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ORAL FOCAL MUCINOSIS- A LITERATURE REVIEW

Dental Science

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ABSTRACT

Oral focal mucinosis is an uncommon soft-tissue lesion of unknown cause which was first described by Tomich in 1974. Oral focal mucinosis (OFM) is a rare disease of unknown aetiology, and most commonly occurs in adult women. Commonly seen in adults during the fourth and fifth decade of life. Oral focal mucinosis appear as a nodular lesion covered with mucosa which is of a round or oval shape, pink white in color, with no clinical signs and shows predilection for the mucosa overlying bone, and keratinized mucosa is almost involved exclusively. The pathogenesis of OFM is suggestive of overproduction of hyaluronic acid by fibroblasts at the expense of collagen production. Histopathologically OFM is differentially diagnosed as myxoid nerve sheath tumor & odontogenic myxoma based on special stains & IHC markers OFM is diagnosed.

KEYWORDS

soft-tissue lesion, oral focal mucinosis, hyaluronic acid.

INTRODUCTION:

OFM is a rare disease where the connective tissue undergoes a focal myxoid degeneration(1). Oral focal mucinosis is an uncommon soft-tissue lesion of unknown cause which was first described by Tomich in 1974. (2)(3)The oral counterpart of dermal lesion coetaneous focal mucinosis (CFM) or cutaneous myxiod cyst is Oral Focal Mucinosis (OFM).(4) Oral focal mucinosis can be a sessile or pedunculated mass. It is a rare disease of unknown aetiology, and most commonly occurs in adult women. women, during the fourth and fifth decade of life(4). but can also be seen in children(5). Localization is most common in the gingiva, secondarily in the hard palate and varies in diameter between 1-2 cm (6,7)Oral Focal Mucinosis (OFM) has no distinctive clinical features and is most often clinically thought to be fibroma, pyogenic granuloma, mucocele or similar lesions (8)They appear as a nodular lesion covered with mucosa which is round or oval shape, pink white in color, with no clinical signs(6). oral focal mucinosis is presented as an innocuous soft tissue swelling that may be either pedunculated or sessile.(7)There is a predilection for the mucosa overlying bone and keratinized mucosa which is almost involved exclusively, with 80% of lesions developing on the gingiva and other locations involved are palate, alveolar mucosa and tongue.(9)The pathogenesis of OFM is suggestive of overproduction of hyaluronic acid by fibroblasts at the expense of collagen production.(1,9) Generally oral focal mucinosis is characterized by a localized, sub-epithelial, non-encapsulated area of loose, myxomatous connective tissue encircled by normal collagen bundles. The myxomatous areas show minimal to absent reticulin fibers and fragmented collagen fibers replaced by mucin.(10)

MATERIAL AND METHODS:

The systematic review of oral mucinosis was carried out using Pubmed and Google scholar databases. The keywords used were (mucinosis, oral mucinosis and oral mucinosis AND oral cavity) and all articles from 1981- 2016 were taken for initial evaluation.

Inclusion criteria:The initial search yielded 56 articles for the search terminologies. After close scrutiny of their abstract for the relevance on our topic 21 articles were short listed. Only full text article were included which summed up to 12 case studies. The articles included were case reports and case series with emphasis on histopathology of the lesion.

Exclusion Criteria: Case reports other than oral mucinosis& occurred other than humans were excluded from the search. Other than full text articles were excluded for the study. All selected articles were reviewed in full text after gaining access.

The purpose of this review was to delineate the clinical, histopathological, treatment & recurrence of oral mucinosis in order to aid its diagnosis and alleviate confusion between other tumours in this category.

RESULTS:

In the present systematic review the following 12 articles from the year 1981-2016 enlisted in (table1)(7),(11),(12),(9),(13),(14),(4),(2),(3),(8),(1),(15) are studied. In this review the following headings like the number of cases, histopathology, treatment, follow up & recurrence are discussed. Of the 12 articles 5 articles didn't delineate the recurrence of mucinosis. Among the articles reviewed many articles had dealt with special stain & immunohistochemistry which help to confirm the diagnosis of oral mucinosis.

