



Riga-Fede disease like ulcers in old age: A case report

İleri yaşta Riga-Fede benzeri ülserler: Bir olgu sunumu

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Abstract

Riga-Fede disease (RFD) is a traumatic, reactive benign disorder characterized by persistent ulceration on the tip or ventral surface of the tongue, seen mainly in infants and children. Lesions tend to develop after the eruption of natal or primary incisors, resulting from repetitive traumatic damage due to backward and forward movements of the tongue over the lower incisors. A literature survey has revealed a very limited number of reported cases of RFD in adults. Herein we reported a 70-year-old female patient who developed RFD-like ulcers on the tongue and buccal mucosa during the previous two months, while under treatment of dental implants. Histopathological examination and direct immunofluorescence of the ulcers and periulcer area did not yield a specific diagnosis. The lesions were resistant to systemic steroid treatment, however, after applying for a soft dental plate nightly for protection of the tongue and buccal mucosa, all ulcers completely healed in two months. With regard to the presented patient, we have reviewed the cases of RFD or RFD-like ulcers reported in adults and discussed the factors contributing to ulcer formation in our patient.

Keywords: Dental treatment, old age, oral ulcer, Riga-Fede disease, trauma

Öz

Riga-Fede hastalığı (RFH) esas olarak bebek ve çocuklarda sıklıkla dilin ucu veya alt yüzünde ortaya çıkan reaktif, travmatik, ülseratif benin bir hastalıktır. Lezyonlar neonatal, natal veya primer kesicilerin çıkışından sonra oluşma eğilimindedir ve dişlerin dil üzerinde tekrarlanan öne arkaya hareketinin sorumlu olduğu düşünülmektedir. Literatürde erişkin RFH olgularına çok ender rastlanmaktadır. Burada 70 yaşında bir kadın hastada diş implantları uygulanırken son iki aydır dilde ve yanak mukozasında ortaya çıkan RFH-benzeri ülserler bildirilmektedir. Ülser ve çevresinden yapılan histopatolojik ve direkt immünofloresan incelemeler özgün bir tanıya işaret etmemiş, lezyonlar sistemik steroid tedavisine cevap vermemiş, dili ve yanağı koruyan yumuşak gece plağı uygulamasıyla iki ay içinde tam düzelme olmuştur. Bu hasta dolayısıyla erişkinlerde bildirilen RFH veya RFH-benzeri ülser olguları gözden geçirilmekte ve sunulan olguda lezyonların gelişiminde rol oynaması muhtemel faktörler tartışılmaktadır.

Anahtar Kelimeler: Diş tedavisi, ileri yaş, oral ülser, Riga-Fede hastalığı, travma

Introduction

Riga-Fede disease (RFD) is a benign ulceration located frequently on the tip or ventral surface of the tongue, which is reactive to repeated trauma¹. This entity was first described in 1881 by the Italian physician, Antonio Riga. Since histologic

studies and additional cases were published by F. Fede in 1890, it has been subsequently known as RFD².

It is mainly seen in infants and small children, and there are very few reports of traumatic ulcers similar to RFD in adults¹⁻⁴.

Herein we report an elderly female patient with RFD-like ulcers and discuss the possible contributing factors.

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Case Report

A 70-year-old woman referred to our outpatient clinic for evaluation of the painful oral ulcers present for two months. She had been under treatment of dental implants for several teeth on the upper and lower chin, during the last nine months. She told that some of the implanted teeth on the upper chin repeatedly traumatized the tongue, as they were relatively loosened due to wearing of the temporary adhesive. She also declared bruxism at night. She was admitted to a primary care unit for oral ulcers and was treated with oral methylprednisolone 32 mg/day. After two weeks of treatment, no healing was observed. Her medical history revealed hypertension, diabetes mellitus, hypothyroidism, hypercholesterolemia, rheumatoid arthritis, and depression. Accordingly, she had been using insulin glargine, gliclazide, metformin, levothyroxine, olmesartan, atorvastatine, and diclofenac. Before the onset of oral ulcers, the patient had used duloxetine for one month, and stopped this medication two weeks after the development of ulcers. She had used various oral antiseptics and local anesthetics for symptomatic relief, and after using a solution comprising hydroxybenzenesulfonic acid, hydromethoxybenzenesulfonic acid, and sulfuric acid, she described severe burning, pain, desquamation and yellowish membranes on the ulcers. She denied smoking and alcohol consumption.

