EOSINOPHILIC ULCER OF THE TONGUE - A RARE AND DISTINCT ENTITY

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ABSTRACT

Eosinophilic ulcer of the tongue is a benign, ulcerative lesion of the tongue with a distinct histopathological appearance. It manifests as an ulcer with elevated margins. In infants and children it is termed as Riga-Fede disease. Though the pathogenetic mechanisms implicated in its development are poorly understood, it possibly results from repeated trauma to the tongue. The lesion is generally self limiting and usually responds well to conservative management. We document this distinct and rare entity in a 36-year old female patient.

Keywords*: Eosinophilic ulceration of tongue, Riga-Fede Disease, traumatic tongue ulcer, tongue ulceration with eosinophilia*

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INTRODUCTION

Eosinophilic ulcer of the tongue (EUOT) also termed as as Riga-Fede disease (RFD) is a benign, ulcerative lesion resulting from the repetitive trauma of contact to the mucosal surface of the tongue with the teeth. This entity was first described in 1881 by the Italian physician, Antonio Riga, and as additional cases were subsequently published by F. Fede in 1890, it has been known as Riga-Fede disease.[1,2] Although this terminology is primarily used to describe the condition in children, similar clinical and histopathological findings can also be found in adults. [3,4,5] The cause of eosinophilic ulcer of the oral mucosa is unknown. Trauma is a reported trigger in 39%. The ulcer usually occurs on sites where trauma from teeth is common and it is seen in the age group most likely to have damaged teeth and dentures that may cause trauma. However, most simple traumatic ulcers in the mouth do not show the characteristic clinical and histological features of eosinophilic ulcer. Therefore it has been suggested that trauma may allow as-yet-unidentified infections, toxins or foreign proteins to enter and trigger the characteristic inflammatory reaction in susceptible people. Another theory is that is represents a CD30+ lymphoproliferative disorder.The clinical significance lies in the fact that it may be mistaken for malignancy.

CASE HISTORY

A 37 - year old female presented to the outpatient department with chief complaint of painful ulcerated nodule in the left lateral aspect of the tongue. Patient complained of pain while chewing food. There was no cervical lymphadenopathy and no other significant past medical history. On oral examination there was a small solitary polypoidal lesion measuring 1x1 cms with surface ulceration and induration. The base appeared white yellow. The edges were raised and firm. The surrounding area showed redness. There was no evidence of enlarged lymph nodes and tonsils. A provisional diagnosis of papilloma of the tongue was considered. An excision biopsy was done and sent for histopathological examination.

Grossly a polypoidal mass measuring 1x1x0.5cm was received which was completely embedded. Microscopic sections showed tissue lined by stratified squamous epithelium with prominent area of ulceration (Fig.1). The sub epithelium showed dilated blood vessels (Fig.2). Sheets of inflammatory cells predominantly eosinophils admixed with neutrophils and lymphocytes were seen (Fig. 3). In view of the typical location and the characteristic histopathological appearance a diagnosis of

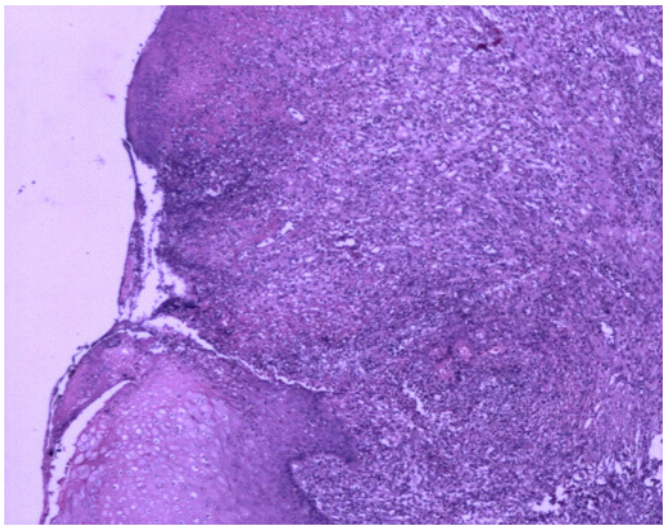

Fig.1: Photomicrograph showing mucosa of the tongue lined by stratified squamou epithelium with adjacent ulceration. H&E X 40

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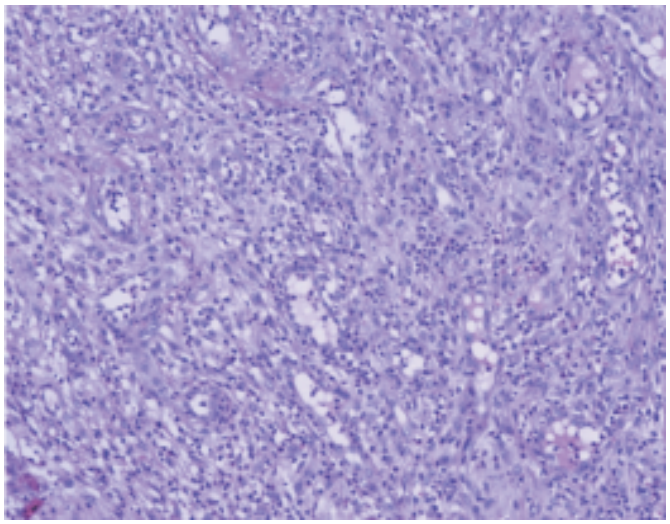

Fig.2: The sub epithelium shows dilated blood vessels surrounded by mixed inflammatory infiltrate. H&E X 100

