Riga-Fede Disease Associated with Syndactyly and Oligodactyly: A rare Occurrence

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Background: *Eruption of first primary tooth starts on an average at around six to seven months of age. Presence of teeth at birth or within a month after birth is considered rare. Natal teeth are those present in the oral cavity at the time of birth. These teeth can cause ulcers on the ventral surface of the tongue, lip, and the mother's breast characterizing the Riga Fede Disease. Exact aetiology of natal and neonatal teeth is still unclear. The treatment depends on its mobility of teeth as it is associated with the risk of aspiration or swallowing, whether the natal tooth is supernumerary or primary, causing any problems in breast feeding, presence of soft tissue injuries on tongue of the child or mother's breast and overall health of child.*

Case presentation: *A fifteen day old girl reported with large ulceration on ventral surface of tongue due to sharp natal teeth present at mandibular anterior region which was also associated with feeding difficulties. Along with Riga Fede disease Syndactyly and Oligodactyly in left and right legs respectively was also observed. Extraction of the teeth has been carried out and complete healing of ulceration has been achieved within 30 days.* ***Conclusion:*** *The present case report describes a rare occurrence of Riga Fede disease with Syndactyly and Oligodactyly and highlights its symptomatology and therapeutic approach.*

Keywords: *Natal teeth, Riga-fede disease, Syndactyly, Oligodactyly*

INTRODUCTION

Eruption of first primary tooth starts on an average at around six to seven months of age.[1] Presence of teeth at birth or within a month after birth is considered rare. These teeth are called natal teeth when they are present at birth and neonatal teeth when they erupted during the neonatal period (first 30 days of life). Several terms have been used in the literature to designate teeth that erupt before the normal time, such as congenital teeth, fetal teeth, predecidual teeth, and dentitia praecox.[2,3]

Exact etiology of natal and neonatal teeth is still unclear. Few of the hypotheses that explains the etiology are dominant autosomal inheritance, endocrine disturbance resulting from pituitary, thyroid, and gonads; excessive or increased resorption of overlying bone resulting in early teeth eruption; poor maternal health, endocrine disturbances, febrile episodes during pregnancy, and congenital syphilis.[4]

The presence of lower incisors at birth may lead to the possibility of swallowing and aspiration[4] and development of Riga-Fede Disease (RFD), which is also called traumatic ulcers that may be located on the ventral surface of the tongue, lip, and on mother's breast.[5] The lesion initiates as an ulcerated area that, with the repetition of trauma, can evolve into an enlarged fibrous mass with ulcerative granuloma appearance. This lesion makes it difficult for the infant to suck and feed, putting the baby at risk of nutritional deficiencies.[5]

It is important that professionals have a thorough knowledge of etiology, clinical features and presentation of RFD as natal teeth can also be associated with an underlying syndrome. Proper assessment, diagnosis, and treatment in such cases is always beneficial for overall development of the child. Failure to diagnose and treat properly may result in inadequate intake of nutrients and dehydration by the baby, increasing the potential for infection at the site.[6]

The present case report describes a rare occurrence of Riga Fede disease with Syndactyly and Oligodactyly and highlights its symptomatology and therapeutic approach.

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Case report

A fifteen days old girl reported with the complaint of difficulty during feeding due to presence of mandibular anterior teeth since birth. The infant's mother had noticed ulcerations present on ventral surface of tongue two days back (Figure 1). Margins of the natal teeth were very sharp and mobile due to which mother had difficulty in feeding and the child remained cranky all the time. No history was given of any systemic problems, fever, dehydration and nutritional deficiencies by mother. Prenatal, natal, post-natal and family history revealed no relevant findings. No developmental disorders or congenital syndromes were observed in immediate family of infant. Medical history was not significant. General physical examination revealed fused toes on left foot (Figure 2) and on right limb only 3 digits were present with cleft in between big toe and second toe (Figure 3) which was diagnosed as syndactyly and oligodactyly respectively.