Dermatologic examination showed an oval, sharply circumscribed, deep ulcer of 3x2 cm in diameter, covered with a yellowish membrane and surrounded by a pinkish white halo (Figure 1 a). Similar but smaller and superficial ulcers were present on the left side of the tongue and on both buccal mucosae (Figure 1 b,c). No regional lymphadenopathy was detected. Other skin areas, genital mucosa, nails and hair were normal. Histopathologic examination of the ulcers showed slightly

proliferated non-keratinized stratified epithelium, subepithelial vascular proliferation and edema (Figure 1 d). Direct immunofluorescence examination performed on a biopsy specimen from the intact mucosa adjacent to the ulcer showed negative staining with immunoglobulin A (IgA), IgG, IgM, and C3. Laboratory tests yielded the following abnormal results: Fasting glucose 239 mg/dL, hemoglobin A1c (HbA1c): 12.2%, blood urea nitrogen: 38 mg/dL, gamma glutamyl transferase: 163 U/L, thyroid stimulating hormone: 0.002 µIU/mL, serum vitamin B12: 1001 pg/mL, erythrocyte sedimentation rate: 25 mm/h, Hb: 11.6 g/dL, hematocrit: 33.8%, leukocyte: 14.800/µL.

Based on the history of previous dental treatments in addition with histopathological and immunofluorescence findings, we considered the lesions as traumatic. All the systemic and topical agents used for the oral ulcers were stopped. The patient started to wear a soft dental night guard, prepared at our dental clinic specifically for her oral and dental anatomy. After one month, pain was alleviated, and ulcers started to heal. A control visit at the second month showed that all ulcers were healed with atrophic white scars (Figure 2 a,b,c). Written informed consent was obtained from the patient.

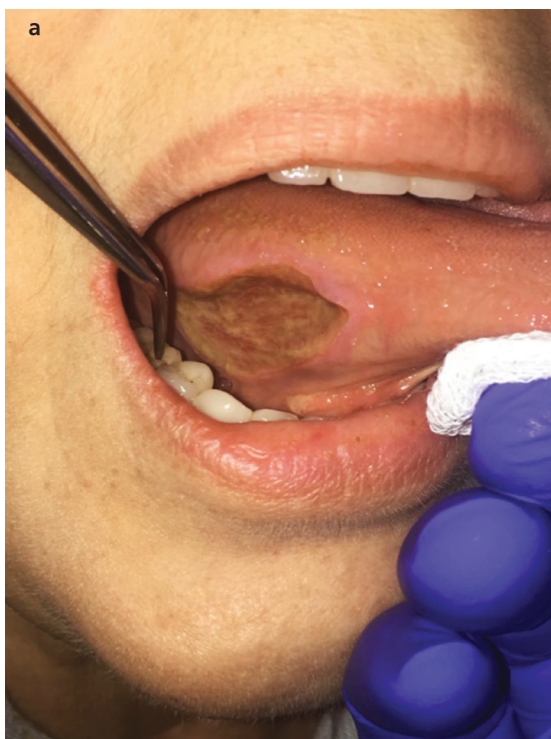


Figure 1. a) Deep and large ulcer covered with yellow-colored membrane on the right side of the tongue



Figure 1. b,c) Similar but smaller and shallow ulcers on the left side of the tongue and buccal mucosa

