CASE REPORT

Oral Focal Mucinosis of the Tongue

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Oral focal mucinosis (OFM) is an uncommon clinicopathological entity which is considered to be the oral counterpart of cutaneous focal mucinosis and cutaneous myxoid cyst. It is comprised of a clinically elevated mass with a histological feature of localised areas of myxomatous connective tissue. The present study adds a rare case of OFM of the tongue to the literature, and we present a review of the most characteristic oral myxomatous lesions. (Pathology Oncology Research Vol 4, No 4, 304–307, 1998)

Key words: focal mucinosis, oral

Introduction

Oral focal mucinosis (OFM) was first described in 1974 by Tomich.[24] He reported 8 cases of the oral counterpart of cutaneous focal mucinosis and cutaneous myxoid cyst and presented the clinicopathological and histochemical features of this new entity. Clinically the lesions appeared as asymptomatic round elevations. Histologically, tissue sections showed a localized area characterized by an accumulation of hyaluronic acid in the connective tissue surrounded by a relatively dense but normal collagenous fibrous connective tissue. Stellate fibroblasts and capillaries were widely separated by myxomatous tissue.[22] Since the original paper by Tomich, only two additional reports describing 2 cases[19] and 15 cases[1] of OFM have appeared in the literature. To our knowledge only two cases of OFM of the tongue have been previously reported. In this paper we report another case of OFM of the tongue, and we present a review of the most characteristic oral myxomatous lesions.

Case Report

Clinical findings

A 68-year-old male, presented a lesion which was located on the anterior ventral tongue and which he had for three years. The clinicians described it as whitish in color, asymptomatic soft swelling. The lesion was treated by simple excision.

Pathologic features

Gross – The lesion presented as a small, sessile, smooth-surfaced, rubbery, elevated mass. On cut section it had a glistening gray-white appearance, and was 0,5 cm in diameter.

Light microscopy – Specimen was fixed in 10% buffered formalin and embedded in paraffin. Sections were cut to 3 μm in thickness and stained with haematoxylin and eosin (HE). The sections used for immunohistochemical staining with monoclonal antibodies to S-100 protein, desmin and vimentin, were deparaffinised and incubated overnight at 4°C with the primary antibody. An avidin-streptavidin complex staining kit (LSAB K680 DAKO) was used according to the manufacturer's instructions. Peroxidase activity was visualized with 3-amino-9-ethilcarbazole (AEC) and sections were weakly counterstained with Mayer haematoxylin. For negative control the sections were incubated with non-immune serum instead of the primary antibody. The following histochemical methods were employed on additional tissue sections: Gomori's method for reticulin, the PAS and alcian blue (at pH 2,5) methods for mucosubstances, and the toluidine blue method for metachromasia at pH 3.

On the HE staining, the histologic features of the lesion consisted, of a fairly well localized area of myxomatous connective tissue. The border zone was only partially well delineated and there were areas of gradual transition from the myxomatous tissue to the surrounding relatively dense but normal collagenous fibrous connective tissue *(Figure*

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Figure 1. Photomicrograph showing the demarcated interface between the lesion and the connective tissue. (HE, x200)

1). Moreover, the myxomatous zone extended to the overlying epithelium and produced definite flattening of the rete ridges. The capillaries and small blood vessels present in the myxomatous area appeared to be relatively few compared to these in the adjacent connective tissue. The myxomatous tissue consisted of collagen fibers separated by homogeneous mucinous material with scattered ovoid, fusiform, or stellate fibroblasts *(Figure 2)*. Delicate fibrillar processes extended from the fibroblast cytoplasms into the surrounding matrix. Cleft-like spaces or small cavities (pools) were present within the mucinous material. The result obtained with alcian blue staining at pH 2,5 was positive, indicating the presence of acid sulfated mucosubstances. Alcianophilia was not observed after treatment with hyaluronidase. PAS reaction was negative and metachromasia was observed after staining with toluidine blue at pH 3. Reticulin fibers were not found within the myxomatous areas except around the blood vessels; instead, these fibers were visible within the surrounding connective tissue. The cellular component of the lesion investigated by immunohistochemical staining showed a negative reaction for S-100 protein and desmin antibodies, whereas it was positive for vimentin antibody.