DISCUSSION:

Oral Focal Mucinosis (OFM) is an oral counterpart of dermal lesion known as cutaneous focal mucinosis (CFM) or cutaneous myxiod cyst which is misdiagnosed as intraoral myxoma(10)& is of unidentified aetiology(16). It is an uncommon soft tissue tumor commonly found on the gingiva and presents as a painless, sessile or pedunculated mass which is of the same colour as the surrounding mucosa.(16)(17)OFM has a predilection for the mucosa overlying bone, and keratinized mucosa is almost involved exclusively, with 80% of lesions developing on the gingiva and the remainder on the palate; other locations are the alveolar mucosa and tongue(9,16) The pathogenesis of OFM is suggested to be the 'overproduction of hyaluronic acid by the fibroblasts at the expense of collagen production' but cause of this overproduction is not known.(18)The histopathological features of OFM demonstrate a submucosal, well-localised but non-encapsulated nidus of very loose, myxomatous or 'mucinous' connective tissue. Other superficial lesions may produce atrophy and loss of rete ridges of the overlying squamous epithelium. Fibroblasts are seen least within the mucinous area, often signifying delicate, fibrillar processes. The mucinous zone is much less vascular than the adjoining connective tissues, and inflammatory cells are not connected with the lesion except as a perivascular infiltrate of lymphocytic T-cells at the periphery.(6) From the review of 12 case reports 28 cases were reported, considering the 8 case reports with only abstract, the total cases reported till 2016 is about 65 cases. Among the available data for the site of occurrence gingiva is the most common site of occurrence. Among the 28 cases reported 20 cases showed the gingiva as the site of occurrence & 17 cases females were commonly affected. Kinjal

Deepak Rambhiaetal in 2016 reported a case of OFM which showed verrucous papules that coalesced to form a finger-like projection(15).Eran Gabay et al in 2010 reported a case of OFM showing cervical root resorption in relation to the OFM. A OFM was reported after surgically assisted RME was performed(19)G V Sowmya et al in 2015 reported a case of OFM that showed a gradual increase in size of the lesion in a span of 3 months(1). A case of OFM in a 2 year old child with bilateral involvement of palate was reported by Woo J ea al in 2015(20).Talacko AA et al in 2014 reported 2 cases of OFM with ulceration(21)

Narayana etal in 2009 reported 7 cases of OFM of which 6 were reported in gingiva & 1 in palate(22).Buchner A et al in 1990 reported 15 cases on OFM of which gingiva & alveolar mucosa are the most common sites affected(23). There was no recurrence noted in the cases reviewed.

Oral mucinosis have a clinical & histopathological differential diagnosis these are ruled out by clinical examination, H&E section, special stains & IHC markers. Clinically differential diagnosis is classified as inflammatory origin, tumors, malignant or metastatic & non-plaque related lesions. Inflammatory origin are gingivitis, fibrous hyperplasia, peripheral giant cell granuloma, epulis fissuratum, pyogenic granuloma. Tumors are peripheral ossifying fibroma & peripheral ameloblastoma. Non-plaque related lesions are irritation fibroma & fibrous hyperplasia, these can be ruled out histopathologically.(9) Histopathologically OFM is differential diagnosed as myxoid nerve sheath tumor & odontogenic myxoma, based on special stains like positive alcian blue & negative PAS staining and IHC marker like S-100 negativity and vimentin positivity help in diagnosis of OFM.(7). In myxoid nerve sheath tumor S-100 is positive & in H & E it is more circumscribed, has fibrous septa between multiple myxoid nodules, and has more plump stromal cells.(10)

CONCLUSION:

Oral focal mucinosis is rare soft tissue tumors which have various clinical differential diagnosis like peripheral giant cell granuloma, peripheral ossifying fibroma & peripheral ameloblastoma and histopathologically OFM is differentially diagnosed as myxoid nerve sheath tumor & odontogenic myxoma. So this is to throw light on this rare lesion, so while considering a soft tissue lesion mainly in gingiva, buccal mucosa, OFM should be considered in diagnosis. Special stain and IHC marker like alcian blue positivity & vimentin positivity respectively is considered in diagnosis of oral focal mucinosis.

Table 1:

