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Case Report

ORAL FOCAL MUCINOSIS OF TONGUE- A RARE CLINICAL PRESENTATION WITH REVIEW OF LITERATURE

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ABSTRACT
Oral focal mucinosis is a unique presentation that is considered as an oral counterpart of cutaneous mucinosis. It is associated with excessive production of hyaluronic acid by the fibroblasts resulting in the loss of collagen and myxoid degradation of connective tissue. This entity is prevalent in adults, fourth and fifth decades with a female predilection of 5:3. It commonly occurs in the keratinized mucosa (gingiva and hard palate). Surgical excision is the preferred modality of treatment and recurrence of the lesion is rare. The Literature revealed only eleven cases of oral focal mucinosis in India which included five cases in mandibular gingiva, three cases in maxillary gingiva, and three cases in hard palate. Only five cases of oral focal mucinosis with site specificity to the tongue are reported all over the world. This report presents the sixth case of OFM in the tongue worldwide and the first case in India. This report highlights clinical features, differential diagnosis, histopathological features and management of Oral Focal Mucinosis (OFM) of tongue in a 42 year old female patient.

Keywords: *Myxoid, Oral Focal mucinosis, Sessile, Tongue*

INTRODUCTION
Oral Focal Mucinosis is an exceptional non neoplastic soft tissue tumor like growth present in the oral cavity, most commonly in the keratinized mucosa. It is commonly described as the counterpart of cutaneous focal mucinosis (Saito *et al.,* 1985), a tumor like growth notable in the face and extremities. Oral focal mucinosis was first observed by Johnson and Helwig in the year 1966 and is distinguished from other clinical entities with their unique histological presentation, which they coined the term "cutaneous focal mucinosis" (Johnson and Helwig, 1966). In the year 1974, Tomich observed soft tissue growths at various sites in the oral cavity and on further investigation; it had similar histopathological presentation of focal myxoid degeneration in the connective tissue. He coined the term Oral Focal Mucinosis (OFM) (Tomich, 1974). The etiology of this clinical entity is attributable to factors such as local trauma to the site, excessive production of hyaluronic acid from the fibroblasts (Tomich, 1974). OFM occur in adults and with a higher incidence in females than males. They are asymptomatic soft tissue growth; without any distinctive clinical features from other soft tissue growth. But histopathological examination is distinctive as it reveals increased production of hyaluronic acid by the fibroblasts (Vipin and Singh, 2012). Malignant transformation is uncommon in oral focal mucinosis. The Treatment for OFM is surgical excision and recurrence is rare (Vipin and Singh, 2012). This case report presents the clinical features, differential diagnosis, histopathological features and management of Oral focal mucinosis in a 42 year old female patient.

CASE
A 42-year old female patient presented to the department of oral medicine and radiology with a burning sensation and growth in the right lateral surface of the tongue. The patient had a history of the burning sensation of the tongue for the past two months and noticed the growth in tongue one month earlier. The burning sensation has been gradual in onset with mild intensity at initial onset period, later it had progressed to severe burning sensation on intake of hot or spicy food, with dryness of the mouth and impingement of the sharp tooth on the corresponding lower side. In regards to the growth, she had noticed a small growth which gradually attained the size at presentation and the growth hampered normal

mastication. The past medical history revealed that she was diagnosed with diabetes mellitus before five years and under regular medication. She has undergone extraction in the left lower back region of the jaw before one year due to pain and the past dental history was non contributory.

General examination revealed that her built was moderate and on intraoral examination; soft tissue examination revealed a single, well-defined sessile growth present on the right postero-lateral border of the tongue, measuring approximately 2.0 x 1.7 cm in dimension [Fig:1a,b]. The growth was pale pink in color and with a yellowish slough at the centre suggestive of central ulceration. The peripheries appeared smooth and the surrounding surface appeared normal without any secondary changes to the region [Fig:1c]. Anteriorly it extended 4 cm from the tip of the tongue, posteriorly 2 cm from the circumvallate papilla region and laterally at the level of a grossly destructed right lower third molar with the sharp cusps approximating the lesion. On palpation, all the inspectory findings were confirmed; it was soft to firm in consistency, sessile without induration, non tender, non compressible, non reducible with no bleeding or any pus discharge. Hard tissue examination revealed a grossly destructed 48, restored 16 and 17, fixed partial denture in relation to 31, 41 and a partially edentulous area in relation to 36.

