Riga Fede Syndrome: Review of Literature with Report of Three Cases
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Abstract
Riga-Fede disease is a chronic traumatic ulceration presenting on the ventral surface of the tongue in neonates and infants, frequently associated with natal, neonatal, or primary lower incisor teeth. Treatment option is either conservative or extraction of natal and neonatal tooth. In the present three cases extraction was done because of difficulty in breast feeding and fear of aspiration due to the mobility of the teeth. At the follow up of all cases the lesion had resolved and the infant was feeding normally.

Key Words: Deciduous; Mouth Diseases; Natal Teeth; Oral Ulcer; Riga-Fede; Stomatitis.

Introduction:
Riga-Fede disease (RFD) is a chronic traumatic ulceration presenting on the ventral surface of the tongue in neonates and infants, frequently associated with natal, neonatal, or primary lower incisor teeth.1,2 The lesion was first described in 1881 by Italian physician Antonio Riga and in 1890 by Fede who performed histological studies of the lesions.3 It has therefore become known as Riga-Fede disease. A variety of other terms that have been applied to this lesion includes sublingual growth in infants, lingual traumatic ulceration, traumatic atrophic glossitis, traumatic granuloma of the tongue, traumatic ulcerative granuloma with stromal eosinophilia, and sublingual fibrogranuloma. It may interfere with proper sucking and feeding and put the neonate at risk for nutritional deficiencies.4 The exact aetiology of natal or neonatal teeth is unknown, it is thought be due to infection, febrile states, trauma, malnutrition, superficial portion of the tooth germ, hormonal stimulation and maternal exposure to environmental toxins.5

Case reports
Case 1:
A twenty day old male infant was referred to the Department of Pedodontics and Preventive Dentistry by his paediatrician with the chief complaint of a tooth present in the lower anterior region since birth. Family history was non-contributory. Intraoral examination revealed an ulcerated lesion on the anterior border of the tongue along with a tooth like structure present in the mandibular anterior region with grade III mobility (Fig 1a). The tooth was extracted because of difficulty in breast feeding and fear of aspiration due to its severe mobility. Extracted tooth showed well developed crown and a short root. (Fig 2a) The patient was recalled after two weeks and on examination a complete and uneventful healing of the extraction socket was observed along with healed ulcer (Fig 1b). Further it was reported by parents that, he was feeding normally without any post-operative complications.

Case 2:
The parents of fourteen days old child reported to Department of Pedodontics and Preventive Dentistry with chief complaint of ulcerated area on the anterior border of the tongue of their child. The mother complained of child exhibiting pain during suckling and could not be able to nurse the child. Clinical examination revealed a whitish tooth like structure in the anterior region of mandibular arch, exhibiting grade I mobility (Fig 1c). On palpation of involved area pain was felt. Examination of the rest of intraoral mucosa revealed no other lesion. The cause of this ulceration was due to natal tooth. Radiographic examination showed that a normally developing succedaneous tooth germ was present below the tooth like structure (natal tooth). Hence, extraction of teeth was chosen as treatment of choice over more conservative treatment which shows slower healing. Extraction of the offending natal tooth was carried out under topical anaesthesia. Extracted tooth showed well developed crown and a very short root. (Fig 2b) On one week follow-up it was
observed that child was feeding normally and the ulcer showed signs of healing (Fig 1d).

**Case 3:**
A 24 day old male infant was referred to the Department of Pedodontics and Preventive Dentistry for evaluation with complaint of two teeth in the lower jaw since birth and difficulty in breast feeding. Oral examination revealed an ulcerated lesion on the ventral surface of tongue along with bilateral natal teeth in mandibular anterior region (Fig 1e). The tooth exhibited an opaque whitish coloration with grade I mobility. The crown size and the appearance of the gingiva seemed to be normal. A diagnosis of natal tooth in association with Riga-Fede disease was made. Examination of the rest of intraoral mucosa revealed no other lesions. Radiographic examination revealed a natal tooth in the mandibular anterior region, devoid of root portion along with a normal developing primary lower incisor tooth germ below it. The medical history was non-contributory. As the tooth was the suspected cause of the chief complaint, it was extracted under topical anaesthesia. Extracted tooth showed well developed crown and a very short root. (Fig 2c) Two weeks later, on examination a complete and uneventful healing of the extraction socket was observed along with healed ulcer (Fig 1f). The baby appeared to be much more contented and the mother reported that he was feeding normally.

