Riga-Fede disease: report of a case and review

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Self-mutilation of tongue is a type of self-injurious behavior. Ulcers of the lingual frenum in neonates with natal lower incisors are referred to as Riga-Fede disease. In this paper a case of Riga-Fede disease in a ten-month infant male with lower central incisors is reported. The ulcer resolved after the sharp incisal edges were smoothened and topical triameinolone was applied. As this lesion may be confused or associated with other serious disorders, a review of medical and dental literature was included.
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INTRODUCTION

Ulcerative lesions are common in children and usually caused by accidental biting, hard foods, appliances, or following dental treatment or other trauma. Non-accidental injuries are may caused by child abuse or children purposely traumatize their oral structures. This paper presents a case of an ulcerative lesion occurring on the lingual frenum and ventral tip of the tongue of a 10-month-old infant.

REPORT OF A CASE

A 10-month-old male child was referred by an otolaryngologist because of an "inflammatory lesion the tongue which had not healed following penicillin therapy."

An intraoral exam showed an ulcerated and swollen area on the lingual frenum and ventral tip of the tongue. Mucosa within the lesion had areas covered with yellow-gray, glistening exudate and areas of erythematous mucosa without exudate (Figure 1).

Examination of the rest of the intraoral mucosa revealed no other lesions. The patient had his lower central incisors recently erupted. The mother said that he often suddenly woke up crying, suggesting that the lesion resulted from "nocturnal" tongue biting, and the patient perceived pain. The pediatrician revealed that the medical history was not contributory.

Based on the mentioned findings the lesion was diagnosed as a Riga-Fede disease.

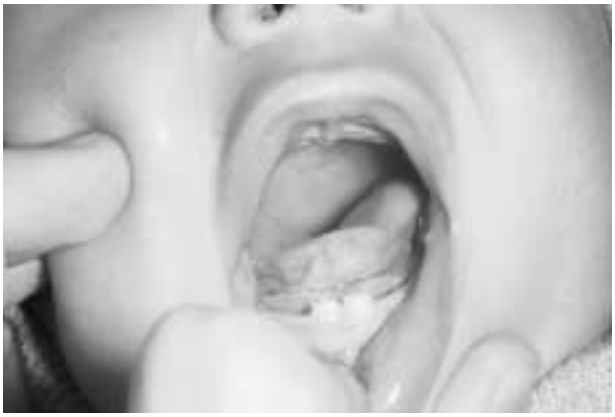

Figure 1. Clinical appearance of a Riga-Fede ulcer on the ventral surface of the tongue of a ten-month infant.

TREATMENT

The consecutive steps (options) suggested to eliminate the probable cause were:

1. observing the condition as many of these ulcers resolve without treatment, especially if the infant has normal cognitive abilities,
2. smoothing sharp incisal edges of the central incisors and another consult with the pediatrician may be in order if the smoothing of the teeth does not eliminate the problem.
3. placing domes of composite resin or splint over the teeth, or
4. extracting of the offending teeth may still be necessary if the lesion persists.

After the mother gave her consent, sharp teeth edges were smoothed to lessen local irritation (Figure 2). Topical applications of triameinolone acetonide in emollient (Kenalog™ in Orabase, Bristol Myers Squibb) were prescribed to help healing. Triamcinolone acetonide (Kena-

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Table. Major causes of tongue ulceration

Recurrent aphthous
Glossitis from drugs and nutritional disorders
Dental and electrolytic
Rheumatic diseases and Behcet's syndrome
Agranulocytosis and neutropenia
Malignant ulcers
Systemic histoplasmosis granuloma with tongue ulceration
Viral and bacterial infections
Lichen planus
Child abuse
Oral self mutilation (Self-injurious behavior)
- Familial dysautonomia (Riley-Day syndrome)
- Lesch-Nyhan syndrome
- Congenital indifference/insensitivity to pain
- Cerebral palsy
- Neuro-hematologic syndrome (Hoyeraal-Hreidarsson syndrome)
- Emotional problem