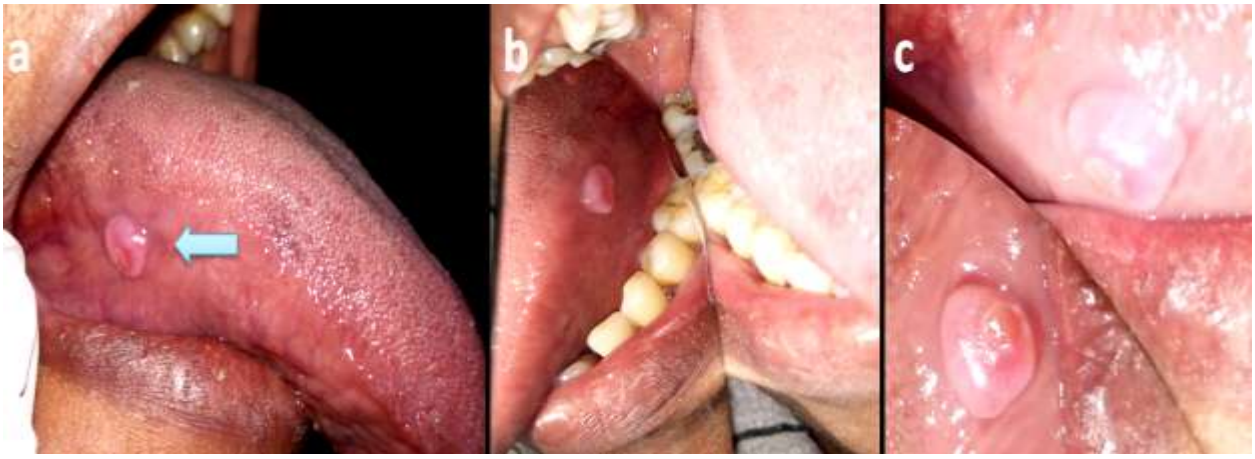

Figure 1: (a,b,c) reveals a growth in the right posterior lateral border of the tongue (a).The growth had impingement of the sharp tooth on the corresponding lower side (b).The growth was pale pink in color and with a yellowish slough at the centre suggestive of central ulceration. The peripheries appeared smooth and the surrounding surface appeared normal without any secondary changes to the region (c).

The regional lymphnodes were non palpable. On the basis of history and clinical examination, the lesion was provisionally diagnosed as traumatic fibroma and differential diagnosis of traumatic ulcerative granuloma with stromal Eosinophilia (TUGSE), pyogenic granuloma, fibroepithelial polyp and solitary neurofibroma were considered.

Informed consent was obtained from the patient followed by routine biochemical/hematological investigations which revealed hemoglobin - 12gms/dl; erythrocyte sedimentation rate - 3mm/hr; bleeding time - 2 minutes 10 seconds; clotting time - 3 minutes 40 seconds; fasting blood glucose level - 110mg/dl and postprandial blood glucose level - 130mg/dl. The patient was explained about the nature of the growth, recurrence rate, standard treatment plan and prognosis of the disease. Excisional biopsy was implemented in the right posterior lateral border of the tongue and complete excision of the lesion with

marginal clearance of two millimeter (mm) was performed and the grossly decayed 48 was extracted. Histopathological examination was advised and microscopic examination using H&E stain revealed hyperplastic parakeratinized stratified squamous epithelium exhibiting arcading pattern of proliferation, covering a mass of connective tissue. The underlying connective tissue showed myxoid areas with stellate fibroblasts. [Fig: 2a, b, c]. the presence of myxoid areas in connective tissue and the clinical appearance were correlated to arrive at the final diagnosis of Oral Focal Mucinosis.

