Riga–Fede Disease associated with Natal Tooth

Deepak Khandelwal, Namita Kalra, Rishi Tyagi, Amit Khatri, Dhiraj Kumar, Komal Gupta

ABSTRACT

Eruption of tooth at about 6 months of age is a significant milestone in a child’s life and is an emotional event for parents. However, a tooth present in the oral cavity at the time of birth can lead to a lot of delusions. Natal and neonatal teeth are conditions of fundamental importance not only for a dental surgeon, but also for a pediatrician since their presence may lead to numerous complications. Early detection and treatment of these teeth are important as they may induce tongue deformity, dehydration, inadequate nutrients, and growth retardation. Riga–Fede disease (RFD) is a reactive mucosal disease due to repetitive trauma of tongue by anterior primary teeth during forward and backward movements. This article reports a case wherein a natal tooth has led to the development of ulcer on ventral surface of tongue in a 4-week-old infant.

Keywords: Natal teeth, Riga–Fede disease, Ulceration.

INTRODUCTION

Developmental milestones are anxiously awaited by every parent, particularly in the first year of their child’s life. One such milestone is the eruption of their child’s first tooth, which starts at, on average, around 6 months old.1 The presence of teeth at birth or within a month after delivery is rare. They are classified into two groups according to the time of eruption, as natal teeth—those present at birth—and neonatal, the teeth that erupted in the first 30 days of the child’s life.2 They can cause feeding problems, ulceration of the ventral part of tongue and frenulum, loosening, and risk of aspiration. About 90% of these teeth are observed to be primary and only 10% are supernumerary.3 The development of traumatic ulcers on the ventral surface of the tongue, often caused by repetitive traumatic injuries due to backward and forward movements of the tongue over the mandibular anterior incisors, is known as RFD.2,4 Trauma to the tip or ventral surface of the tongue is a complication of natal and neonatal teeth, and occurs in 6 to 10% of cases of natal teeth.5

The RFD was first identified by Riga, an Italian physician, in 1881 and described in 1890 by Fede, who performed the histological studies of the lesion. The RFD can be asymptomatic or occasionally occurs with pain, with both sexes being equally affected. Although common in infants (between 1 week and 1 year of age), it has also been reported in older patients and patients with acquired immunodeficiency syndrome.4

The lesion initiates as an ulcerated area, which upon repetition of trauma can evolve into an enlarged fibrous mass with ulcerative appearance. Lesion makes it difficult for the infant to suck and feed, putting the baby at risk of nutritional deficiencies.2

The prevalence of natal teeth was found to be of 1:2000 and neonatal teeth of 1:3500.6 Most common tooth involved is mandibular central incisors (85%) followed by maxillary incisors (11%).4 Because of its rare occurrence, in the past this anomaly of eruption was associated with superstition and folklore, being related to good or bad omens.7

Several methods of treatment for RFD have been reported. Historically, the most common has been extraction. However, it can be managed in some cases without extraction also.5

These teeth stimulate the interest of both parents and health professionals because of their clinical characteristics.7 Failure in diagnosis and proper treatment may result in dehydration and inadequate intake of nutrients by the baby, increasing the potential for infection at the site.2

This report aims to present the occurrence of RFD associated with natal tooth and the treatment approach in a 1-month-old infant.

CASE REPORT

A 4-week-old female infant was brought by the parents to the Department of Pedodontics and Preventive Dentistry, University College of Medical Sciences and Guru Teg
Bahadur Hospital with a complaint of a tooth present in the lower front region of the mouth and ulcer on tongue. The ulcer had been present for 14 days. The child was experiencing difficulty in feeding. Her parents reported that the child had inadequate nutrients intake due to difficulty in suckling and intermittent bleeding from the tongue. There was a clear association of the ulcer with a partially erupted tooth present in the lower incisor region (Fig. 1). The medical history revealed that she was healthy at birth with normal full-term delivery, but there was a mandibular anterior natal tooth present at the time of birth. The parents did not consult any dentist as neither the mother nor the child had any problem during feeding during the first 2 weeks. The mother reported that the child had been neither sleeping nor able to suck the breast normally since 2 weeks. Infant was unable to suckle the mother’s nipple for feeding. The tooth appeared well formed and was mobile. A familial history of natal teeth, involving one of her three siblings, was elicited. The family history for developmental disorders and congenital syndromes was found to be negative.

The intraoral examination revealed the crown of a natal tooth in the mandibular anterior region with grade II mobility and ulceration on the ventral surface of the tongue. The physical examination revealed a circular lesion of 3 mm diameter, covered with a gray-white fibrinous plaque located on the ventral surface of the tongue, in contact with the lower central incisors (Fig. 1). The intraoral mucosa revealed no other lesions. Hematological examinations including complete blood count and clotting test were done, and all the values were within normal range. A pediatrician consultation was made and 1 mL of vitamin K was administered intramuscularly to prevent excessive bleeding. The parents refused permission to perform a tongue biopsy. Therefore, histopathological examination could not be carried out. Diagnosis of RFD was made based on the history and clinical features.