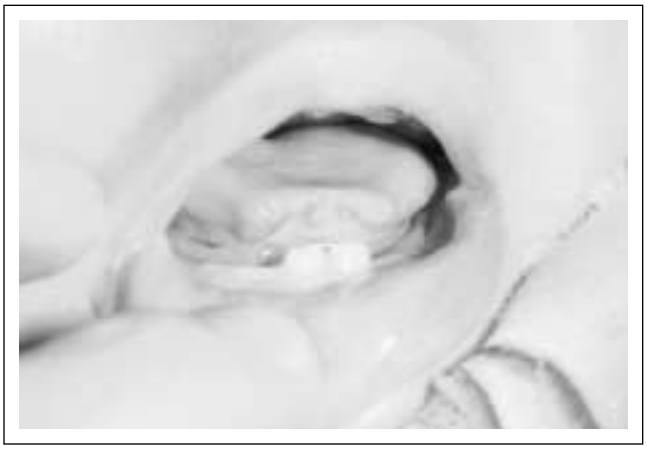

Figure 2. Treatment includes removal of the irritation source by smoothing sharp incisal edges of the lower central incisors.

log) is a synthetic corticosteroid which possesses anti-inflammatory, antipruritic, and antiallergic action.[1] The emollient dental paste (Orabase) acts as an adhesive vehicle for applying the active medication to the oral tissues. The vehicle provides a protective covering which, may serve to temporarily reduce the pain associated with oral irritation.

The next day, the mother reported that the pain and swelling had lessened and a few days later the ulcer was healed. Eliminating the sharp edges of the incisors and using topical medication allowed healing and confirmed the initial diagnosis of a traumatic ulcer of the ventral surface of the tongue, traditionally known as Riga-Fede disease.

DISCUSSION

The traumatic lesion of the tongue is most frequently an ulcerated, indurated mass of short duration, which may suggest a primary malignancy or fungal infection. In patients, who are unable to provide an adequate history, such as infants, the determination of a definitive clinical diagnosis is especially difficuh.

Most mouth ulcers are caused by trauma or are aphthous. Table lists major causes of tongue ulceration.

Recurrent aphthous ulcers (RAU) can frequently and severely affect the lateral margins and tip of the tongue. The clinical and histological appearance of traumatic ulcers is similar to that of aphthous ulcers.[2]

The differentiation between aphthous lesions and trauma is based on the history and the relationship of the lesion to a source of irritation. Also, any linearity in ulceration suggests a traumatic cause.

Many of the British studies, according to Aldred *et al*.[3] have suggested that RAU is associated with nutritional deficiency states. However, shallow but persistent ulcers appearing traumatic in origin and affecting the ventral surfaces of the tongue may be associated with a nutritional deficiency or a blood dyscrasia. Pernicious anemia can be accompanied by shallow ulcers appearing traumatic in origin. Ulcers associated with pernicious anemia may have little or no erythema present because of a depressed marrow inflammatory response.[4]

When ulcers persist, blood tests for full blood, hematocrit, hemoglobin, vitamin B_{12} folate, serum ferritin are important and should be used to exclude deficiency states.

A complete blood count and differential white count also serve to rule out myelosuppressive disease, such as leukemia. In such a case, mucosal ulcerations occur most frequently in traumatized areas such as the lateral borders of the tongue. Lesions appear as penetrating ulceration and are most often ovoid.[2] The borders may be raised and the base yellowish because of necrosis. Unlike in traumatic lesions, a peripheral band of erythema is usually absent again because of the inability to mount an effective inflammatory response.

Laboratory studies in addition to the biopsy are indispensable as the oral manifestations of blood disorders, such as leukemia, are not sufficient to cause most dentists to suspect hematological malignancies. This was clearly showed in a study by White[5] conducted many years ago.

A traumatic ulcer involving the lateral border of the tongue may resemble carcinoma. However, oral malig-

nancies present as expanding, painless ulcers. There are often rolled, hard borders around the lesion, and the surface of the ulcer may be smooth or pebbly and somewhat erythematous.

Probably one of the most powerful indicators of malignancy is that the ulcers do not heal in 10 to 14 days. Thus, an important axiom in dealing with oral ulceration is to perform a biopsy of any lesion that does not heal spontaneously within 10 days to two weeks following its initial appearance.