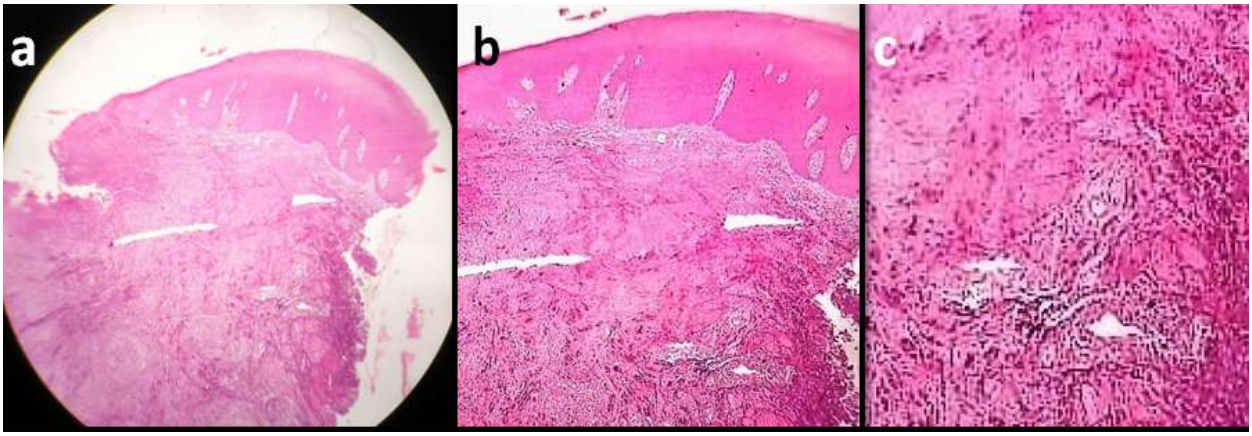

Figure 2: Photomicrograph of histopathological section at 10x, H&E stain revealed hyperplastic parakeratinized stratified squamous epithelium exhibiting arcading pattern of proliferation, covering a mass of connective tissue. The underlying connective tissue showed myxoid areas with stellate fibroblasts.

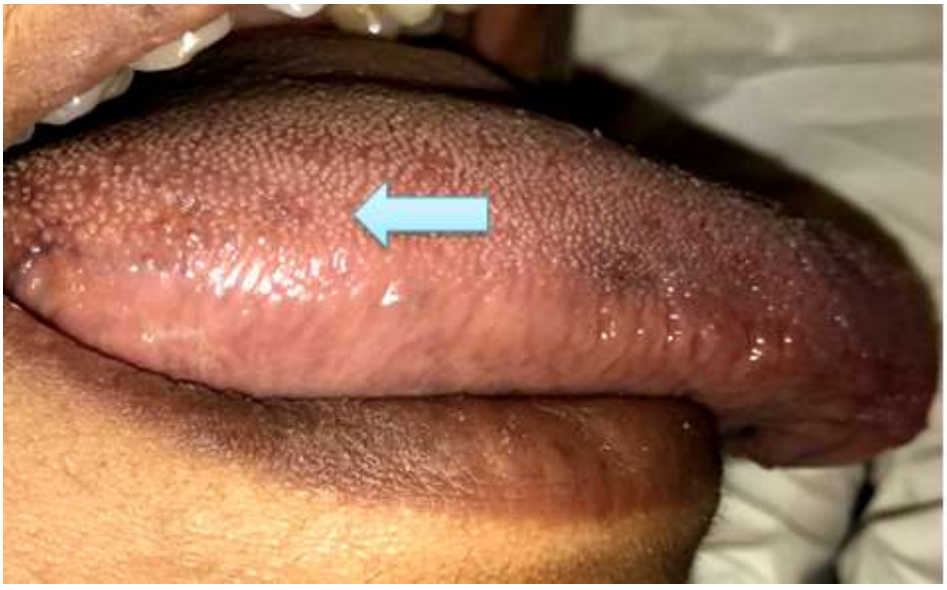

Figure 3a: It reveals post operative site with no signs of inflammation or recurrence after three month follow up

DISCUSSION

Case Report

Oral Focal Mucinosis (OFM), a rare clinical entity, is considered as the counterpart of cutaneous focal mucinosis (CFM) in oral cavity (Buchner *et al.,* 1990). It is misdiagnosed with other soft tissue growths such as fibroma, focal epithelial hyperplasia, mucocele when presented on the tongue (Mattsson and Lindberg, 2017). About ninety cases of human OFM have been reported, affecting children and adults ranging from 2 years to 88 years (Mattsson and Lindberg, 2017).The cases reported in the tongue are tabulated in [Table-1]

