2). The fibroblasts in the myxomatous area were oval, fusiform- or star-shaped, interlaced with delicate fibers. In the adjacent connective tissue, there were dilated blood vessels and moderate perivascular and sub-epithelial mononuclear inflammatory infiltrates.

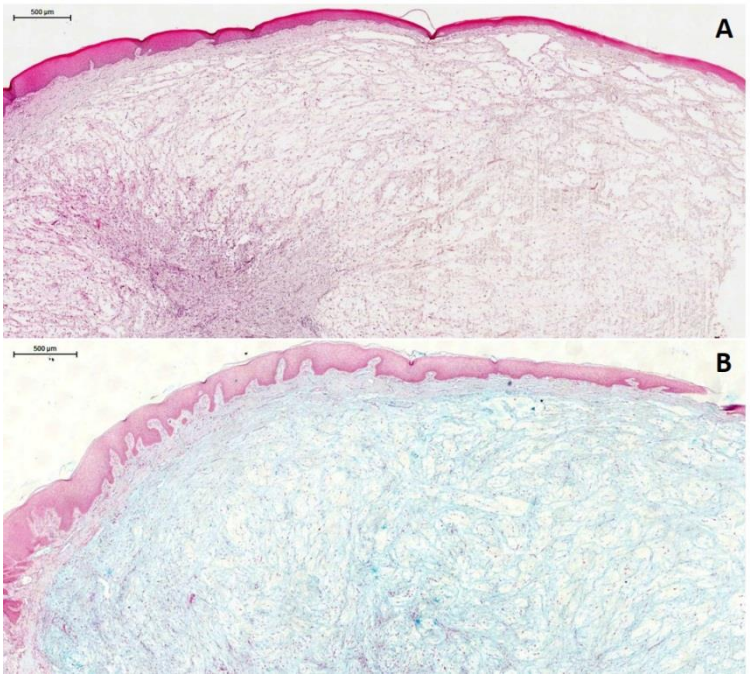

Figure 2: Photomicrograph (H&E-A) in which loose connective tissue with a myxomatous aspect is observed, positive for Alcian Blue staining (B), 100x.

Case 2

The patient, a 35-year-old man, noted a gingival swelling, without painful symptoms or local bleeding, after implant placement surgery to replace the left central incisor, four years previously. On clinical examination, a nodule with whitened color, measuring approximately 0.5 cm, was seen in the alveolar ridge mucosa (Figure 3). The clinical diagnosis hypotheses were fibrous hyperplasia and fibroma.

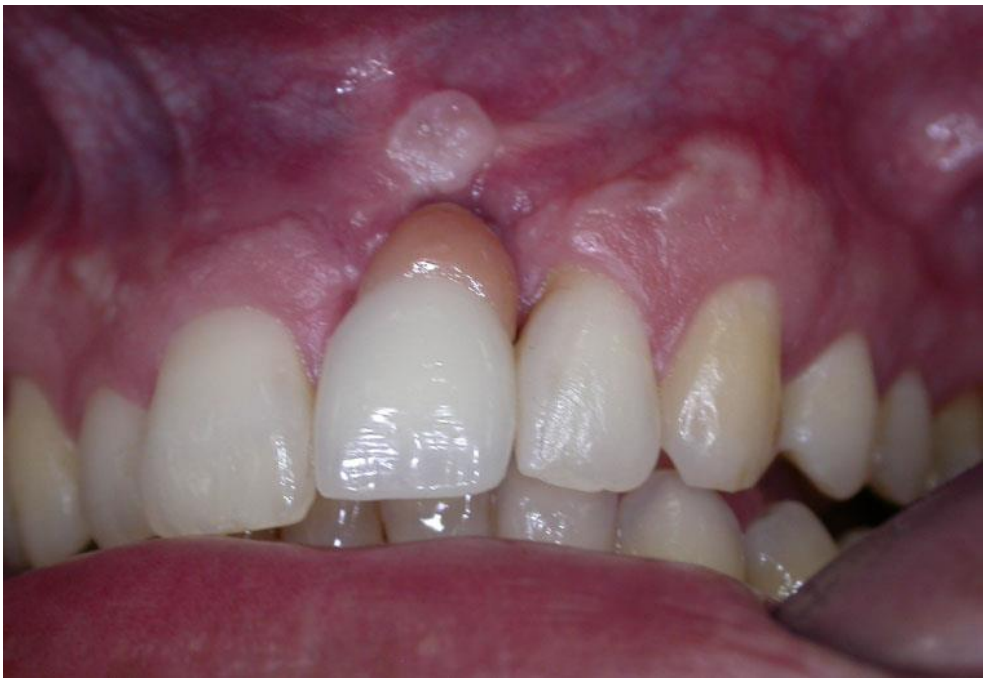

Figure 3: Clinical image - Case 2. A nodule with whitened color, measuring approximately 0.5 cm, is noted in the alveolar ridge mucosa.