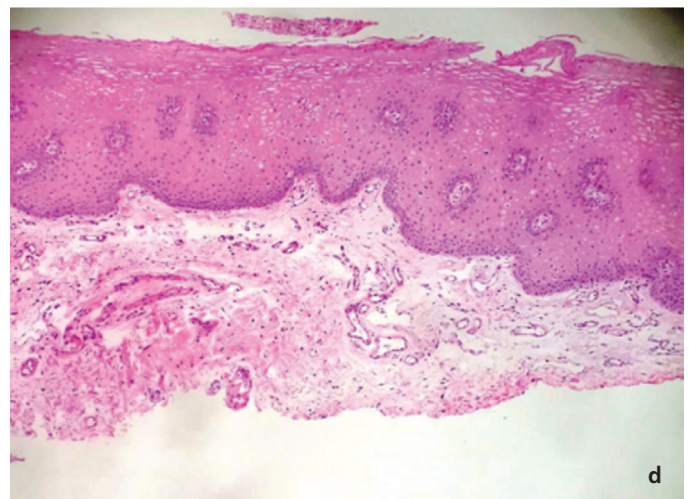


Figure 1. d) Histopathologic examination revealed slightly proliferated non-keratinized stratified epithelium, subepithelial vascular proliferation, and edema



Figure 2. a) The large ulcer healed with a prominently depressed whitish scar



Figure 2. b,c) Smaller ulcers completely healed with slightly atrophic scars

Discussion

Differential diagnosis of oral ulcers includes several diseases such as traumatic and infectious ulcers, tumors, immunobullous disorders, Behçet's disease, contact stomatitis, and reactive proliferative processes⁵.

Traumatic ulcers may develop due to physical, thermal or chemical effects. Acute traumatic ulcer is the most frequent cause of oral ulcers and mainly related with accidentally mucosal biting or trauma from food with sharp edges. These ulcers spontaneously heal in a few days⁵. Chronic traumatic ulcers are caused by repeated trauma of teeth with pointed or sharp edges, ill-fitting dentures and braces. The lesions are

located frequently on the ventral or lateral surfaces of the tongue and sometimes on the buccal mucosa^{5,6}.

Riga-Fede Disease (RFD) is a rare type of chronic traumatic oral ulcer seen after eruption of natal, neonatal or primary teeth. A "PubMed" research revealed only 61 published cases of RFD in infants and children by this time. The ulcer is caused by repetitive movements of the tongue on especially the lower, anterior incisors^{1,7}. The ulcer is mostly located on the ventral surface of the tongue or on the frenulum, however can occasionally occur on the lateral surface of the tongue, lip, or other mucosal areas^{1,8}. RFD may be associated with several neurologic and developmental disorders including cerebral palsy, microcephaly, encephalopathy, Down syndrome³, Tourette syndrome, Lesh Nyhan syndrome, Riley-Day syndrome⁹, and macroglossia^{1,10}. If natal, neonatal or early erupted primary teeth are present, the onset of oral ulcer is between the first week-6 months of life. Approximately 1/4 of the cases have neurologic disorders, and in these patients ulcer appears usually later, after 6th and 8th months of life¹.

Dermatologic examination reveal an asymptomatic or painful, indurated, superficial or deep ulcer with a white or yellowish fibrinopurulent membrane on its base^{1,3}. Perilesional erythema and raised white borders peripheral to ulcer are usually present⁷⁻¹⁰. RFD is commonly presents as a solitary ulceration¹. Histopathological evaluation shows ulcerated mucosa with granulation tissue, submucosal mixed inflammatory infiltration with lymphocytes, macrophages, mast cells and many eosinophils^{1,11}. After the elimination of trauma, ulcer heals with or without scarring, according to the degree of tissue damage^{1,3,7-11}.

Although the histopathologic features are identical to RFD, the causes of oral traumatic ulcers in the adults differ being related to the presence of broken teeth or ill-fitting prosthetic material. Up to date, there are only 3 adult cases reported under the heading of RFD or RF-like ulcers. One of them was a 70-year-old woman who presented with an ulcer on the lateral surface of the tongue caused by an ill-fitting denture. Histopathologic examination revealed leukoplakia without dysplasia, and the ulcer healed in two weeks after removal of the denture⁴. The other case was an 38-year-old man who developed hypoesthesia and left lateral tongue deviation after ischemic stroke and right hemiparesis. Repetitive trauma due to neurologic deficit led to a large ulcer located on the left border of the tongue. Histopathologic examination showed reactive changes of the epithelium, ulceration, granulation tissue, and sparse eosinophils. The ulcer healed completely in two months after neurologic recovery¹². The third case was a 40-year-old man with advanced stage of AIDS, who had a painful ulcer on the dorsal surface of his tongue, present for two months. Physical examination revealed a broken tooth above the ulcer. Histopathologic findings included granulation tissue surrounded by eosinophils. With the aid of a protective tooth device, the ulcer healed after four months¹³.