Scores of treatments have been tried in the attempt to cure "benign" oral ulcerations.[6] Some of the treatments include antibiotics, gamma globulin, steroids, vitamin therapy, topical anesthetics, and numerous other drugs and ointments. None of which was credited with more than partial success.

N-butyl-2-cyanoacrylate based tissue adhesive was found to adhere successfully to moist living tissues, forming a seal and allowing effective healing. It was also used to "cement" medicaments in place for an extended period, and found to bring about pain relief. Recently, it was employed as a non-suture method for closing wounds in oral surgery and found, as an animal study by Hunsuck *et a1*.[8] showed in 1970, to be well tolerated by the tissue and permitted hemostasis and normal healing of incisions.[9]

REVIEW OF THE LITERATURE

Riga-Fede is an interesting and quite characteric disease that is mainly observed in males with two distinct peaks in incidence during either infancy or adulthood.[10] An important aspect is that this lesion may be the initial presentation for some serious underlying medical problems; therefore, we consider it is imperative to review the pertinent medical and dental literature.

Term

In 1857 Cardarelli described the condition, which was later referred to by several Italian authors as "afta cachettica"." Its chief characteristics were ulceration on the lingual frenum of infants associated with general wasting, leading to "exitus lethalis". Riga, in 1881, was the first to really draw the medical world's attention to the malady, while the first histological examination was that of Fede in 1890.[11] It has subsequently been known as "Riga-Fede disease".

Other authors have referred to this lesion as "Riga's disease", "sublingual growth in infants", "sublingual ulcer", "sublingual granuloma", "reparative lesion of the tongue", "neonatal sublingual traumatic ulceration", and "traumatic atrophic glossitis".[12-14]

Interestingly, a lesion with similar clinical characteristics and identical histological features has been reported as "traumatic eosinophilic granuloma".[15] However, the latter lesion has been reported to occur during late adulthood and not restricted in location to the tongue; it may occur in the cheek-buccal mucosa, the vestibule, gingiva, or palate. Since these lesions have identical histomorphology and clinical characteristics and frequently associated with a history of trauma, Elzay[10] suggested the term "traumatic ulcerative granuloma with stromal eosinophilia (TUGSE).

This entity should have an appropriate, descriptive term because similarity of terms may result in confusion and inappropriate treatment.

ETIOLOGY

Most investigators believe that these lesions result from mechanical trauma produced by the tongue thrusting against erupted teeth during nursing. Some reports related an association with either a sharp tooth, a dental restoration, or a dental appliance.[10]

This possible etiology is supported by the fact that the most frequent location is the tongue. This may be explained by the fact that the mammalian tongue is an exceptionally mobile and functional structure and, as a result, is frequently traumatized during mastication and phonation.

Bhaskar and Lilly[16] reported seven reactive tongue lesions and, in the same paper, published the results of an experimental study of traumatic tongue lesions in rats. The experimental findings reported by Bhaskar and Lilly[16] and by Shteyer *et al*.[17] on traumatized rat tongues strongly implicated trauma as a causative factor for eosinophilic granuloma of the human tongue.

On the basis of a similar investigation, however, von Domarus *et al*.[18] considered this conclusion to be incorrect. Rather, they concluded that the etiology of eosinophilic granuloma of the tongue as that of other eosinophilic granulomas in man is still unknown.

Tang *et al*.[19] reported that, although the cause remains unknown, trauma appears to be a significant contributing factor. They postulated that, subsequent to a traumatic break in the epithelium, a viral or toxic agent invades the mucomuscular tissues as well as the capillary endothelial cells. Resultant swelling and degeneration of the endothelia cause occlusion of the capillary lumina and produce focal isehemia. They further suggested that the proliferation of capillaries and myoblasts may be compensatory, and the infiltration of histiocytes and eosinophils is reactive and reparative, all promoting healing of the lesion.[19]

Elzay[10] accepted the concept that trauma has some etiological role in the development of TUGSE lesions, but he commented, "If trauma is the sole cause, why do we not observe reactive lesions with stromal eosinophilia associated with other oral ulcerative processes?' He suggested that the pathogenesis of TUGSE lesions commences with ulceration resulting from some form of trauma, which permits the ingress of microorganisms, toxins, or foreign protein into the connective tissue. These substances, in predisposed persons, induce an adverse inflammatory process resulting from an exaggerated mast cell-eosinophil reaction sim-

ilar to that noted in the studies on bronchial asthma. The exact mechanism is as yet unknown. This mechanism would account for the unique histological and clinical features noted in TUGSE lesions and yet notably lacking in the plethora of other of ulcerative lesions of the oral mucosa.

