

Case for diagnosis

Caso para diagnóstico

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CASE REPORT

A 38-year-old Caucasian man was referred to our Dermatology Department for a painless ulcer on the left border of his tongue that had appeared 2 weeks before. The lesion was 2 cm in diameter and had an infiltrated base, elevated borders and the cen-



FIGURE 1: Large tongue ulcer with an infiltrated base, elevated borders and the center covered with fibrinous exudate

ter covered with fibrinous exudate (Figure 1). There was no palpable regional adenopathy and the remainder mucocutaneous examination was normal. Three weeks before, the patient had suffered an ischemic stroke with right hemiparesis conditioning hypoesthesia and left lateral tongue deviation. He was a heavy smoker and denied sexual risk behavior. Serologic tests for HIV 1 and 2 and syphilis were negative, as it was the search for *Treponema pallidum* by PCR in a biopsy sample.

Histological examination of a biopsy specimen from the border and the bottom of the ulcer was performed, showing reactive changes of the epithelium, ulceration with granulation tissue and scant presence of eosinophils, consistent with traumatic ulcer. There was complete recovery of neurological deficit and the lesion cleared after 2 months, without recurrence at follow up 10 months later (Figures 2 and 3).

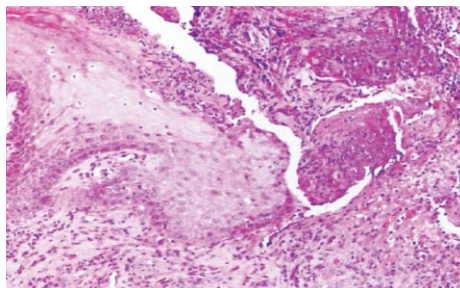
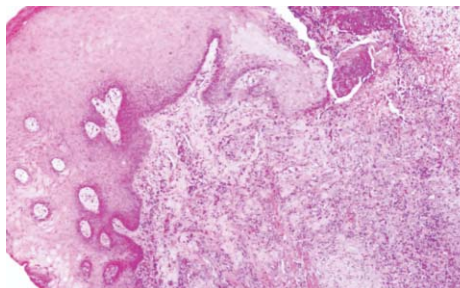


FIGURE 2: Histological examination of the border and bottom of the ulcer showing reactive changes of the epithelium, ulceration with granulation tissue and scant presence of eosinophils (Hematoxylin eosin, left - 40x, right - 100x)



FIGURE 3: Resolution of the lesion after neurologic recovery (left - after one month, right - after 2 months)

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DISCUSSION

Injuries to the oral mucosa may cause traumatic ulcers that occur most commonly on the tongue, lips or buccal mucosa. When the causative agent persists, the ulcer may become chronic, reactive and exophytic, resembling squamous cell carcinoma.¹ The ulceration may remain for years and can result in long-lasting tongue deformity.²

Riga-Fede disease (RFD) is a rare, benign disorder characterized by reactive ulceration of the oral mucosa, especially located on the tongue, and is commonly associated with repetitive dental traumatism. The designation of RFD applies specifically to children younger than 2 years with developmental or neurologic disorders causing hypotonia and lack of pain sensation. On microscopic examination, it reveals chronic ulceration and granulation tissue with mixed inflammatory cell infiltrate composed of lymphocytes, histiocytes, neutrophils and eosinophils. Early recognition of this entity is important because it may be a sign of an underlying neurological disorder.³ Similar clinical and histopathological findings have been reported in adults as oral traumatic granuloma or Riga-Fede-like disease usually caused by broken teeth or ill-fitting dental prosthesis.^{4,5} A broad variety of terms has been used to describe RFD, such as eosinophilic ulcer of the oral mucosa, sublingual fibrogra-

nuloma, sublingual growth in infants, lingual traumatic ulceration, traumatic atrophic glossitis, traumatic granuloma of the tongue, and traumatic ulcerative granuloma with stromal eosinophilia.⁵ Treatment of the disorder consists in removing the causative agent or minimizing the trauma by applying protective acrylic over the offending teeth and modifying behavior.⁵

If ulcerating lesions of the oral mucosa persist, biopsy is mandatory to exclude neoplastic causes such as squamous cell carcinoma, which accounts for 90-95% of the oral mucosa malignancies and is localized mostly on the tongue, and also leukemia, lymphoma or granular cell tumor. Other causes should be considered in the differential diagnosis of oral mucosa ulcers, such as inflammatory causes (major aphthous ulcer, eosinophilic ulcer), infectious causes (primary syphilis, tuberculosis, fungal infection) and iatrogenic causes (chemotherapy, radiotherapy, chronic tongue ulcers in bisphosphonate-associated osteonecrosis of the jaws).^{6,7}

In our case, clinical and histological data, as well as the healing of the lesion after resolution of the traumatic factor related to neurological deficit, allowed the diagnosis of traumatic ulcer of the tongue, Riga-Fede-like.

This rare entity should be considered in the differential diagnosis of oral mucosa ulcers to avoid excessive diagnostic or therapeutic measures. □

Abstract: Riga-Fede disease is a rare, benign disorder characterized by reactive ulceration of the oral mucosa associated with repetitive dental traumatism. It was first described in children with neurologic disorders and is very rare in adults. This case report describes the occurrence of a large ulcer of the tongue, resembling squamous cell carcinoma, in an adult with hemiparesis. The lesion cleared after neurologic recovery. This case highlights the importance of considering this disorder in the differential diagnosis of oral mucosal ulcerations.

Keywords: Mouth mucosa; Nervous system diseases; Neurologic manifestations; Oral ulcer; Paresis; Ulcer

Resumo: A doença de Riga-Fede caracteriza-se por ulceração reativa da mucosa oral associada ao traumatismo dentário repetitivo. Foi inicialmente descrita em crianças com déficit neurológico, sendo muito rara em adultos. O presente caso descreve o aparecimento de uma úlcera grande, semelhante ao carcinoma espinocelular, na língua de um adulto com hemiparesia. A lesão teve resolução completa após a recuperação neurológica. O caso salienta a importância de considerar esta doença no diagnóstico diferencial das úlceras da mucosa oral.

Palavras-chave: Doenças do sistema nervoso; Manifestações neurológicas; Mucosa bucal; Paresia; Úlcera; Úlceras orais

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