After excisional biopsy, microscopic characteristics similar to those described in Case 1 were observed, with a myxomatous connective tissue area containing star-shaped fibroblasts, surrounded by fibrous connective tissue, well delimited, however, not encapsulated (Figure 4).

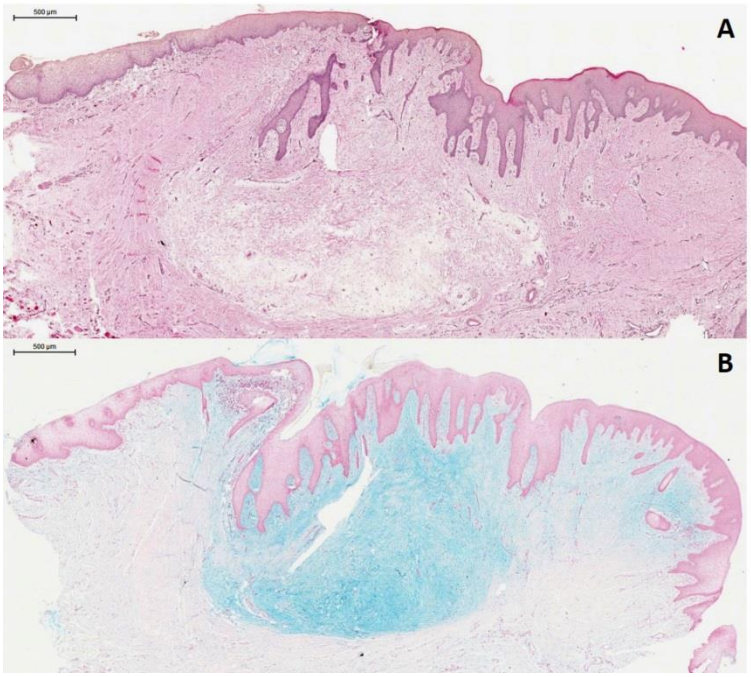

Figure 4: Photomicrograph (H&E-A) in which myxomatous tissue is observed, shown by Alcian Blue staining (B), limited by fibrous connective tissue, 100x.

The cases reported were diagnosed as OFM. In addition HE staining, Alcian Blue (pH 2.5) staining was performed, which showed a mucoid substance in both cases. The patients were followed up, without history of recurrence. The cases reported were the only cases of OFM found in our service in the period from 1962 to 2015 (12,722 cases), representing 0.015% of the diagnosed lesions.

Results

A search was conducted in the PubMed, Scopus and Web of Science databases, by using the following keywords: oral focal mucinosis and myxomatous lesions, evaluating the articles published in the English language from 1974 up to March, 2015.

Twenty-one articles were selected, totaling 65 cases of OFM (Table 1). Of these cases, 63.07% occurred in female patients; 67.69% were located in the gingiva; 13.84% in the palate; 9.23% in the tongue; 6.15% in the buccal mucosa and 3.07% in the lip. The mean age of patients was 39.29 years (standard deviation: 15.49). The most prevalent clinical diagnoses were fibroma and fibrous epulis, in 31.42% and 14.28%, respectively, of the cases in which this information was present. The following diagnoses were also found: mucus extravasation phenomenon (mucocele), ossifying fibroma, papilloma, periodontal abscess and mucoepidermoid carcinoma. The duration of the lesions ranged between 4 months and 4 years. In our cases, the lesions were treated by simple surgical excision, without recurrence, which was also observed in the literature review, with the exception of one case located in the gingiva in a 27-year-old woman [7].