Table 1: Cases reported in the tongue

Author, Year	Age/Sex	Site	Duration	Provisional Diagnosis	Stain	Treatment	Recurrence
Tomich,1974	45/M	Tip of Tongue	2 months	Mucocele	H & E PAS-Alcian Blue	Excision	No
Buchner *et al.,* 1990	50/M	Anterior ventral tongue	2 months	Fibroma	H&E PAS-Alcian blue	Excision	No
Soda *et al.,* 1998	68/M	Anterior Ventral surface of the tongue	3 years	Asymptomatic swelling	H & E Ab-S-100 PAS-Alcian Blue	Excision	No
Aldred *et al.,* 2003	55/M	Tip of Tongue	3 months	Fibroepithelial polyp	H & E Ab-S-100 PAS-Alcian Blue	Excision	No
Pacifici *et al.,* 2012	62/F	Tip of the tongue	US	Fibrous growth	H & E Ab-S-100 PAS-Alcian Blue	Over all excision by 810nm laser diode	No
Mattson *et al.,* 2017	88/F	Dorsal surface of the tongue	US	Focal epithelial hyperplasia	H & E stain Ab-S-100	Excision	No

Females are affected than males with a predilection ratio of 5:3 (Woo and Cheung, 2015). They occur most frequently in the mucosa which is overlying the bone and in keratinized mucosa. From the previous literature, gingiva is the most common site and mandibular gingiva is more commonly reported than the maxillary gingiva. The hard palate, buccal mucosa, tongue and the lip has very limited occurrence (Madhusudhan *et al.,* 2010). In India eleven cases of OFM are reported so far of which five cases are of mandibular gingiva, three cases of maxillary gingiva, and three cases are of hard palate (Kumar *et al.,*

2017). Site specificity to tongue revealed only five cases reported so far. This report is the sixth case of OFM in the tongue worldwide and the first case in India to be reported. The etiology is still unknown with few factors contributing to the formation of OFM .Local trauma to the site is a validating factor, Neto et al and Gnepp et al reported OFM due to orthodontic treatment (Neto *et al.,* 2014 and Gnepp *et al.,* 1990). In our case, local trauma has played a role, which leads to the pathogenesis of this reactive process and attributed to the accumulation of hyaluronic acid between the collagen fibers, eventually replacing most of the collagen, and usually resulting in formation of small cystic spaces (Gnepp *et al.,* 1990). They are non-specific and appear as a single nodule, exophytic, elevated, sessile, pedunculated or verrucous growth and asymptomatic (Rambhia and Khopkar, 2016).Various clinical diagnosis are considered such as traumatic fibroma which is one of the most common lesion on the lateral surface of the tongue, along the line of bite (Reamy et al,2010) caused due to chronic irritation from the sharp cusp tip of the approximating tooth. The traumatic ulcerative Granuloma with Stromal Eosinophilia was taken into account as there was a yellow purulent membrane present in the center of the lesion and fibroepithelial polyp was considered as it's a reactive lesion owing to irritation (Paul *et al.,* 2016). Neurofibroma presents similarly as a solitary asymptomatic growth on the tongue (Roy *et al.,* 2015).

Radiographic investigations depend on the site of involvement (Gabay *et al.,* 2010). Gingival OFM may reveal angular bone loss. Histological examination of OFM shows few reticular fibers within the myxomatous area, except for those associated with blood vessels. The mucinous material is alcianophilic at pH 2.5 and negative at pH 0.4 (Tobouti *et al.,* 2018). Metachromasia with toluidine blue was observed at pH 3.0 and absent at pH 1 (Lee *et al.,* 2012). The Hematoxylin and Eosin stain is the widely used stain for identification of OFM and Alcian blue staining contributory for confirmation of hyaluronic acid presence (Amanda-Katarinny-Goes Gonzaga DH and de Oliveira IP, 2018). To differentiate between myxoid neural lesions from OFM, further Immuno-histochemical staining with S-100 is done occasionally (Amanda-Katarinny-Goes Gonzaga DH and de Oliveira IP, 2018). The histological differential diagnosis of OFM includes the soft tissue myxoma. It shows an extensive network of reticular fibers, whereas little reticulum is present in focal mucinosis. Myxoma, in some instances, exhibits an infiltrative growth pattern while focal mucinosis 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 focal mucinosis (Higuchi *et al.,* 2019).

