Traumatic Lingual Ulcer in a Child with Precocious Riga-Fede Disease: Case Report and Review of The Literature

Michele Callea[1]*, Pamela Armi[1], Mehmet Sinan Dogan[3], Dian Agustin Wahjuningrum[2], Francisco Cammarata Scalisi[4], Gaia Rosati[1], Roberta D'Avenia[1]

1. Pediatric Dentistry Unit, Meyer Children's University Hospital, Florence, Italy.
2. Department of Conservative Dentistry, Faculty of Dental Medicine, Universitas Airlangga, Surabaya, Indonesia
3. Pediatric Dentistry Department, Harran University, Faculty of Dentistry. Sanliurfa, Turkey.
4. Regional Hospital of Antofagasta, Chile.

Abstract

We report on a case of traumatic lingual ulcer in a child with Riga-Fede disease discussing the treatment and focusing on the importance of an early diagnosis of this entity and the possible concomitant clinical and genetic associations, reviewing the most recent literature in regard. It is caused by repeated forward and backward movements of the tongue over the lower incisors in newborns and infants.

It is most commonly associated with neonatal or natal teeth in newborns. These teeth can cause ulcers on the ventral surface of the tongue, lip, and the mother's breast characterizing the Riga-Fede disease. We reported a 2-month-old female who had an ulcerated area on the ventral surface of the tongue. Her mother complained of pain during breastfeeding and had to interrupt it. No biopsy was done in view of the particular site of the lesion and the clinical evidence of diagnosis. Based on these findings, the baby was diagnosed with Riga-Fede Disease.

Indeed, a clinical consultation has been required to extract her teeth under local anesthesia and healing of the ulcerated area was observed within two weeks. At a following control, two weeks later, the lesion was fully recovered and the infant restarted feeding normally.

Case report (J Int Dent Med Res 2022; 15(4): 1724-1726)

Keywords: Neonatal teeth, Riga Fede disease, natal teeth, lingual ulcer, follow up.

Received date: 20 July 2022 **Accept date:** 07 September 2022

Introduction

Riga-Fede disease (RFD) is an extremely rare, benign inflammatory disorder characterized by traumatic ulceration of the oral mucosa. It is caused by repeated forward and backward movements of the tongue over the lower incisors in newborns and infants.[1] A case of Riga-Fede disease with lesion in the upper jaw has also been reported.[2] It is most commonly associated with neonatal or natal teeth in newborns. Natal teeth are those present in the oral cavity at the child's birth. These teeth can cause ulcers on the ventral surface of the tongue, lip, and the mother's breast characterizing the Riga-Fede disease.

Corresponding author:
Michele Callea,
Pediatric Dentistry and Special Dental Care Unit
Meyer Children's Hospital, Florence, Italy.
E-mail: mcallea@gmail.com

A recent classification divides the disease in precocious Riga-Fede disease and late onset Riga- Fede disease depending; the precocious one occurring in the first six month of life, based on the presence of natal or neonatal teeth; the second one after the sixth month of age with the occurrence of the first dentition caused by neurodevelopmental disorders.[3] The treatment depends on the possible clinical manifestations: the tooth's mobility and the risk of aspiration or swallowing; whether it is causing interference in breastfeeding; breast and oral soft tissue injuries and the general child's health.[1,4-7] Riga-Fede disease could be associated with neurological and genetic disorders like Down Syndrome (OMIM 190685), Fabry disease (OMIM 301500), Niemann-Pick C disease (OMIM 257220), bilateral harlequin syndrome (OMIM 242500), unilateral Horner syndrome (OMIM 143000), hereditary sensory and autonomic neuropathy (HSAN) type IV, also known as congenital insensitivity to pain with anhidrosis (OMIM 256800), glutaric aciduria type 1 (OMIM 231670), Lesch Nyhan syndrome (OMIM 300322), Riley

day syndrome (OMIM 223900), Tourette syndrome (OMIM 137580) and ectodermal dysplasia-like syndrome with clinodactyly and oligodactyly.[3,9-14] Since it can clinically mimic multiple oral malignant and benign disorders, a correct and early diagnostic assessment is paramount.[1-14]

Case Report

A 2-month-old female was referred to the Unit of Pediatric Dentistry for Special Need of Meyer Children Hospital, Firenze, Italy, to evaluate an ulcerated area on the ventral surface of the tongue (Figure 1).

