

Management of Riga-Fede disease: a case report

**Guillermo Machuca MD, DMD¹, Sonia Rodríguez DMD²,
María-Patrocínio Vargas DMD², Cira Suárez DMD² and
Pedro Bullón MD, DMD³**

¹Professor, ²Assistant Professor, Department of Special Patients and Periodontics; ³Professor and Chairman, Department of Oral Medicine and Periodontics; University of Seville, School of Dentistry, Spain

Abstract

The treatment of a case of traumatic ulceration of the tongue (Riga-Fede disease) is described. A 12 month-old female patient, with cerebral palsy, presented with a widespread non-cicatrised yellowish-white ulcer on the ventral surface of the tongue, which was causing pain and refusal to eat. A very conservative approach was adopted, based on the use of an acrylic lower mouth guard, the smoothing of the incisal edges of the lower incisors, and the topical application of an obtundant gel.

Key words: Riga-Fede disease, cerebral palsy, oral health

Introduction

Riga-Fede disease is a benign ulcerative process of the tongue and frenulum, as a result of repetitive trauma induced by the erupting primary lower central incisors. The lesion usually begins as an ulcerated area on the ventral surface of the tongue, which with repeated trauma, progresses to an enlarged, fibrous lesion with the appearance of an ulcerative granuloma (Elzay, 1983). To qualify as Riga-Fede disease it is necessary for the patient to be less than two years old; over this age the term should be oral traumatic granuloma. Although this lesion was first described in 1881, few cases have been published but a number of therapeutic options are available such as smoothing of sharp incisal edges, using topical agents for protection, composite restoration, use of mouthguard/acrylic splints or in extreme cases, tooth extraction. The present case report attempts to throw some light on an apparently simple problem, which is sometimes difficult to solve.

Case report

A 12 month-old patient was admitted to the Odontology Faculty in the University of Seville, Spain for diagnosis and treatment of her oral problems. The clinical history of the patient revealed the following findings: at birth there was a breech presentation, the umbilical cord around the neck, meconium stained liquid and vacuum-extractor delivery. The patient then went on to progressive neurological deterioration. Psychomotor development was abnormal. The patient presented with global, limited mobility and was unable to sustain a normal posture for her age, with poor



Figure 1. Traumatic ulcer on the ventral surface of the tongue and its proximity to the sharp edges of the primary lower central incisors.

control of her head. The infant has limited mobility in upper and lower limbs and was unable to put her hands in her mouth. There was a diagnosis of hypoxicischaemic encephalopathy.

The patient was referred for evaluation of a painful ulcer on the ventral surface of the tongue, including the lingual frenulum, of two months duration, during which she refused to eat, even refusing to drink water on occasions, leading to serious dehydration (*Figure 1*). The ulcer appeared as her mandibular incisors had erupted. The parents were extremely concerned. The patient was seen to have an exaggerated and continuous sucking reflex, with repeated thrusting of the tongue towards the exterior of

the oral cavity. This lingual reflex seemed to be the one that initiated the ulcer by moving the ventral surface of the tongue over the sharp edges of the semi-erupted lower incisors.

Differential diagnosis:

It is necessary to carry out a differential diagnosis to eliminate other soft tissue lesions that can be malignant, such as sarcoma (Eichenfield *et al.*, 1990). In the case presented, a biopsy of the lesion was not taken because the parents refused consent to sedate the patient. However the clinical presentation of the lesion indicated its benign nature.

It must be borne in mind that this lesion, which appears in early infancy, can be a manifestation of important neurological lesions, such as familial dysautonomia syndrome and Lesch-Nyhan syndrome, (Zaenglein *et al.*, 2002). The latter was not part of the differential diagnosis in this case because Lesch-Nyhan syndrome only affects boys.