The extraction of natal tooth was selected as treatment of choice, over more conservative treatments for the rapid resolution of the lesion and to prevent a potential teeth ingestion or inhalation by the infant with the consequent penetration into airways and lungs, should the teeth become dislodged during nursing procedures due to their great mobility (grade II).

With parental consent, the extraction was carried out under topical local anesthesia and was followed by curettage of the socket to prevent the development of the dental papilla cells ("residual natal tooth") to continue. The teeth showed normal size and shape, whitish opaque color, and absence of complete root formation (Fig. 2).

The complete healing of the tongue lesion took 2 weeks (Fig. 3). Patient was kept under regular follow-up. No complication or recurrence was observed.

**DISCUSSION**

Tooth eruption at about 6 months of age is a significant phase in terms of functional and psychological changes in the child’s life. The presence of teeth at birth or within a...
month after birth is a rare condition. The RFD involving ulceration of the underside of the tongue is a relatively uncommon complication of natal teeth. The RFD is a chronic traumatic ulceration presenting on the ventral surface of the tongue, frequently associated with natal, neonatal, or primary mandibular incisor teeth during continual protrusive and retrusive movements. The condition is most commonly observed in newborns and infants, and usually coincides with the eruption of the primary teeth; however, the symptoms can be observed immediately after birth with natal and neonatal teeth. Other complications of natal teeth may include possibility of swallowing and aspiration, injury to mother’s breast, and inconvenience during suckling. Kinirons described a peculiar situation of birth of a baby with a natal tooth and the presence of a sublingual ulcer seen immediately after birth probably caused by suction during intrauterine life.

In the present case, the patient had irritability and difficulties with breastfeeding because of the injury on the ventral surface of the tongue, which could eventually generate nutritional damage among other problems. Since the patient’s parent did not give permission to perform radiographic examination, the diagnosis was based on history and physical examination.

van der Meij et al stated that once the clinician is familiar with RFD, the history and clinical features are most often so typical that there is seldom any need for additional histopathological examination. The exact etiology of the early eruption of these teeth is unknown, but the most acknowledged etiology behind the tooth eruption before the chronological period is considered to be the superior placement of tooth germ, whereas literature also proposes role of inheritance as dominant autosomal trait, endocrine factors and poor maternal health, infections, nutritional deficiency, fever, endocrine disorders, superficial position of tooth germs, and osteoblastic activity in the area of dental germs.

The natal and neonatal teeth may be related to hereditary syndromes, such as congenital pachyonychia, Sotos syndrome, Hallermann–Streiff syndrome, chondroectodermal dysplasia, craniofacial dysostosis, syndrome of Wiedemann, and Meckel–Gruber syndrome. In the present case, the patient had no other congenital abnormalities, but there was the other case of natal teeth in the family.

Although usually associated with natal or neonatal teeth, it may also occur in older infants after the eruption of the primary lower incisors. Males are usually more affected with RFD. Ventral surface of tongue is the most common site of occurrence of the ulcer. Failure to diagnose the lesions can lead to tongue deformity, dehydration, and inadequate nutrient intake, resulting in poor development. Lesion is present as erythema surrounding a centrally removable yellow and fibrinopurulent membrane. Borders appear as a rolled white hyperkeratotic margin immediately adjacent to the ulceration. Based on these characteristics, the differential diagnosis of RFD should include myofibroma, granular cell tumor, pyogenic granuloma, agranulocytosis, electrical or chemical trauma.

The presence of natal and neonatal teeth may be a source of doubt about the treatment plan. The RFD is not an indication for extraction. The preferred treatment of RFD includes smoothing of rough incisal edges or the placement of composite resin over the incisal edges. Additional histopathological examination is recommended to rule out any other pathology. Early extraction of a primary natal tooth might lead to overcrowding of the permanent teeth, and esthetic, functional, and phonetic problems.

The decision to retain or extract a natal and/or neonatal tooth should be evaluated in each case, keeping in mind scientific knowledge, clinical common sense, and parental opinion after the parents are properly informed about all aspects involved in this situation. In RFD, although the aspect of the lesion might be impressive, its nature is relatively benign and can most often be treated with conservative measures only.

CONCLUSION

The presence of natal teeth is rare, but can cause RFD. Early diagnosis and treatment of these teeth are of utmost importance due to risk of aspiration, irritation, and trauma to soft tissues and even compromises the child’s ability to feed, which may result in malnutrition and dehydration in the infant.
REFERENCES