Table 1. Clinical cases of oral focal mucinosis related in the literature in the period between 1974 to March, 2015

Author	Age/Gender	Localization	Duration	Clinical Diagnoses
Tomich, 1974 [1]	40/F	Palate	5-10 years	Fibroma
	31/F	Gingiva	1 year	NA
	16/M	Gingiva	NA	Fibroma
	-/F	Buccal mucosa	1 year	Papilloma
	45/M	Tongue	2 months	Mucocele
	28/M	Alveolar mucosa	NA	Fibroma
	22/F	Hard palate	4 months	Mucocele
	19/F	Hard palate	4 months	Fibroma
Saito et al, 1985 [6]	35/M	Gingiva	3 months	Fibroma
	50/F	Gingiva	NA	Fibroma
Gnepp et al, 1989 [4]	4/F	Hard palate	NA	NA
Buchner et al, 1990 [2]	30/M	Gingiva	5 years	NA
	32/F	Gingiva	1 month	NA
	22/F	Gingiva	1 month	NA
	53/F	Gingiva	NA	NA
	16/F	Gingiva	NA	NA
	43/M	Gingiva	NA	NA
	61/F	Alveolar mucosa	NA	NA
	37/F	Alveolar mucosa	NA	NA
	41/F	Gingiva	NA	NA
	37/F	Gingiva	3 years	NA
	46/M	Gingiva	1 year	NA
	38/F	Hard palate	1 year	NA
	46/M	Gingiva	3 years	NA
	50/M	Tongue	2 months	NA
Soda et al, 1998 [11]	68/M	Tongue	3 years	NA
Iezzi et al, 2001 [13]	48/M	Gingiva	8 months	Periodontal abscess
Aldred et al, 2003 [14]	38/F	Lip	NA	NA
	30/F	Gingiva	1 month	Fibrous epulis
	16/F	Gingiva	4 months	Fibrous hyperplasia
	56/F	Buccal mucosa	NA	NA
	60/F	Mouth	>1 years	Squamous papilloma
	49/M	Gingiva	10 years	Polyp
	31/F	Gingiva	6 months	Giant cell granuloma
	52/M	Gingiva	1 year	Fibrous epulis
	74/M	Lip	NA	NA
	40/F	Gingiva	4 months	Fibroma
	55/M	Tongue	3 months	Fibroepithelial polyp
	37/F	Gingiva	3 months	NA
	35/F	Gingiva	1 year	Fibrous epulis
	33/F	Gingiva	1 year	Fibrous epulis
	68/M	Gingiva	1 year	Fibroepithelial polyp
Talacko et al, 2004 [15]	63/F	Buccal mucosa	NA	Sinus or fistula
	24/M	Gingiva	NA	NA
Germano et al, 2008 [16]	35/M	Gingiva	NA	NA
Soares de Lima et al, 2008 [3]	36/F	Gingiva	4 months	Fibroma, POF, gingiva hyperplasia
Narayana et al, 2008 [7]	37/F	Gingiva	NA	Fibroma, POF
	54/F	Gingiva	NA	POF
	49/M	Hard palate	NA	Mucoepidermoid

				carcinoma, Pleomorphic adenoma
	27/F	Gingiva	NA	Fibroma, cyst
	26/M	Gingiva	NA	Fibroma
	32/F	Gingiva	NA	Gingival cyst
	48/F	Gingiva	NA	Gingival cyst
Gabay et al, 2010 [9]	44/F	Gingiva	3 years	POF, fibrous hiperplasia, GCG
Madhusudhan et al, 2010 [17]	50/M	Gingiva	2 months	Gingival epulis
	26/F	Gingiva	3 months	Fibrous epulis
Pacifici et al, 2012 [12]	62/F	Tongue	NA	NA
Lee et al, 2012 [5]	17/F	Gingiva	NA	NA
Bharti et al, 2012 [8]	32/F	Palate	4-5 months	Fibrous Hyperplasia
Ena et al, 2013 [18]	26/M	Gingiva	1 year	NA
	36/F	Gingiva	NA	NA
Tekkesin et al, 2013 [19]	19/M	Palate	NA	NA
Neto et al, 2014 [20]	20/F	Gingiva	NA	NA
Woo et al, 2015 [21]	2/F	Palate	3 months	Palatal exostosis, POF, fibrous hyperplasia, pleomorphic adenoma, lymphoma, Langerhans cell histiocytosis
Sowmya et al, 2015 [22]	54/M	Gingiva	NA	NA
Case 1	35/F	Gingiva	4 months	POF, lipoma, fibroma
Case 2	35/M	Alveolar ridge	4 years	Fibrous hyperplasia, fibroma

(F= female, M= male, NA= not available, POF= peripheral ossifying fibroma, GCG=giant cell granuloma).

