Riga-Fede Disease

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Quick Response Code

doi: 10.5866/2014.611475

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Article Info:

Received: October 10, 2013
Review Completed: November 8, 2013
Accepted: December 7, 2013
Available Online: February, 2014 (www.nacd.in)
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ABSTRACT:

The term Riga-Fede disease has been used to describe chronic traumatic ulceration that occurs on the ventral surface of the tongue by natal and neonatal teeth in infants. It is important to diagnose the lesion and treat it by eliminating the source of trauma.

Key words: Riga-Fede disease, natal teeth, neonatal teeth.

INTRODUCTION

Riga-Fede disease was first described by Antonio Riga, an Italian physician, in 1881. Histological studies and additional cases were subsequently published by Fede in 1890. It has therefore become known as Riga-Fede disease. Riga-Fede disease is ulceration of the ventral surface of the tongue caused by trauma from continual protrusive and retrusive movements over the lower incisors. It is frequently associated with natal or neonatal teeth but may also occur in older infants after eruption of the primary lower incisors. It is important to note that this lesion may be the initial presentation of some serious underlying medical problems, such as Lesch-Nyhan syndrome and familial dysautonomia.^{1,2}

REVIEW OF THE LITERATURE

Riga-Fede is an interesting and quite characteristic disease that is mainly observed in males with two distinct peaks in incidence during either infancy or adulthood.² An important aspect is that this lesion may be the initial presentation of some serious underlying medical problems; therefore, we consider it is imperative to review the pertinent medical and dental literature.

TERMINOLOGY

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 1890.³ 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”, “traumatic granuloma of the tongue”, “traumatic ulcerative granuloma with stromal eosinophilia” and sublingual fibrogranuloma.

ETIOLOGY

These lesions result from mechanical trauma produced by the tongue thrusting against erupted teeth during nursing.³ 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.

Elzay suggested that the pathogenesis of Riga Fede 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 similar to that noted in the studies on bronchial asthma.²

Tang et al. 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 isohemia. 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.⁴

The experimental findings reported by Bhaskar and Lilly⁵ and by Shteyer et al.⁶ on traumatized rat tongues strongly implicated trauma as a causative factor for eosinophilic granuloma of the human tongue.

HISTOLOGICAL FEATURES.

Riga Fede disease is histopathologically characterized by an ulcerated mucosa with granulation tissue and a mixed inflammatory infiltrate consisting of lymphocytes, macrophages, mast cells and an abundant number of eosinophils, the latter being the most typical of this entity. 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.⁷ It can mimic other diseases, including malignancy;⁸ 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 the Riga Fede lesions.²

SYNDROMES ASSOCIATED.

- Lesche- Nyhan Syndrome.
- Tourettes syndrome.
- Adreno genital syndrome
- Cleft lip and palate
- Craniofacial dysosotosis.
- Craniostosis
- Ectodermal dysplasia
- Ellis van creveld syndrome.
- Epidermolysis bullosa
- Hallerman strieff syndrome.
- Jadassohn lewandowsky syndrome.
- Multiple steacystoma.
- Pallister-Hall syndrome.
- Pfieffer syndrome.
- Pierre robin syndrome.

- Polydactyl type 2
- Sotos syndrome.
- Steatomacystoma multiplex
- Van der woude
- Walker Warburg syndrome
- Weidemann Rautenstrauch syndrome.
- Familial dysautonomia.

DISCUSSION

Riga Fede disease is a reactive traumatic mucosal disease characterized by persistent ulceration of the oral mucosa. It develops as a result of repetitive trauma of the tongue by the anterior primary teeth during forward and backward movement. The lesion begins as an ulcerated area on the ventral surface of the tongue.¹⁰ Riga-Fede disease is initiated by the erosion produced by the sawing of the lingual frenum on the sharp, cutting edges of lower central incisors. With repeated trauma and secondary infection, granulation tissue forms and produces a tumor like mass, which is usually disc-shaped and about 1mm in diameter. In the center, there is usually an ulcerated area covered with fibrinous pseudomembrane. The lesion sometimes causes difficulty in nursing but there are no other symptoms. There is no associated lymphadenopathy or adjacent inflammation. Domingues-Cruz et al. proposed using a classification of the disease where in 'precocious Riga fede disease' defines those occurrences associated with (neo) natal teeth in the first 6 months of life, where no relation with neurologic disorders was found, and 'late Riga Fede disease' refers to those instances which typically start after 6-8 months of life, with the first dentition, usually the lower incisors. In the former, the existence of (neo) natal teeth, together with the instinctive sucking reflex and the tendency for the tongue to protrude favor the development of the disease. In the latter, the importance of recognition of the condition is due to its possible relationship to neurologic disease.¹¹ Several terms have been described in the literature which erupt before the normal time, such as congenital teeth, foetal teeth, precocious teeth, and dentitia praecox. According to the definition presented by massler, taking only the time of eruption as reference, natal teeth are those observable in the oral cavity at birth and neonatal are those that erupt during the first 30 days of life.

They are more common in the mandibular arch than the maxillary and in the mandibular, they are most commonly seen in the incisor region than the canine and the molar segments.¹²

DIFFERENTIAL DIAGNOSIS OF ULCERATED, INDURATED MASSES OF THE ORAL MUCOSA IN INFANCY.

LOCAL NEOPLASIA

- granular cell tumour
- myofibromad
- sarcoma
- extra-nodal lymphoma

INFECTION

- congenital syphilis
- tuberculosis

HEMATOLOGICAL DISORDER

- agranulocytosis

TRAUMATIC

- mechanical (Riga Fede disease)
- electrical
- chemical¹³.

TREATMENT.

Table 1. Management options for Riga-Fede disease.

Treatment	
Conservative	<ul style="list-style-type: none"> • Do nothing. • Modify feeding behaviour or feeding device • Wean the infant • Treat symptoms—Kenalog in Orabase. • Smooth off incisal edges of lower incisors • Use Stomahesive Wafers • Place domes of composite resin over the offending teeth
Radical	<ul style="list-style-type: none"> • Extract lower incisors • Treat the lesion by radiation. • Apply liquid nitrogen • Excise the lesion

Treatment of Riga Fede disease has varied over the years. In case of mild to moderate irritation to the tongue, conservative treatment such as smoothing the incisal edge with an abrasive instrument is advocated to alleviate feeding difficulties or complications like Riga-Fede disease. Alternatively, a small increment of composite may be bonded to the incisal edges. Extraction may be needed in cases where the Riga Fede disease remains unhealed, where the involved tooth is hyper mobile, increasing the chance of aspiration. Although many investigators have mentioned the possibility of aspiration of these teeth, this risk, in reality, is an unlikely possibility since there are no reports in the literature of the actual occurrence of aspiration. However, cases of spontaneous tooth exfoliation have been reported. If extraction is indicated, the child should be at least ten days or above. Otherwise prophylactic administration of vitamin K (0.5 - 1.0 mg, IM) is advocated before and after extraction, since vitamin K is essential for the production of prothrombin in the liver as there could be risk of haemorrhage.^{1,14,15,16} Riga-Fede can be managed in some cases without extraction. Elimination of the sharp edges of teeth may 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.

CONCLUSION

The early diagnosis of this clinical picture is vital in order to avoid more extensive/ longer periods of pain and possible hospitalization. This condition may interfere with proper feeding which, in turn, may pose potential risks to infants due to nutritional deficiency. Consequently, proper dental management for these patients must be considered.

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