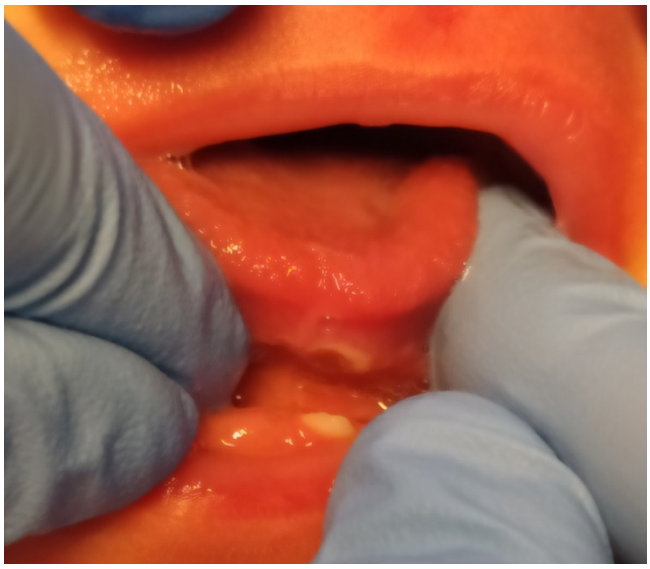

Figure 1. On the lower jaw is visible the crown of the neonatal tooth and an ulcerative lesion is present at the ventral surface of the tongue.

The lesion was elliptical, 1 cm by 1 cm in diameter. The mother complained of pain during breastfeeding and had to interrupt it. Oral examination revealed one tooth crown in the mandibular anterior region: a lower left mandibular tooth of whitish colour, cutting edge and shape resembling a primary incisor with mobility was evident. Examination of the rest of intraoral mucosa revealed macroglossia. Hypertelorism, flame angioma were noticed. The tooth was already present at birth, at 41 weeks of gestation. The mother reported in her clinical records history of oligohydramnios during her pregnancy. The maternal grandmother had a benign mammal neoplasia; paternal grandfather and paternal uncle were pace-maker carriers due to a severe cardiopathy.

No biopsy was done in view of the particular site of the lesion and the clinical evidence of diagnosis. Based on these findings, the baby was diagnosed with Riga-Fede Disease. Indeed a clinical genetic consultation has been required to understand and define a possible association between the clinical phenotype of the 2-month-old female and the occurrence of neonatal tooth. The tooth was extracted under local anesthesia (Figure 2) and healing of the ulcerated area was observed within two weeks. At a following control, two weeks later, the lesion was fully recovered and the infant restarted feeding normally.

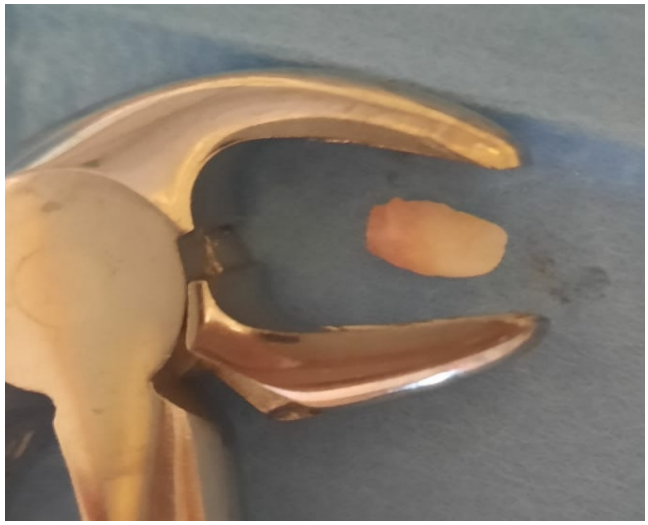

Figure 2. The extracted neonatal tooth.

Discussion

The neonatal tooth causing lingual lesion was first described by A. Riga, an Italian physician in 1881. Additional case and histologic studies were published by F. Fede in 1890. Ulceration of the ventral surface of the tongue in newborn babies or infants is most commonly related to natal or neonatal teeth, as illustrated in this case. In fact, major complication from neonatal teeth is the ulceration on the ventral surface of the tongue caused by tooth' sharp incisal edge.

Although uncommon, these teeth are found to have several clinical implications, from manifestations such as the ulceration of the tongue as above mentioned, to neuro-developmental disorders. This constant trauma may create an ulceration sufficient to interfere with proper feeding and put the neonate at risk for nutritional deficiencies.