Discussion

OFM affects adult women, in the majority of cases. However, some authors have shown their occurrence in younger patients as well. Gnepp et al. [4] reported the case of a 4 year-old girl, who was submitted to surgery for correction of cleft lip and palate and during the course of surgery, a nodule was found in the palate. This was diagnosed as OFM after biopsy and there was no recurrence. Lee et al. [5] reported the case of a 17-year-old girl with swelling in the region of the incisive papilla.

The majority of lesions were described as asymptomatic nodules with a color similar to that of the adjacent mucosa, measuring up to 2 cm in the largest dimension [6]. On the other hand, Talacko et al. [15] reported two cases in which there was ulceration on the surface of the OFM, resulting from occlusal trauma.

From the clinical point of view, it is not possible to distinguish OFM from other lesions. Their characteristics lead to differential diagnoses such as fibroma, fibrous hyperplasia and pyogenic granuloma, prevalent among the cases reviewed in the literature. It is also possible for OFM to resemble peripheral ossifying fibroma, peripheral ameloblastoma and minor salivary gland tumors [3, 7, 8]. No case was observed to be clinically diagnosed as OFM, and the basis for final diagnosis was the histopathological exam.

The etiology of OFM remains unknown, however, Gabay et al. [9] raised the hypothesis of association with trauma, on reporting a case of the lesion located in the attached gingiva over the root of a tooth with external resorption in its cervical third. The thinking about the pathogenesis remains the same as was suggested by Tomich in 1974 [1]: an overproduction of hyaluronic acid by fibroblasts may lead to the development and growth of the lesion [2].

In the microscopic exam of OFM, oval, fusiform- and star-shaped fibroblasts were found in the myxomatous area, which may sometimes cause atrophy of the epithelium. Few capillaries are found in the lesion, generally present in the collagenous tissue around the myxomatous area, which sometimes presents perivascular inflammatory infiltrate [2, 3]. Microscopic examination of the two cases reported here revealed a relatively delimited, but non-encapsulated area, consisting of connective tissue with a myxomatous aspect, exhibiting a large amount of extracellular matrix permeating fibroblasts that showed a fusiform or a star-shaped appearance. There was a scarcity of vessels present in the myxomatous region in comparison with the quantity in the peripheral region. The inflammatory infiltrate was predominantly mild.

Histologically, among the differential diagnoses of OFM, the following can be included: odontogenic myxoma, mucocele or mucus extravasation phenomenon, nerve sheath myxoma or neurothekeoma, neurofibroma with myxomatous regions and plexiform neurofibroma. We are able to distinguish odontogenic myxoma based on the histological, clinical and radiographic characteristics. Differentiation is simple, since this lesion infiltrates into the surrounding connective tissue, instead of being well delimited, and has a remarkable presence of reticular fibers [2, 11]. Mucocele is a common lesion of minor salivary gland origin, differentiated from OFM due to the presence of granulation tissue [11].The mucus extravasation phenomenon is described by spilled mucin surrounded by a granulation tissue, with histiocytic cells usually included in the mucoid material. These findings are not present in lesions diagnosed as focal oral mucinosis [11, 23], and were not found in our cases either. Moreover, minor salivary glands are present throughout the oral cavity, except for the anterior region of hard palate and gingiva/alveolar ridge mucosa [24], where the presented lesions were found. Therefore, the location is also important in the differential diagnosis.

Nerve sheath myxoma or neurothekeoma has abundant alcianophilic content, numerous mast cells, cells positive for S-100 protein and lobular aspect, differing from the lesion in question. In the same way, it is possible to distinguish OFM from neurofibroma with myxomatous areas [1, 12]. Therefore, a routine histological analysis may not always establish the diagnosis of these lesions, due to the similarities among them [10], and complementary histochemical or immunohistochemical analysis is necessary in some cases.

In spite of its rarity, pathologists and clinicians must include OFM in differential diagnoses of lesions in the gingiva and palate, the locations of their most frequent occurrence. Owing to their nonspecific characteristics, clinical diagnosis of OFM is extremely difficult, emphasizing the need for performing biopsy in order to conduct the case in the most adequate manner.

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