# Riga-Fede disease: A histological study and case report

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## ABSTRACT

Acute traumatic ulcerations and granulomas of the oral mucosa may result from physical damage via sharp foodstuffs, accidental biting, or talking. Most ulcerations heal within days. Others become chronic, reactive, and exophytic. A histopathologically unique type of chronic traumatic ulceration is the traumatic ulcerative granuloma with stromal eosinophilia (TUGSE). TUGSE exhibits a deep "pseudoinvasive" inflammatory reaction. This lesion may occur under the tongue in infants as a result of chronic mucosal trauma caused by mandibular anterior primary teeth during nursing and is termed Riga-Fede disease (RFD). The clinical presentation many resemble squamous cell carcinoma causing concern. RFD, although not uncommon, is not frequently reported. Thus, dental practitioners are unfamiliar with such lesions. We present a large Riga-Fede lesion in an infant along with the clinical management.

**Key words:** Riga-Fede disease, traumatic granuloma, ulcerative granuloma

Riga-Fede disease (RFD) is a chronic traumatic ulceration presenting on the ventral surface of the tongue in neonates and infants, frequently associated with natal, neonatal, or primary lower incisor teeth.[1-8] Chronic traumatic ulcerations mostly involve the tongue, lips, and buccal mucosa. However, the anterior ventral surface of the tongue is the most common site. Individual lesions present as erythema surrounding a centrally removable, yellow, fibrinopurulent membrane. Often, the lesion develops a rolled white hyperkeratotic border immediately adjacent to the ulceration. Occasionally, however, the underlying proliferative granulation tissue results in a raised exophytic lesion similar to a pyogenic granuloma. The pseudolymphomatous pattern, termed atypical histiocytic granuloma, may be clinically misdiagnosed as a malignancy.[1,8,9]

## CASE REPORT

An 8-month-old infant boy suffering from cerebral palsy (CP) presented with a large exophytic lesion located on the anterior ventral part of the tongue [Figure 1]. He was referred to us by an ENT specialist. Examination revealed the infant's disability to suckle the mother's nipple for feeding. Laboratory tests previously performed (CBC, FBS, BUN, CR, CT, BT, U/A, Ca, K, PT, PTT) were normal. Under general anesthesia (GA), a wedge-type incisional biopsy was performed for diagnosis and to rule out malignancy. Contact of the anterior part of the tongue with the newly erupted anterior mandibular teeth was suspected to be the cause. Thus, the incisal edges of the anterior mandibular teeth were grinded round and covered with composite resin material after etching conveniently at the same time while still under GA. Atypical histiocytic ulcerative granuloma (RFD) was confirmed via histopathological diagnosis. The lesion resolved 2 weeks later [Figure 2]. Although recurrence is a possibility, it has not recurred in our case (2 years post operatively). This may be because the baby is no longer nursed.

## DISCUSSION

Simple chronic traumatic ulcerations occur most often on the tongue, lips, and buccal mucosa (sites usually injured by the dentition). RFD describes a traumatic ulceration presenting on the ventral surface of the tongue in neonates and infants typically between 1 week and 1 year of age.[1-6] Although usually associated with natal or neonatal teeth, it may also occur in older infants after the eruption of the primary lower incisors.[7-8] RFD has a significant male predominance.[1] Most reported Riga-Fedes have been on the anterior ventral surface of the tongue. However, the dorsal surface may also be affected. Ventral tongue lesions contact the mandibular anterior incisors whereas dorsal lesions contact the maxillary incisors. Although cases have been seen on the gingiva, buccal mucosa, floor of mouth, palate, and lip, the tongue remains to be the most common site of involvement. The ulcerations appear very similar to simple traumatic ulcerations; however, on occasion, underlying proliferative granulation tissue can result in

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a raised lesion similar to a pyogenic granuloma [Figures 3 and 4].[1] A pseudolymphomatous pattern termed atypical histiocytic granuloma may occasionally be misdiagnosed as lymphoma. The infiltrate is at times worrisome on light microscopy and immunohistochemical studies may be required to rule out this possibility (by demonstrating a polyclonal mixture of B lymphocytes, T lymphocytes, and histiocytes).[1,8-11]