Tang *et al.*[19] suggested that viral serology and isolation and immunofluorescent assays for such lesions be attempted in future studies. Elzay[10] recommended that future studies ascertain whether or not eosinophil major basic protein is present in either the afflicted tissue or sputum of patients with TUGSE lesions. It may also be of interest to determine whether TUGSE patients have either asthma or some other hypersensitivity condition.

HISTOLOGICAL FEATURES

Microscopically, the variable features of traumatic tongue lesions have been recognized by some authors and have been generally described as fibroblastic and histiocytic proliferations, which infiltrate between striated muscle bundles. Degenerating and regenerating muscle cells and varying numbers of eosinophils, neutrophils, and plasma cells further complicate the histomorphology.[14] It can mimic other diseases, including malignancy;[20] therefore, it is a lesion which clinicians and pathologist should be familiar.

Bizay was the first to report that stromal eosinophilia was accompanied by increasing numbers of mast cells when compared to their presence in control tissues. Based on this finding, he suspected that the eosinophil associated with cellular destruction in bronchial asthma might similarly contribute to the cellular damage noted in TUGSE lesions.[10]

MEDICAL DISORDERS

Aldred *et al.*[3] stated that Riga-Fede disease occurs almost exclusively in children with cerebral palsy (CP). Indeed, Riga-Fede persists in infants who have severe CP and they cannot control spasticity of the tongue hence the repeated ulceration.

Besides the possibility of cerebral palsy, Lesch-Nyhan syndrome,[22,23] familial dysautonomia, or congenital indifference to pain may be the underlying problem and the tongue biting may be the initial presentation.

Lesch-Nyhan syndrome (LNS) is an X-linked recessive disorder of purine metabolism. The prevalence of LNS is estimated to be one in 10,000 males. LNS is characterized clinically by hyperuricemia, growth impairment, spasticity, mental retardation, and self-mutilation.[21] The disease is usually diagnosed at the onset of self-mutilatory behavior. Therefore, the suspicion of LNS to the presence of self-mutilatory behavior, such as lip or tongue biting, should be considered. Otherwise, the correct diagnosis may be late or missed altogether.

Oro-dental abnormalities are quite frequent in familial dysautonomia, or FD, (Riley-Day syndrome). Self-injurious behavior is caused mainly by profound sensory loss. Mass *et al.*[22] reported that dental trauma was found in 59% of 22 patients with FD. Rakocz *et a1.*[24] reported a case of FD with Riga-Fede's disease. Later, Eichenfleld *et al.* reported another case.[25]

Congenital indifference to pain also is usually manifested in children by a history of unrecognized trauma, indifference to painful stimuli, or self-mutilation.[26] Awareness of the orodental self-mutilation as the primary teeth erupt in these patients is important as the patient is usually normal with respect to intelligence, development, psychological adjustments, and other sensory perceptions.[23]

TREATMENT

Treatment of this lesion has varied over the years. Historically, the most common modality of treatment has been excision of the lesion and/or extraction of the offending teeth.[10] Bhaskar and Lilly[16] reported a case treated by radiation, while Welbom[15] reported a case received no treatment. All cases were reported to have healed without recurrence. Tomizawa *et a1.*[27] found that mesial inclination of the neighboring teeth and narrowness of the extracted space were observed in the cases where extraction was performed at an early age.

Riga-Fede can be managed in some cases without extraction.[28] Elimination of the sharp edges of teeth was found to be helpful. Also, success has been reported with coverage of the teeth with light cured composite resin to provide a smooth rounded surface for the tongue to pass over. However, because of inadequate enamel surface area for resin bonding combined with the difficulties of field control, achieving adequate retention of the resin could be questionable. Should the restoration fail, the composite resin could also be swallowed or inhaled. Parents should be made aware of this possible complication.

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