Discussion

Oral lesions of myxomatous nature are relatively rare. Such lesions include OFM, soft tissue myxoma, nerve sheath myxomas, odontogenic myxoma, myxomatous change in fibrous lesions, neurofibroma with myxomatous change, oral mucous escape reaction or mucocele, and the recently described ectomesenchymal chondromyxoid tumor of the anterior tongue. OFM was first described in 1974 by Tomich. Since the original paper by Tomich only two additional reports by Buchner and Saito describing respectively 15 and 2 cases of OFM have appeared in the literature but in all papers only 2 cases were localized in the tongue. In 1965, Johnson et al[9] called attention to the cutaneous myxoid cyst which occurred on the dorsal aspect of the fingers. Successively, in 1966, Johnson and Helwig[10] described the cutaneous focal mucinosis which occurred on the face, trunk, or extremities and in 1974, Tomich described the oral counterpart of the cutaneous myxoid cyst and cutaneous focal mucinosis. (Histochemistry was used to detected veticular filters and to determine the nature of the mucinous material. The latter proved to be hyaluronic acid.) On the basis of the histological and histochemical features, Tomich, suggested that all three lesions are identical and that they differed only in their anatomic location. Further, he proposed that the term focal mucinosis, rather than myxoid cyst, be applied to these lesions which developed as a result of fibroblastic overproduction of hyaluronic acid due to unknown stimulus. Combining all cases reported in the literature, we have revealed that the most common location of OFM is the gingiva and it si very ravely located in the tongue. The main histological differential diagnosis of OFM is soft tissue myxoma. The soft tissue myxoma is a true neoplasm

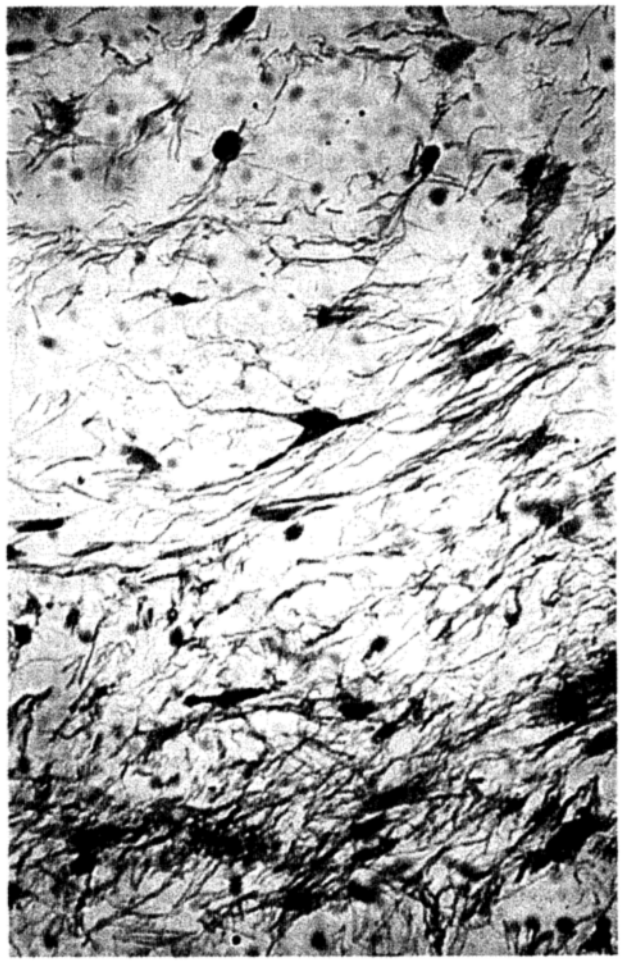

Figure 2. Photomicrograph showing delicate fibrillar processes extending from fibroblast cytoplasm. (HE, x400)