The standard treatment followed for OFM is surgical excision because there is need for complete excision of the OFM lining and to eliminate the recurrence (Narayana and Casey, 2009). The alternate treatment options are LASER of 810nm (Pacifici *et al.,* 2012) and 320nm which showed promising result without any recurrence (Ena *et al.,* 2013). Conventional surgical excision is preferred over Laser treatment as it gives us the precise incision margin and best wound healing at the site (Bhatsange *et al.,* 2016). We excised surgically due to its advantages and there is no report of malignant transformation (Iezzi *et al.,* 2001).

CONCLUSION

OFM in the tongue is an uncommon clinical presentation. The preoperative diagnosis of this lesion is difficult due to its rarity. There are many similar clinical presentations of oral focal mucinosis and hence histopathological examination is mandatory for definitive diagnosis. OFM does not undergo malignant transformation. To avoid recurrence it should be surgically excised and causes of trauma must be resolved.

REFERENCES

Aldred M, Talacko A, Ruljancich K, Story R, Newland S, Chen S, O'grady J, Bergman J, Smith A, Dimitroulis G and Redman J (2003). Oral focal mucinosis: report of 15 cases and review of the literature. *Pathology*, **35**(5)393-6.

Amanda-Katarinny-Goes Gonzaga DH and de Oliveira IP (2018). Clinicopathological study of oral focal mucinosis: a retrospective case series. *Medicina Oral, Patologia Oral Y Cirugia Bucal* **23**(4) e401. https://doi.org/10.4317/medoral.22291

Bharti Vipin and Jagmohan Singh (2012). Oral focal mucinosis of palatal mucosa: A rare case report. *Contemporary Clinical Dentistry* **3**. Suppl 2:S 214-18.https://doi.org/10.4103/0976-237X.101098

Bhatsange A, Meshram EP, Waghamare A, Shiggaon L, Mehetre V, and Shende A (2016). A clinical and histological comparison of mucosal incisions produced by scalpel, electrocautery, and diode laser: A pilot study. *Journal of Dental Lasers* Jul 1; 10(2) 37. https://doi.org/10.4103/2321-1385.196962

Buchner A, Merrell PW, Leider AS, Hansen LS (1990). Oral focal mucinosis. *International journal of oral and maxillofacial surgery* **19** 337-40. https://doi.org/10.1016/S0901-5027(05)80076-1

Ena S, Manjari N, Anirban C and Ramesh A (2013). Oral focal mucinosis: a rare case report of two cases. *Ethiopian Journal of Health Sciences* **23**(2) 178-81. https://www.ajol.info/index.php/ejhs/article/view/91250

Gabay E, Akrish S and Machtei EE (2010). Oral focal mucinosis associated with cervical external root resorption: a case report. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology* **110**(4) e75-8. https://doi.org/10.1016/j.tripleo.2010.04.002

Gnepp Douglas R,Carole Vogler, Cirilo Sotelo-Avila and Izak H. Kielmovitch (1990). Focal mucinosis of the upper aerodigestive tract in children. *Human Pathology* **21**(8) 856-8. https://doi.org/10.1016/0046-8177(90)90056-B

Higuchi Y, Tsushima F, Sumikura K, Sato Y, Harada H, Kayamori K, and Ikeda T (2019). Diagnosis and treatment of oral focal mucinosis: a case series. *Journal of Medical Case Reports* **13**(1) 108. https://doi.org/10.1186/s13256-019-2033-8

Ichiro Saito, Fumio Ide, Takeshi Enomoto and Itsuro Kudo (1985). Oral Focal Mucinosis. *Journal of oral and maxillofacial surgery* **43**(5) 372-374. https://doi.org/10.1016/0278-2391(85)90259-9.