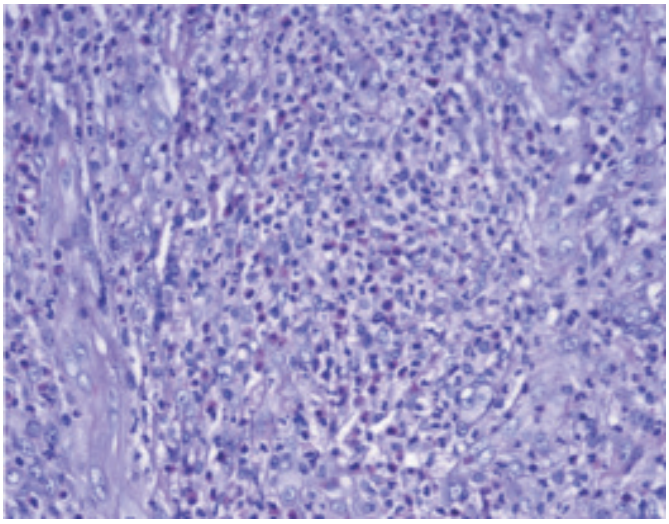

Fig.3: High power shows numerous scattered eosinophils with bright reddish pink cytoplasm H&E X200

eosinophilic ulceration of tongue also called Riga- Fede disease was rendered. Follow up at the end of 6 months showed no recurrence.

DISCUSSION

Eosiphilic ulcer of the tongue or Riga- Fede disease is a rare and distinct entity. It is a reactive mucosal disease as a result of repetitive trauma to the tongue by the anterior primary teeth during forward and backward movement Though these lesions are microscopically identical, the causes of trauma differ in the adult population as they may be related to the presence of broken teeth or ill-fitting prosthetic material in the oral cavity. It usually follows a benign course. Early recognition of this entity is important because it may be the presenting sign of an underlying neurological disorder.[1] Other associations include Lesch-Nyan syndrome, Tourette syndrome, familial dysautonomia, microcephaly, macroglossia and tongue biting. In infants new ulceration on the ventral surface of tongue is commonly associated with trauma, eruption of primary lower incisors with repetitive tongue thrusting habit, and in children with familial dysautonomia. Several factors such as superficial position of the tooth germ, infection or malnutrition, febrile states, hormonal stimulation, hereditary transmission of a dominant autosomal gene, osteoblastic activity inside the tooth germ and hypovitaminosis have been commonly implicated.[7]

Our patient showed a single lesion in the tongue with no other associated condition. A broad variety of terms have been used to describe RFD, such as eosinophilic ulcer of the oral mucosa, sublingual fibrogranuloma, sublingual growth in infants, lingual traumatic ulceration, traumatic atrophic glossitis, traumatic granuloma of the tongue, and traumatic ulcerative granuloma with stromal eosinophilia.[1,2,3,4,6]

It has been suggested that trauma may allow as-yet-unidentified infections, toxins or foreign proteins to enter and trigger the characteristic inflammatory reaction in susceptible people. Another theory is that it represents a CD30+ lymphoproliferative disorder.

It is reported that EUOT exhibits a slight female predominance in most of the cases with a peak incidence between the sixth and seventh decades of life.[6] Our case was a female patient in the fourth decade.

Histopathology shows an ulcerated surface with an underlying mixed inflammatory infiltrate including lymphocytes, macrophages, mast cells and a predominance of eosinophils. Atypical histiocytic granulomas may also be seen.

The differential diagnosis includes aphthous ulcer of the mouth and squamous papilloma with ulceration. Rarely malignant ulcer may have to be considered. Eosinophilic ulcer of the oral cavity usually heals by itself within one month, but can persist up to one year. Recurrences have rarely been reported.

Rarely, the associated ulceration may remain for years and can result in long-lasting tongue deformity.[1,2]

Treatment is conservative management with minimizing the repetitive trauma and preventing the provoking factor. EUOT is a self limiting disease which usually resolves in a few weeks to months. However the most frequently performed therapy is surgical excision. No further recurrence is noted after excision.

We document this rare case of eosinophilic ulcer of the tongue to emphasize the clinical and histopathological aspects that are distinct and relevant for diagnosis and treatment of this interesting condition.

REFERENCES

1. Zaenglein AL, Chang MW, Meehan SA, Axelrod FB, Orlow SJ. Extensive Riga-Fede disease of the lip and tongue. J Am Acad Dermatol 2002;47: 445–7
2. Baroni A, Capristo C, Rossiello L, Faccenda F, Satriano RA. Lingual traumatic ulceration (Riga-Fede disease) Int J Dermatol 2006;45:1096–7
3. Ceyhan AM, Yildirim M, Basak PY, Akkaya VB, Ayata A. Traumatic lingual ulcer in a child: Riga-Fede disease. Clin Exp Dermatol 2009;34:186–8.
4. Cunha VS, Zanol JDR, Sprinz E. Riga-Fede;like disease in an Acquired immune deficiency disease syndrome patient. J Int Assoc Physicians AIDS Care 2007:6;273-4.
5. Uwe Wollina, Riga-fede –Like disease in a 70 year old woman. Indian J Dermatol 2010;55:92-4.
6. Segura S, Pujol RM. Eosinophilic ulcer of the oral mucosa: A distinct entity or a non-specific reactive pattern? Oral Dis 2008;14:287–95.
7. N Ghimire, IK Maharjan, A kumar, B Koirala. Riga-Fede Disease. Health Renaissance 2013;11;174-6.