Meningitis and Riga-Fede Disease: An Unusual Condition

ABSTRACT

Background Riga-Fede disease (RFD) is a benign inflammatory disorder characterised by the appearance of a traumatic ulceration of the oral mucosa. Early detection of RFD and its adequate management are very important.

Case report The authors present an unusual case of RFD with concomitant Staphylococcus aureus meningitis. A 36-day-old female infant was referred to the emergency room of the Hospital of the University of Siena for a 4-day history of high fever. Clinical evaluation revealed the presence of lingual ulceration caused by natal tooth. Few hours later, clinical manifestations were overshadowed by neurological symptoms. The cerebrospinal fluid examination showed the presence of Staphylococcus aureus. The wound healing after extraction of the tooth and the antibiotic therapy have been important for the resolution of this case. A conservative approach is preferable for natal teeth, but in this case the extraction was suggested since a more radical treatment was more likely to avoid major complications.

Keywords Meningitis; Natal teeth; Neonatal teeth; Traumatic granuloma; Traumatic ulcer.

Introduction

Riga-Fede disease (RFD) is a rare disorder described in 1881 by Antonio Riga and in 1890 by Francesco Fede. RFD is also known as “traumatic oral granuloma” or “traumatic eosinophilic ulcer” because of the presence, in histological sections, of a peculiar inflammatory infiltrate with abundant eosinophils [Zaenglein et al., 2002].

The disease occurs in newborn and infant children and is commonly associated with the eruption of natal or neonatal teeth, but it can also occur at a later time, when normal eruption of deciduous teeth occurs. In particular, we use the definition of RFD in cases that occur in children under two years of age whereas for the other cases we usually use the term of “oral granuloma” or “eosinophilic ulcer”. Any surface of the oral mucosa can be affected, but the most common location in the Riga-Fede disease is the ventral surface of the tongue. The lesion begins as a small ulcerated area and thereafter, because of the continuous trauma, becomes larger with irregular bottom and hardened edges. Chronic trauma represents the most important etiopathogenetic factor for the formation of the "eosinophilic" inflammatory infiltrate [Baghdadi, 2001]. We report a RFD case diagnosed through history and clinical features together with its clinical management.

Case report

A 37-day-old female infant was referred to the emergency room of the University Hospital of Siena for a 4-day history of fever (38.6°C). The parents reported that the child had inadequate nutrient intake, difficulty in suckling, and bleeding on the ventral surface of the tongue every morning. Clinical evaluation showed the presence of an erythematous plaque of 1 cm diameter on the ventral surface of the tongue associated with the primary lower left central incisor (Fig. 1). All routine laboratory tests including biochemical analysis, complete blood counts, and immunological study, were found to be normal.

Few hours later there was a rapid deterioration of the general conditions. The infant became drowsy and hyporeactive to stimuli, although pupillary reflexes to light and accomodation were preserved. She also exhibited increased fever (39.5°C) and heart rate (198...
FIG. 2 Reduction of the lesion after 1 week.

beats/min), and bilateral crackles. No cardiac murmurs and no abdominal tenderness were found. New blood tests showed an increase of C-reactive protein (17mg/dl), erythrocyte sedimentation rate (60 mm/h), blood glucose (144 mg/dl) and neutrophil cells (81.4%). Few hours later, the clinical manifestations were dominated by neurological symptoms such as trunk hypertonia and downward deviation of the eyes (“sunset eyes”).

Because of persistent systemic signs (especially fever) and of a clear neurological picture, a suspicion of meningitis was raised and a lumbar puncture was performed. At the cerebrospinal fluid (CSF) examination, the cell count was 5,000/mm², with a prevalence of polymorphonuclear leukocytes, glucose was 40 mg/dl and the proteins were 250 mg/dl; bacteriological analysis of the CSF sediment showed the presence of the Staphylococcus aureus. Testing for bacterial sensitivity was done, and an intravenous antibiotic (Ceftriaxone 50 mg/kg/day) was administered. Because the oral lesion could result in dehydration and inadequate nutrient intake for the infant, dental consultation was required. Considering the high degree of mobility of the tooth, which caused tongue ulceration, extraction was performed to eliminate the associated trauma.

Twenty-four hours later the overall health status of the patient greatly improved, with reduction of fever. One week later the ulcer was significantly reduced (Fig. 2); three weeks later it had completely healed.

Discussion

In RFD the lesion begins as a small ulcerated area which, due to the continuous trauma, becomes larger with irregular bottom and hardened edges. The differential diagnosis of oral ulcers includes bacterial (primary syphilis and tuberculosis) and fungal infections (ulcerative candidiasis), recurrent aphthous stomatitis, allergies, immunologic diseases, genetic disorders, haematological disorders such as agranulocytosis or pernicious anaemia and malignant conditions [Baroni et al., 2006; Toy, 2001; Campos-Munoz et al., 2006; Schmitt-Kopple et al., 2002]. Early diagnosis of RFD is recommended, because ulceration of the tongue may also be due to serious underlying neurological disorders [Eichenfield et al., 1990; Baghdadi 2002; Mangano et al., 1994; Toy, 2001]. In general, biopsy is required for definitive diagnosis of RFD [Zaenglein et al., 2002; Baghdadi, 2001]. In this case considering the age of the infant, the particular site of the lesion and the clinical features typical of RFD, the histopathological examination was not performed. The positive results obtained after treatment ruled out other diagnostic hypotheses.

Early detection of RFD and its adequate management are very important. Ulceration induced by continuous trauma is likely to interfere with proper nutrition and therefore causes delay in growth. Moreover, waiting for the lesion to heal spontaneously may, in the long run, result in cleft deformity, which is a permanent mutilation [Terzioglu et al., 2003]. Several management options for RFD have been described: oral disinfectant, corticosteroids, smoothing the incisal surface of the tooth, the construction of a protective shield or teething ring [Slayton, 2000]. Conservative treatment is preferable to dental extraction, because tooth removal may delay the eruption of permanent tooth, but healing of the lesion is slower. In the case presented here extraction of the lower incisor has been suggested as a more radical treatment choice to avoid major complications and to minimise the associated trauma. The urgency of the case dictated the need for a rapid resolution of the oral lesion that interfered with proper feeding and put the infant at risk for nutritional deficiencies thus worsening the complex clinical state. Besides, the ulceration of the tongue was the only wound present on patient and it was also suspected to be the origin of the bacterial infection. Staphylococcus aureus is a facultative anaerobic, Gram-positive coccus, it is frequently part of the skin flora and so we hypothesized that during breast-feeding the ulcer had allowed the spread of the bacterium. Extraction of the tooth and the consequent healing of the ulcer together with appropriate intravenous antibiotic therapy were crucial for the solution of this case.

References