Iezzi G, Rubini C, Fioroni M, and Piattelli A (2001). Oral focal mucinosis of the gingiva: case report. *Journal of Periodontology* **72** 1100-2. **https://doi.org/10.1902/jop.2001.72.8.1100**

Johnathan Woo and Wa Sham Cheung (2015). Bilateral oral focal mucinosis on the palate of a 2-year-old child: a case report *International journal of paediatric dentistry*, **25**(1) 70-2. https://doi.org/10.1111/ipd.12098

Johnson WC,Helwig EB (1966). Cutaneous focal mucinosis: a clinicopathological and histochemical study *Archives of Dermatology* **93**(1) 13-20.

Lee JG, Allen G, Moore L and Gue S (2012). Oral focal mucinosis in an adolescent: a case report. *Australian dental journal* **57**(1) 90-2. https://doi.org/10.1111/j.1834-7819.2011.01649.x

Madhusudhan AS, Nagarajappa Das, Manjunatha BS, Swati Saawarn and Charan Babu HS (2010). Oral focal mucinosis: report of two cases. *Revista Odonto Ciência* **25**(3) 310-3. https://doi.org/10.1590/S1980-65232010000300018

Mattsson U and Lindberg P (2017). Oral focal mucinosis of the tongue: A rare clinical entity?.*Oral and Maxillofacial Surgery Cases* **3** 1-4. https://doi.org/10.1016/j.omsc.2017.01.006

Narayana N and Casey J (2009). Oral focal mucinosis: review of the literature and seven additional cases. *General dentistry* **57**(2) e11-3.

Neto JR, Sendyk M, Uchida LM Nunes FD and de Paiva JB (2014). Oral focal mucinosis associated with surgically assisted rapid maxillary expansion. *American Journal of Orthodontics and Dentofacial Orthopedics 145(4):534-8. https://doi.org/10.1016/j.ajodo.2013.02.035*

Nilesh Kumar,Huzaifa S Kothi,Rahul Patil and Pramod RC (2017). Oral focal mucinosis of posterior maxilla. *Journal of oral and maxillofacial pathology* **21**(2) 273. *https://dx.doi.org/10.4103/jomfp.JOMFP_242_15*

Pacifici L, Meleo D, Pompa G, Pacifici A, Gambarini G and Testarelli L (2012). Oral focal mucinosis of the tongue: a rare clinical case. *European Journal of Inflammation*, **10**(1) 111-5.https://doi.org/10.1177/1721727X1201000113

Paul S, Anuroopa P, Kulal R and Savita S (2016). Report of an unusually Huge Fibroepithelial Polyp managed with Diode Laser. *JOHSR* **6** 52-5. https://www.johsr.com/doi/pdf/10.5005/jp-journals-10042-1021

Rambhia K and Khopkar U (2016). Verrucous oral focal mucinosis. *Indian journal of dermatology, Venereology and Leprology* **82**(3). https://doi.org/10.4103/0378-6323.174384

Reamy BV, Derby R and Bunt CW (2010). Common tongue conditions in primary care. *American Family Physician* **81** 627-34.

Roy P, Chakraborty S, Das S and Roy A (2015). Solitary neurofibroma at the base of the tongue: A rare presentation. *Indian journal of dermatology* **60** *497. https://dx.doi.org/10.4103%2F0019-5154.164374*

Soda G, Baiocchini A, Bosco D, Nardoni S and Melis M (1998). Oral focal mucinosis of the tongue. *Pathology & Oncology Research* **4**(4) 304-7.

Tobouti, Priscila Lie, Fernando Kendi Horikawa, Bruno Fernandes Matuck, Suzana Cantanhede Orsini Machado de Sousa, and Fábio Abreu Alves (2018). Oral focal mucinosis of the hard palate and gingiva. *Autopsy & Case Reports* **8**(4). https://doi.org/10.4322/acr.2018.044

Tomich CE (1974). Oral Focal Mucinosis: A clinicopathological and histochemical study of eight cases. *Oral Surgery, Oral Medicine, Oral Pathology* **38**(5) 714-24. https://doi.org/10.1016/0030-4220(74)90392-2