of primitive mesenchyme which, excluding the cardiac myxoma and the odontogenic myxoma of the jaws, occurs most often in the large muscles of the shoulder and thigh. Intraoral soft tissue myxoma are rare, and their existence has been debated.[1,21,24] However, myxomas have been reported in many oral locations, although the palate is most frequently affected. Mockli et al[13] reported one case of intramuscular myxoma of the tongue diagnosed on the basis of fine needle aspiration cytology. Intraoral soft tissue myxoma have been reported in an autosomal dominantly inherited syndrome consisting of myxomas, spotty mucocutaneous pigmentation (similar to Peutz-Jeghers syndrome), and endocrine abnormalities ("Carney's complex").[2,4,16,18] Oral intramuscular myxomas, including myxomas of the tongue, have been reported in association with this syndrome.[18] Oral myxomas are not encapsulated and may exhibit infiltration into surrounding soft tissue. Disperse stellate and spindle-shaped fibroblasts are found in a loose myxoid stroma with a network of delicate reticular fibers. Although there are major similarities between OFM and myxoma, there are also some differences. Myxoma usually shows an extensive network of reticular fibers, whereas little reticulum is present in OFM. Myxoma exhibits infiltrative growth pattern while OFM usually manifests as a localized area of myxomatous connective tissue. Cleft-like spaces or small pools of mucinous material are not present in myxomas but are a feature in many cases of OFM. The myxoma of oral soft tissue is extremely rare[20,26] and Shafer et al[20] state that most lesions classified as such actually represent myxomatous change in a pre-existing fibrous lesion. Elzay and Dutz[5] who reviewed the literature concerning oral soft tissue myxoma claimed that some cases which were reported under the term oral myxoma[15,23,25] are in reality examples of OFM. Odontogenic myxoma involving the jaws can be distinguished by its histologic features when combined with its clinical and radiographic features. Myxomatous change in pre-existing fibrous lesions is a well-recognized phenomenon, but is not common. OFM can be distinguished from this lesion rather easily, in as much as the latter blends into the surrounding connective tissue rather than being more sharply defined. In addition mononuclear inflammatory cells, as well as a prominent vascular component, are almost invariably present in areas of myxomatous change. The oral mucous escape reaction or mucocele is one of the most common soft tissue lesions of the oral soft tissue. It is a granulation tissue lined pseudocyst that results from spillage of salivary mucin into connective tissue from a traumatically severed minor salivary gland excretory duct. In the tongue, it arises from the glands of Blandin-Nuhn on the ventral surface.[17] It is easily separated from OFM by the presence of the granulation tissue and the mucoid material which contain numerous histiocytic cells with active phagocytosis. Neither reticulin fibers nor hyaluronic acid are present within the mucus filled space. Nerve sheath myxoma was originally described in 1969 by Harkin and Reed[8] as a rare neoplasm characterized by a lobular proliferation of myxomatous tissue separated by fibrous septa. This neoplasm has been observed in the tongue.[12] Neurothekoma was subsequently reported in 1980 and shared histologic features with the nerve sheath myxoma.[7] Most investigators now accept these two lesions as synonymous.[6,11] The nerve sheath myxoma can be identified by the condensed connective tissue around the periphery of the myxomatous area. In addition, this lesion, often demonstrates a lobular pattern[8,12] a feature not observed in OFM. The nerve sheath myxoma contains abundant alcianophilic material and numerous mast cells that were totally lacking in the OFM. Neurofibroma with myxomatous change has been well documented in the oral cavity.[3,8,14] This lesion is histologically and histochemically very similar to the nerve sheath myxoma.[24] Therefore, it is distinguished from OFM in a like manner. Finally the new clinicopathologic entity termed ectomesenchymal chondromyxoid tumor (ECT) of the anterior tongue[22] is well separated from OFM because the latter does not have the lobular architecture, histochemical, or immunohistochemical profile of ECT.

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