Though not designated as RFD or RFD-like ulcers, there is another case report of chronic tongue ulcer related to mechanical dental trauma. This case, a 36-year-old male patient, had an ulcer on the left side of the tongue caused by recurrent biting due to crossbite malocclusion. Histopathological evaluation of the ulcer showed acanthosis and mild epithelial dysplasia. After a specific tongue-retaining device, his chronic ulcer resolved within a few weeks¹⁴.

In our patient the duration, localization and clinical features of the ulcer were consistent with those described for RFD or RFD-like ulcers described in adults. For our patient we preferred to use the term RFD-like ulcers, due to the presence of multiple lesions, while RFD

mostly represents with a solitary ulcer. Histopathologic examination revealed absence of prominent eosinophilic infiltration similar to the abovementioned patients with adult onset RFD. Rapid improvement after protection with soft dental night guard supported the role of trauma.

Eosinophilic ulcer of the tongue (EUT), which is regarded by some authors as RFD seen in adults, should be considered in the differential diagnosis of the ulcers in our case. This entity has several synonyms including sublingual fibrogranuloma, traumatic granuloma of the tongue, and traumatic ulcerative granuloma with stromal eosinophilia. It is a rare, benign ulcer of unknown etiopathogenesis, most frequently seen in patients aged between 50-70 years. Mechanical trauma caused by accidental bite, sharp teeth or hard food plays an important role in about 35-40% of reported cases. EUT is characterized by a single ulcer, however, cases with multiple lesions, though very rarely, have been reported. The tongue is the most common location, but the lips, buccal mucosa, and palate may also be affected. The clinical features of the ulcer are closely similar to RFD¹⁵. Histopathologic examination demonstrates a dense, eosinophil-rich dermal inflammatory infiltrate. Plasmacytes, mast cells, macrophages, CD30+ large atypical mononuclear cells and small lymphoid cells may be scattered in this mixed infiltrate. A deep infiltrate involving submucosa is a typical finding for EUT, which may even extend to muscles and salivary glands. The rapidly developing ulcer persists for several weeks, and then spontaneously heal. Topical and systemic steroid may have beneficial effects^{15,16}.

Our case had an apparent history of trauma, the histologic features of the ulcers were not compatible with EUT, and the lesions were resistant to systemic steroids. We therefore considered this case as RFD-like ulcers seen in an elderly, similar to the previous reported cases under this title, instead of the diagnosis of EUT.

Oral ulcers related with bruxism have been described rarely, especially in children with neurologic disorders¹⁷. Bruxism has been observed in up to 14% of the patients under treatment with antidepressants including paroxetine, venlafaxine, and duloxetine, as an adverse effect¹⁸. Our patient declared bruxism and use of duloxetine before the onset of the oral ulcers. We suggest that repetitive mechanical trauma due to bruxism, and duloxetine treatment might have contributed to the ulcer development. In addition, the patient described an intense mucosal reaction similar to a chemical burn after application of an oral solution containing sulfonated phenol compounds and sulfuric acid which is used for chemical cauterization of oral aphthae¹⁹. It may be speculated that this reaction may have aggravated the severity and course of the ulcers.

Traumatic oral ulcers caused by ill-fitting dentures and pointed teeth can be seen in 2-15% of the elderly²⁰. Therefore it is interesting that RFD or RFD-like ulcers have been rarely reported in adults. An explanation for this discordance may be that the majority of these ulcers are superficial, small and transient. Another reason may be the insufficient awareness of this entity among the physicians.

Though RFD is a traumatic ulcer seen in infants and children, it should be noted that similar lesions may develop in adults.

Ethics

Informed Consent: Written informed consent was obtained from the patient.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.T.M., K.D., K.Ö., Concept: A.T.M., K.D., K.Ö., Design: A.T.M., Data Collection or Processing: K.D., K.Ö., Analysis or Interpretation: A.T.M., K.D., K.Ö., Literature Search: A.T.M., Writing: A.T.M.

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Kaynaklar

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