S no	Type Of Article	Author	Year	Age	Sex	No.of Cases	Site	Size	Histopathology	Treatment	Recurrence & Follow Up
1	Case report with review of literature	Michael J. Aldredetal(7)	1981-2003	38 30 16 56 60 49 31 52 74 40 55 37 35 33 68	F F F F F M F M M F M F F F M	15	Lip Gingiva Gingiva Buccal mucosa Mouth Gingiva Gingiva Gingiva Lip Gingiva Tongue Gingiva Gingiva Gingiva Gingiva	10mm 3mm 8X5mm 10X5mm 9X8X3mm 5mm 15mm 7mm Not given 8X5mm 5mm Not given 5X3X3mm 6mm 10X5mm	Most cases had a circumscribed region of myxoid tissue containing scattered elongated, stellate and ovoid cells. In some specimens the characteristic myxoid tissue merged into the surrounding tissue. Most cases had a border of normal connective tissue between the lesion and overlying epithelium. Occasional cases appeared to have more than one area of mucinous tissue. Mitoses were rare and inflammatory cells were seen infrequently.	----------	19yr 7yr 6weeks Not Given 10 Mon None None None Not Given 1 Week 1mon 8 Mon None None 1 Week *no Recurrence For All 15 Cases
2	Case report	Ichiro saitoetal(11)	1985	35 50	M F	2	Mandibular anterior gingival,labially& lingually gingiva extending from the left mandibular central incisor to the right canine region	labially 7X8X4mm Lingually 10X10X4mm 20X12X7mm	The lesions consisted of fairly well-localized areas of myxomatous connective tissue surrounded by dense fibrous connective tissue in the deep and lateral areas. The myxomatous zone extended to the overlying epithelium and produced a definite flattening or bending of the rate ridges. Same as case 1	Resected Resected	Not given Not given
3	Case report	Giuseppe soda etal(12)	1998	68	M	1	Anterior ventral tongue	0.5 cm diameter	----------	Fully excised	Not given
4	Case report	Giovanna Iezzietal(9)	2001	48	M	1	gingiva of the left mandibular central incisor	1cm	Microscopically, a myxoid connective tissue with spindled and stellate fibroblasts was present. a diffuse positivity of the stroma and the myxoid matrix was present	Fully excised	NO,4 Year follow up
5	Case report	Eran Gabay et al (13)	2010	44	F	1	Gingiva with cervical root resorption	Not given	Histologically, this was a well-circumscribed yet unencapsulated lesion composed of myxomatous and mucoid connective tissue in the submucosa which contained numerous stellate-shaped fibroblasts and small blood vessels	Not completely excised	No,6 months follow up

6	Case report	JG Lee, etal(14)	2012	17	F	1	gingival papilla lingual to the lower left central and lateral incisors	6mm	Histopathologically revealed myxoma-tous connective tissue with a plasma cell infiltrate on haematoxylin and eosin stained sections. Alcian Blue-PAS pH 2.5 showed strong blue staining reveals the presence of hyaluronic acid	Fully excised	NS
7	Case report	Sharma Ena et al (4)	2013	Case 1 26 Case 2 36	M F	2	Gingiva of 22 & 23 Gingiva of distal 21 to mesial 11	15X15mm 8X7mm	Hematoxylin and eosin stained microscopic slides of both cases revealed a stratified squamous hyper-parakeratinized epithelium and the underlying connective tissue stroma was composed of loose fibro-myxoid stroma with stellate shaped fibroblasts. Deeper stroma showed spindle shaped fibroblasts interspersed between thin collagen fiber bundles and numerous small blood capillaries.	Fully excised Fully excised	NO,8 Month follow up NO,6 Month followup
8	Case report with review of literature	Mervasolukte kkesin et al (2)	2013	19	M	1	Palatal side of the maxillary premolar teeth.	2cm	Microscopic examination revealed mild flattening and parakeratosis in rete-ridges in the multilamellar epithelium overlying the parts. Bipolar, fusiform, or star-shaped fibroblasts scattered in a loose mucinous stroma. Some hypercellular cells were observed but no mitosis was observed. Chronic inflammatory cell infiltration was observed at the mild level of lymphocytes in the surrounding connective tissue. positive staining with alsian blue was detected and it was shown that this content was hyaluronic acid.	Fully excised	NO,8 Month follow up
9	Case report with review of literature	Jose RinoNetoetal (3)	2014	20	F	1	attached gingiva	10mm	The histologic sections showed fragments of mucosa that were partially covered with hyperparakeratinized stratified squamous epithelium with the areas of acanthosis . The lamina propria consisted of dense connective tissue permeated by an intense, diffuse mononuclear inflammatory infiltrate with areas of loose myxomatous material that were stained with alcian blue and interpreted as mucin.	Fully excised	NO,2 Year follow up
10	Case report	Siva Kumar M et al (8)	2015	25	F	1	Gingiva in relation to 46	1cm	Histopathologically reveals parakeratinised stratified surface squamous epithelium in association with a nodule of myxomatous tissue associated with spindle cells	Fully excised	NO,1 Year follow up
11	Case report with review of literature	G V Sowmya et al (1)	2015	54	M	1	Gingiva & Vestibular mucosa of 46	2X1cm	H&E-stained microscopic slides of the case revealed a stratified squamous hyper-parakeratinised epithelium and the underlying connective tissue stroma was composed of loose fibromyxoid stroma with stellate-shaped fibroblasts. Deeper stroma showed the myxomatous stroma with plump fibroblasts and no inflammatory cells	Fully excised	NO,1 Year follow up

12	Case report	Kinjal Deepak Rambhiaetal(15)	2016	60	F	1	verrucous lesion on the right buccal mucosa	finger-like projection	Biopsy from the lesion revealed a sub-epithelial, localized non-encapsulated area of loose myxomatous connective tissue stroma with stellate-shaped fibroblasts. The overlying epidermis showed hyperkeratosis, acanthosis and papillomatosis. Alcian blue stain revealed blue staining in the areas of prominent mucin deposition.	Fully excised	NS

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