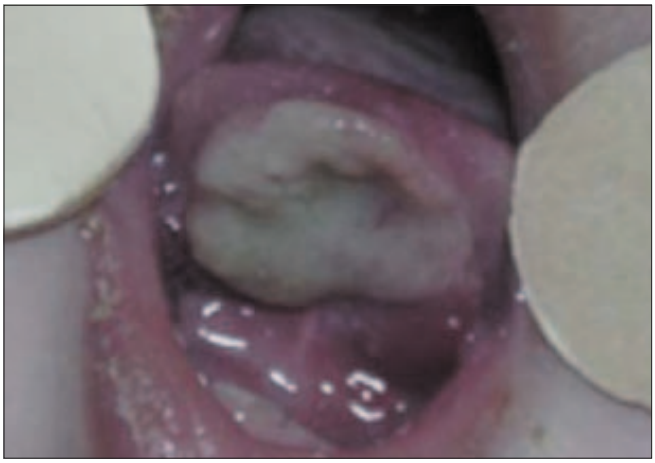

**Figure 1:** Photograph depicting a large 3 cm lesion on the ventral surface of the tongue

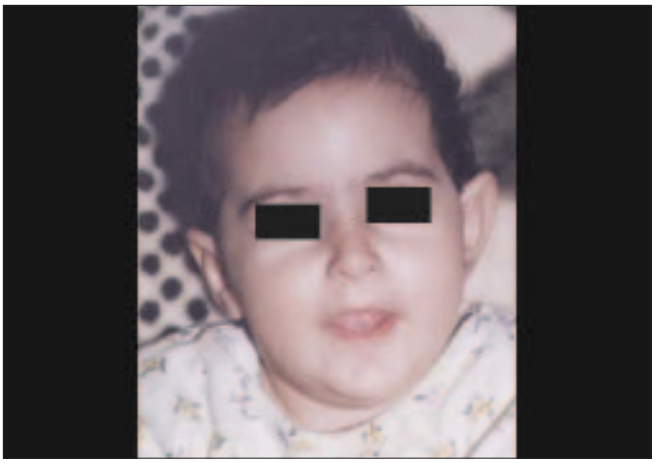

**Figure 2:** Resolution of the lesion

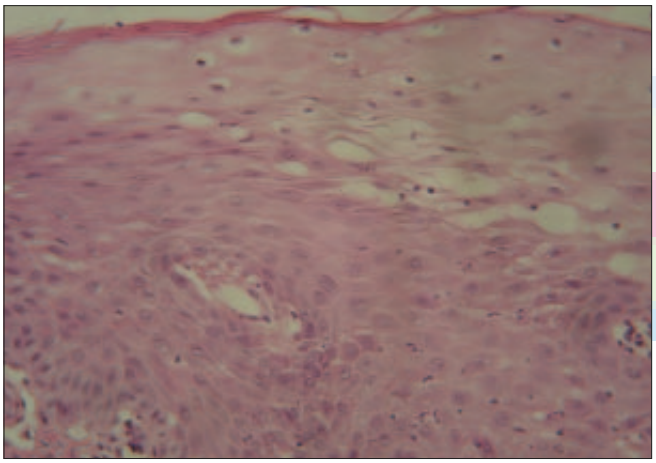

**Figure 3:** Epithelial hyperplasia near the surface of the lesion (H and E, 10x)

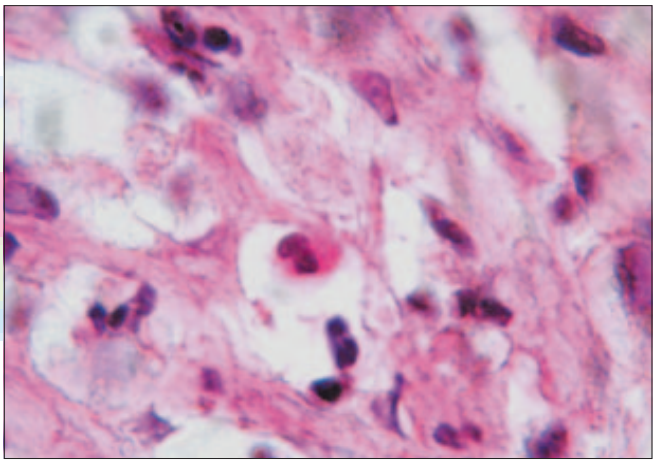

**Figure 4:** High-power photomicrograph of an eosinophil (H and E, 40x)

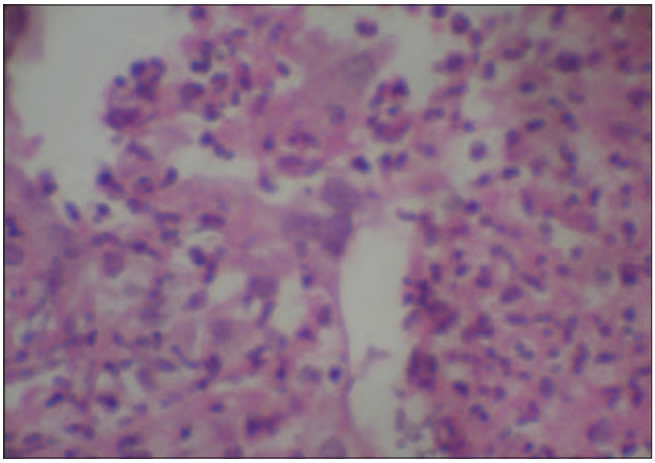

**Figure 5:** Medium-power photomicrograph of several atypical histiocytes and granulation tissue supporting a mixed inflammatory infiltrate (H and E, 20x)

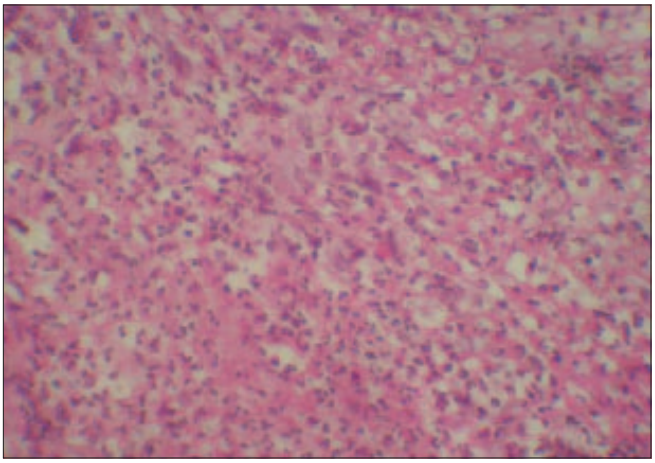

**Figure 6:** Low-power photomicrograph showing atypical cells and eosinophils in the depth of the lesion (H and E, 10x)

Although more common in infants, RFD has been reported in older patients and, recently, in a patient with acquired immunodeficiency syndrome.[12]

### Diagnosis

Biopsy is required for definitive diagnosis of RFD. Microscopic examination reveals a mixed inflammatory reaction under the ulcerated surface, including T cells, large mononuclear cells, and numerous eosinophils. Hence, it is often classified as a subtype of eosinophilic ulcer. The characteristic cellular infiltrates are deep-seated [Figures 5 and 6]. Early recognition of this entity is important in treatment because it may be the presenting sign of an underlying neurological disorder.[8] Our patient had CP, a persisting qualitative motor disorder appearing before age 3 due to non progressive damage to the brain. CP is the most common cause of motor disability in the pediatric age. The different definitions of CP cover motor sequelae secondary to an isolated brain injury that occurs in a developing brain.[10] Dyskinesias of various body parts accompanies CP, including the tongue, which may predispose the infant to RFD when the tongue is involved.[11]

### Treatment

Treatment should begin conservatively and should focus on eliminating the source of trauma. Failure to diagnose and properly treat this lesion can result in dehydration and inadequate intake for the infant.[8,9] For traumatic ulcerations that have an obvious source of injury, the irritating cause should be removed first. Biopsy is indicated for lesions that remain for 2 weeks after removal of the cause.[1] Rapid healing after a biopsy is typical even with large traumatic granulomas. Recurrence is usually not expected. The use of corticosteroids in the management of traumatic ulcerations is controversial. Extraction of the anterior primary teeth is not recommended either. Although tooth extraction has resolved the ulcerations in RFD, it may delay eruption of the permanent dentition. It seems best to retain the teeth if they are stable while eliminating trauma.[1] Construction of a protective shield is usually sufficient to allow resolution.[1-9]

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