Nonetheless, Riga–Fede disease may be associated with other syndromes as reported in literature hence it might be an alert suggesting further investigations to confirm or exclude the co-existence of other nosological entities such as genetic diseases.[3,9-15]

Conclusions

We report on a case of neonatal tooth and Riga-Fede disease, underlying the importance of an early clinical diagnosis, in order to avoid biopsies which may create discomfort in newborns and parents, with difficulties in feeding of newborns; we also would like to stimulate colleagues who daily works with newborns to address to Pediatric Dentists these patients. Furthermore, the occurrence of neonatal teeth should alert a multispecialistic team to provide an appropriate surveillance to manage the neurodevelopmental manifestations which may arise from Riga-Fede Disease. Nonetheless Riga–Fede disease may be associated with other syndromes as reported in literature thus it might be an alert suggesting further investigations to confirm or exclude the concomitance of neurological or genetic syndromes.

Declaration of Interest

The authors report no conflict of interest.

References

1. Jamani NA, Ardini YD, Harun NA. Neonatal tooth with Riga-Fide disease affecting breastfeeding: a case report. Int Breastfeed J. 2018;13:35; doi: 10.1186/s13006-018-0176-7.
2. Çavuş Ş, Özmen B. Riga-Fede disease in the upper jaw in an infant. Dermatol Ther. 2017;30(5):1-3; https://doi.org/10.1111/dth.12517.
3. Domingues-Cruz J, Herrera A, Fernandez-Crehuet P, Garcia-Bravo B, Camacho F. Riga-Fede disease associated with postanoxic encephalopathy and trisomy 21: a proposed classification. Pediatr Dermatol. 2007;24(6):663-5; https://doi.org/10.1111/j.1525-1470.2007.00564.x.
4. Lakshmanan S, Venkataraman S, Singh U. Riga-Fede disease: a mimicker of malignancy. BMJ Case Rep. 2021;14(3):e235976; doi: 10.1136/bcr-2020-235976.
5. Benitez B, Mülli J, Tzankov A, Kunz C. Traumatic ulcerative granuloma with stromal eosinophilia - clinical case report, literature review, and differential diagnosis. World J Surg Oncol. 2019;17:184; doi: 10.1186/s12957-019-1736-z.
6. Samuel SS, Ross BJ, Rebekah G, Koshy S. Natal and Neonatal Teeth: A Tertiary Care Experience. Contemp Clin Dent. 2018;9(2):218-22; doi: 10.4103/ccd.ccd_814_17.
7. Sharma B, Koshy G, Kapoor S. Traumatic Ulcerative Granuloma with Stromal Eosinophila: A Case Report and Review of Pathogenesis. J Clin Diagn Res. 2016;10(10):ZD07-ZD09; doi: 10.7860/JCDR/2016/22265.8657.
8. van der Meij EH, de Vries TW, Eggink HF, de Visscher JG. Traumatic lingual ulceration in a newborn: Riga-Fede disease. Ital J Pediatr. 2012;38:20; doi: 10.1186/1824-7288-38-20.
9. Senanayake MP, Karunaratne I Persistent lingual ulceration (Riga-Fede disease) in an infant with Down syndrome and natal teeth: a case report. .J Med Case Rep. 2014;8:283; doi: 10.1186/1752-1947-8-283.
10. Alobaida S, Lam JM.Bilateral harlequin syndrome, unilateral Horner syndrome, and Riga-Fede disease as presenting features of hereditary sensory and autonomic neuropathy type IV. Pediatr Dermatol. 2021;38(1):213-6; doi: 10.1111/pde.14385.
11. Polat Ekinci A, Kılıç S, Babuna Kobaner G. Early-onset and persistent traumatic granuloma of the tongue (Riga-Fede disease) associated with neonatal teeth and Down syndrome. J Eur Acad Dermatol Venereol. 2019;33(3):e131-e132; doi: 10.1111/jdv.15336.
12. Alamankany A. Riga-Fede disease associated with Fabry's disease and Niemann-Pick C disease in a boy with microcephaly: A case report. Int J Health Sci (Qassim). 2021;15(1):56-8.
13. Sivamurukan P, Chandrasekaran V, Biswal N. Riga Fede Disease with Glutaric Aciduria Type 1. Indian J Pediatr. 2020;87(6):484-5; doi:10.1007/s12098-019-03125-w.
14. Kariya PB, Shah S, Singh S, Buch A Riga-Fede Disease Associated with Syndactyly and Oligodactyly: A rare Occurrence. .J Clin Pediatr Dent. 2019;43(5):356-9; doi: 10.17796/1053-4625-43.5.10.
15. MontanariM, Callea M, Yavuz I, Maglione M. A new biological approach to guided bone and tissue regeneration. BMJ Case Rep. 2013 Apr 9;2013:bcr2012008240.