Oral focal mucinosis of the tongue: A rare clinical entity?

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A R T I C L E   I N F O

Article history:
Received 1 January 2015
Received in revised form 3 November 2016
Accepted 21 January 2017
Available online 23 January 2017

Keywords:
Oral focal mucinosis
Myxoid lesions
Oral pathology

A B S T R A C T

Oral focal mucinosis (OFM) is regarded as a very rare clinical entity with only 62 cases previously reported in the literature. The majority of these cases have been present on gingiva and only 3 cases have been diagnosed on the tongue. The clinical appearance of OFM is relatively non-specific and resembles other and more prevalent conditions like fibroepithelial hyperplasia or other reactive mucosal lesions. The present case report describes OFM in the tongue in an 88-year-old woman. The report addresses the diagnostic considerations of OFM, but also if the lesion is really as rare as the literature indicates.

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1. Introduction

The term cutaneous focal mucinosis (CFM) was coined by Johnson & Helwig in 1966 to describe asymptomatic, white or flesh-colored white papules or nodules on the face, trunk, or extremities [10]. Histopathology showed an increased production of hyaluronic acid with a decreased presence or absence of reticulum and/or collagen. The replacement of collagen and reticular fibers in favor of hyaluronic acid resulted in the formation of small cysts. Johnson & Helwig had previously in 1965 described an entity which they referred to as cutaneous myxoid cyst (CFC) in 40 patients, where lesions appeared on the fingers and toes [9]. Despite a very similar histopathological appearance they chose to regard them as separate entities because of their different anatomical locations.

Oral involvement is rare. Tomich reported 8 cases of oral focal mucinosis (OFM) in a retrospective study in 1974 [21]. The author identified cases that had been diagnosed as soft-tissue myxomas, fibroma or fibrous hyperplasia with myxomatous degeneration. Eight cases were identified which met the diagnostic criteria for CFM defined by Johnson & Helwig. Three lesions were found in the palate, three on gingiva, one in alveolar mucosa and one on the tip of the tongue. These 8 cases were subsequently analyzed by Johnson & Helwig who agreed that the histopathological appearance of the oral lesions “resembled the myxoid cyst as well as the cutaneous focal mucinosis” and suggested that CFM, CFC and OFM were basically the same lesion, but with different anatomical locations [21]. Additional cases have since then been reported [1,3,6,8,15,18,19] and a review on oral focal mucinosis published in 2008 [17] described a total of 48 cases. Since then 14 new cases have been published [2,4,5,11-13,20], giving a total of 62 known cases of OFM.

The most common location for OFM is the gingiva, comprising more than half of the reported cases. Involvement of the tongue is rare with only 3 cases previously reported in the literature [3]; [18,21]. The present case describes a fourth case of oral focal mucinosis of the tongue.

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http://dx.doi.org/10.1016/j.omsc.2017.01.006
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2. Case presentation

An 88-year-old woman was referred for examination of a nodular lesion on the dorsum of the tongue. A tentative diagnosis of focal epithelial hyperplasia (FEH) was suggested by the referring dentist. The patient had recently changed to a new dentist and was not aware of the lesion herself. It could therefore not be established how long the lesion had been present.

Her medical history revealed thyroiditis, hypertension, bradycardia, angina pectoris and pain from the neck. She was medicated with levothyroxine, hydrochlorothiazide, acetylsalicylic acid, felodipine and paracetamol. She had no allergies and was a non-smoker.

Fig. 1. Clinical appearance of a nodular lesion on the dorsum of the tongue. The lesion was solid on palpation and appeared to extend down to the underlying muscle.

Fig. 2. Overview of the tissue specimen. The lesion was well demarcated and extended down to the underlying muscle (a). H&E stain. Magnification × 20. b shows close-up of the lesion in a with loosely connected myxomatous connective tissue and slightly enlarged oval fibroblasts. H&E stain. Magnification × 100.
The clinical examination revealed a broad-based nodular lesion on the dorsum of the tongue of approximately 1 cm in diameter (Fig. 1). The surface was smooth and no inflammatory reaction could be seen in the adjacent tissue. The lesion was solid on palpation and appeared to extend down to the underlying muscle. The clinical appearance was therefore not consistent with a common fibro-epithelial hyperplasia or FEH. The lesion was surgically excised to include a portion of the superficial tongue musculature.

3. Histopathology

The histopathological examination showed a relatively well demarcated area (Fig. 2a) with a loosely connected myxomatous connective tissue with slightly enlarged oval fibroblasts. The lesion was lobulated, and circumscribed, but unencapsulated. The superficial portions of the lesion attenuate overlying rete ridges of the squamous epithelium, and deeply, the lesion involved and displaced skeletal muscle bundles (Fig. 2b). Aside from some blunted rete ridges in the overlying surface epithelium, the squamous epithelium was without significant alteration. The histopathological image was consistent with oral focal mucinosis.

4. Discussion

Oral focal mucinosis (OFM) is regarded as a very rare lesion with only 62 published cases. To the best of our knowledge, there are only 3 previously reported cases of OFM in the tongue and the majority of OFM lesions are found on gingiva. However, the question is if OFM is as rare as the literature indicates. First, the clinical appearance of OFM is rather non-specific. Most of OFM lesions appear as singular nodules and may clinically resemble other diagnostic entities. Published OFM cases were usually given provisional diagnoses such as fibroma, fibrous epulis, fibrous hyperplasia, peripheral giant-cell granuloma, pyogenic granuloma, periodontal abscess or fibroepithelial polyp. In fact, a tentative diagnosis of OFM was not given in any of the cases reported in the literature. The diagnosis can only be made through biopsy and subsequent histopathological examination. Consequently, if the lesion is clinically regarded as harmless and not removed for histopathological examination, it may be diagnosed as some other, and more prevalent, mucosal lesion.

Secondly, we examined the number of cases in our own archive and found that 52 cases had been signed out as OFM since 1988. Out of our 52 cases, 29 occurred in female and 23 in male patients. The different locations were 33 from the gingiva, 9 from the hard palate, 3 from vestibulum, 1 from the floor of the mouth, 1 from the buccal mucosa and the present case from the dorsum of the tongue. This is close to the total number of cases reported in the literature. There are most likely a considerable amount of OFM cases in pathology department archives throughout the world, they are just not reported in the scientific literature.
Thirdly, there are differential histologic diagnoses like neural tumours with myxoid degeneration, neurofibroma, nerve sheath myxoma, mucocoele and odontogenic myxoma to consider, but also myxoid perineuroma and myxoid neurofibroma lesions that could involve skeletal muscle. To differentiate from some of these lesions, immunohistochemical stains for S-100 protein and EMA (epithelial membrane antigen) were made, and since there was no positive staining for any of these antibodies (Fig. 3a and b), we could exclude myxoid perineuroma (EMA+) and nerve sheath myxoma (S-100+) [14]. We further excluded odontogenic myxoma, because it is predominantly found in young adults and involve the bone [16]. All OFM cases reported by Tomich, had previously been signed out as other diagnoses [21]. If the same diagnostic considerations can be applied on other pathologists, the true number of OFM cases may be even higher.

To conclude, it is tempting to speculate that OFM is a relatively rare condition, but not as rare as we may previously have thought.

Conflict of interest statement

There are no actual or potential conflicts of interest including any financial, personal or other relationships with other people or organizations within three years of beginning the submitted work that could inappropriately influence, or be perceived to influence, this paper.

Funding

There is no funding involved in this manuscript.

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