Treatment

The usual treatment is initial smoothing of the incisal edges of the lower incisors. Since the lesion was extremely painful and tongue thrusting reflex uncontrollable, it was decided to fabricate an acrylic mouth guard, so as to reduce the effect of friction of the tongue on the teeth. The impression was taken with a shortened individualised tray, packed with an irreversible hydrocolloid. During this procedure, the infant was held by her mother with her head inclined downwards in order to protect the airway. The tongue protector was fabricated from a soft silicone relining material used for dentures. The protector covered the mandibular primary central incisors and the alveolar ridge, reducing the contact of the dorsum of the tongue (Figure 2). The parents were advised to insert the appliance daily, for the maximum time the patient would tolerate it. In addition, a topical application of a cicatrising gel containing



Figure 3. Patient's presentation, on review 2.5 months after initial presentation.

polyvinylpyrrolidone, maltodextrin, propylenglicol, PEG-40, xanthan gum, potassium sorbate, sodium benzoate, sodium hyaluronate, glycyrrhetic acid, vera aloe and sacarine (Alocclair gel®, Sunstar Lausanne, S.A.) was prescribed three times a day. The clinical improvement was evident within one week; the patient was pain-free and had begun to feed normally, so it was decided to discontinue the cicatrising gel. At the end of a three-week follow up period, the ulcer had cicatrised completely (Figure 3), so it was also decided to discontinue the mouth guard. Although the tongue thrusting reflex remains, there have been no signs of relapse.

Discussion

On the basis of the presenting evidence, the patient was diagnosed with a sublingual traumatic ulcer, or Riga-Fede disease. Although there is little published on this type of lesion, it is probable that it occurs more frequently than is given to understand from the literature. Despite a seemingly trivial occurrence, it has a series of consequences that makes it important for oral health professionals to be aware of (Zaenglein *et al.*, 2002): intense pain, which linked with the incapacity to communicate, whether due to age and/or intellectual impairment, results in failure to eat and drink and consequent dehydration (Slayton, 2000). The timely diagnosis of this clinical picture by the oral health clinician will avoid more extensive periods of difficult feeding/drinking and even hospitalisation.

With respect to treatment, it is worth noting that while some authors (Eichenfield *et al.*, 1990; Slayton, 2000) recommend very aggressive initial treatment, with the removal of the lesion, it is nonetheless true that these same authors report relapses of the same cases. For this reason, a



Figure 2. Acrylic mouth guard for protection, externally secured by means of dental floss and a clip.

very conservative treatment was recommended. Although cerebral palsy was present there was no history of abnormal sensory perceptions. Moreover, in this case, due to the tongue thrusting habit, if the sharp incisal edges of the semi-erupted primary incisors had not been smoothed (Kozai *et al.*, 1998), the clinical picture would have been repeated. The protecting mouth guard used in the first phase of treatment enabled the lesion to heal fully (Kozai *et al.*, 1998). The mouth guard was not cemented in place because a deterioration of breathing function in children of less than 20Kg or 130cm has been reported (Ogasawara *et al.*, 1995). The cooperation of the infant was very poor because the mouth guard was easily dislodged by the tongue. Other authors solve this same problem by placing composites over the edges of incisors so obtaining a similar effect (Goho, 1996); in the present case, using a composite restoration to cover the lower incisors to achieve a smooth surface, was not recommended due to the difficulty of isolating the lower incisors in order to bond them. As other authors have described previously (Terzioglu *et al.*, 2002), if the situation is not resolved quickly, it is necessary to resort to the extraction of the incisors.

Lastly, the use of the analgesic gel to aid in wound healing (Alocclair gel[®], Sunstar Lausanne, S.A.) undoubtedly accelerated the healing process. Other authors have described other types of preparations, such as Kenalog in Orabase or Hydroxypropyl cellulose film for these lesions. (Slayton, 2002; Terzioglu *et al.*, 2002). The product employed was easy to apply in an uncooperative patient and the desired outcome achieved.

Conclusions

The early diagnosis of this clinical picture is vital in order to avoid more extensive/ longer periods of pain and possible

dehydration. The simple conservative treatment proposed avoided other more aggressive and mutilating treatments. Furthermore, this treatment can be of use in establishing the differential diagnosis. If healing is not apparent after a few days, biopsy is recommended.

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Address for correspondence

Prof. Dr. Guillermo Machuca
 Department of Special Patients and Periodontics,
 University of Seville,
 School of Dentistry,
 Clínica Odontológica Universitaria, C/ Avicena s/n,
 41009 Seville,
 Spain.
 Email: gmachuca@us.es