![Figure 1: Nata...](image1)

**Discussion**
Several terms have been used in the literature to designate teeth that erupt before the normal time, such as congenital teeth, foetal teeth, precocious teeth, and dentitia praecox. According to the definition

![Figure 2: Extracted...](image2)
presented by Massler taking only the time of eruption as reference, natal teeth are those observable in the oral cavity at the birth and neonatal teeth are those that erupt during the first 30 days of life. They are more common in mandibular arch than in the maxillary arch and more commonly occur in the incisor region rather than the canine and molar regions. The eruption of more than two natal teeth is rare but few cases of multiple natal teeth have also been reported in the literature. The incidence of natal teeth is usually quoted in the range of 1:2000 to 1:3500 live births. The presence of natal and neonatal teeth is definitely a disturbance of biological chronology whose aetiology is still unknown. It has been related to several factors, such as superficial position of the germ, infection or malnutrition, febrile states, eruption accelerated by febrile incidents or hormonal stimulation, hereditary transmission of a dominant autosomal gene, osteoblastic activity inside the germ are related to the remodelling phenomenon, and hypovitaminosis.

Spouge and Feasby have suggested that clinically, natal and neonatal teeth are further classified according to their degree of maturity
1. A mature natal or neonatal tooth is one which is nearly or fully developed and has relatively good prognosis for maintenance.
2. The term immature natal or neonatal tooth, on the other hand, refers to a tooth with incomplete or substandard structure; it has a poor prognosis.

Hebling classified natal teeth into four categories depending upon the appearance of each natal tooth into the oral cavity as the teeth emerge into the oral cavity:

a. Shell-shaped crown poorly fixed to the alveolus by gingival tissue and absence of a root;

b. Solid crown poorly fixed to the alveolus by gingival tissue and little or no root;

c. Eruption of the gingival margin of crown through gingival tissue.

d. Edema of gingival tissue with an unerupted but palpable tooth

Riga-Fede disease describes a traumatic ulceration presenting on the ventral surface of the tongue in neonates and infants typically between one week and one year of age. Although usually associated with natal or neonatal teeth, it may also occur in older infants after the eruption of the primary lower incisors. Simple chronic traumatic ulcerations occur most often on the tongue, lips, and buccal mucosa, the sites usually injured by the dentition. Most reported Riga-Fede have been on the anterior ventral surface of the tongue. However, the dorsal surface may also be affected. Typically the lesion begins as an ulcerated area on the ventral surface of the tongue with repeated trauma; it may progress to an enlarged, fibrous mass with appearance of an ulcerative granuloma. It may interfere with proper suckling and feeding and put the neonate at risk for nutritional deficiencies. In such instances, dental intervention may be required.

Riga-Fede disease often heralds an underlying developmental or neurologic problem, including familial insensitivities to pain. It has been well described in association with familial dysautonomia. Other neurological disorders related to self-mutilation, such as Lesch-Nyhan syndrome and Tourette’s syndrome. The other syndromes include Adrenogenital syndrome, Cleft lip and palate, Craniofacial dysostosis, Craniosynostosis syndromes, Ectodermal dysplasia, Ellis-van Creveld syndrome, Epidermolysis bullosa simplex, Hallerman-Streiff syndrome, Jadassohn-Lewandowsky syndrome, Multiple stacystoma, Pallister-Hall syndrome, Pfeiffer syndrome, Pierre-Robin syndrome, Polydactyly type II, Rubinstein-Taybi syndrome, Sotos syndrome, Steatocystoma multiplex, Van der Woude syndrome, Walker-Warburg syndrome and Wiedeman-Rautenstrauch syndrome. Therefore, it is important to note that this lesion may be the initial presentation of some serious underlying medical problems.

The various complications related to natal and neonatal teeth include discomfort during suckling, irritation and trauma to infants’ tongue, sublingual ulceration (Riga-Fede disease) laceration of the mother’s breast and risk of aspiration of the mobile teeth. Prolonged gingival irritation from natal or neonatal teeth may cause localized inflammation of the gingiva or fibrous hyperplasia.

Treatment should begin conservatively and should focus on eliminating the source of trauma which is usually the natal or neonatal tooth in case of Riga-Fede disease. Failure to diagnose and properly treat this lesion can result in dehydration and inadequate...
intake for the infant. In case of mild to moderate irritation to the tongue, conservative treatment such as smoothing the incisal edge of the natal or neonatal tooth with an abrasive instrument is advocated.\textsuperscript{18} Alternate therapy would be to bind a small increment of composite to the incisal edges of the tooth.\textsuperscript{11} Extraction may be needed to alleviate feeding difficulties or complications like Riga-Fede disease. Extraction may also be indicated if child’s age is ten days or above and child has appropriate amounts of Vitamin K in the blood. Otherwise prophylactic administration of vitamin K (0.5 - 1.0 mg, I,m) 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.\textsuperscript{19} In most of the cases it is observed that the ulcer resolves uneventful on the removal of the tooth and rarely some intervention is required specifically for the treatment of the ulcer.

In the present cases extraction was done because of difficulty in breast feeding and fear of aspiration due to the mobility of the teeth. At the follow up of all cases the lesion had resolved and the infant was feeding normally.

Conclusion
It should be noted that natal and neonatal teeth are conditions of fundamental importance to pedodontist and paediatrician, as their presence may lead to numerous complications. The child must be monitored to restore the function and aesthetics of the normal primary dentition.

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