Intraoral examination revealed two natal teeth in the mandibular anterior region which were fused. According to Hebling (1997)[7] these teeth can be classified as type 2 i.e. solid crown poorly fixed to the alveolus by gingival tissue and little or no root and. The fused natal teeth were grade 1 mobile and had sharp incisal edges that caused ulceration on the ventral surface of tongue. The lesion was roughly round in shape having 2cm diameter, erythematous and raised, with an indurated border covered by yellowish white slough. Bleeding was not associated with the lesion. (Figure 1) Based on these findings diagnosis of Riga-Fede disease was made. Extraction of fused natal teeth was planned as lesion was quite big and mother was facing difficulties in feeding the child. Pediatrician referral was done before extraction and vitamin K had been administered before extraction as it was not administered after birth. Topical anesthesia was applied and extraction was done. Teeth were completely fused in the crown and coronal ½ of the root length. (Figure 4) Bleeding was controlled by applying gauze pressure. Post extraction instructions were given. Patient was called for follow up after 24 hours.

Extracted fused natal teeth had been sent for histopathological examination which shows presence of eosinophillic structure along with tubules like structure suggestive of dentin like tissue shows columnar cells just beneath the dentinal layer suggestive of odontoblasts. Below the odontoblastic layer it comprises of spindle shaped fibroblasts along with many blood capillaries and few local areas showing odontogenic cell rests suggestive of primitive odontogenic tissue. Overall clinico-histopathological feature is suggestive of natal teeth (Figure 5). Follow up was done for the patient after 15 days and 30 days. Complete healing of the ulcer has been achieved in 30 days (Figure 6).

Figure 1: Ulceration on ventral surface of tongue along with natal teeth.

Figure 2: Syndactyly in left foot with fused third and fourth toe.

Figure 3: Oligodactyly in right foot with only three digits separated by cleft in between big toe and second toe.

Figure 4: Extracted fused natal teeth.

Figure 5: Clinico-histopatholoical feature suggestive of natal teeth (10X magnification)

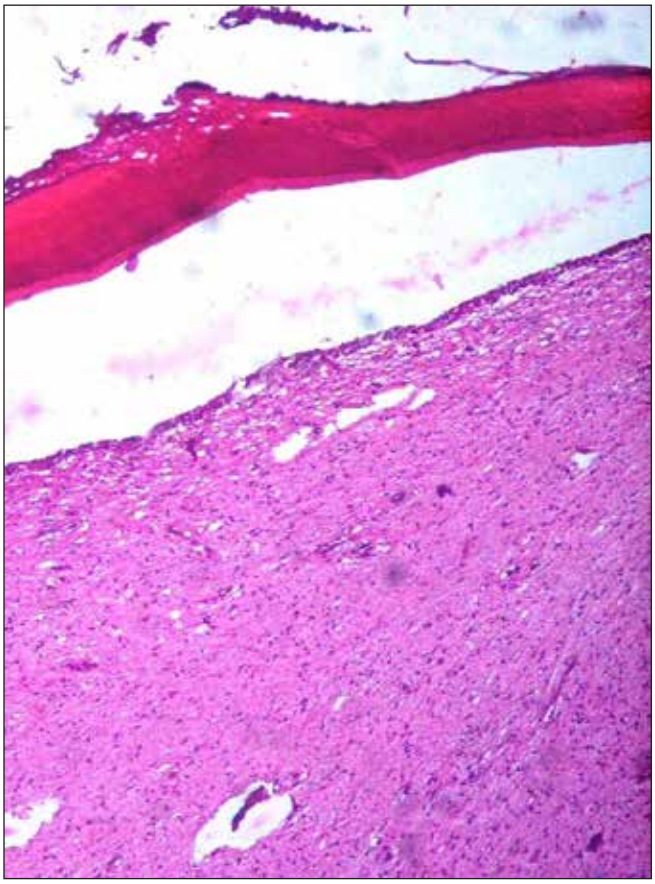

Figure 6: Post-operative photograph showing complete healing of lesion after 30 days.

DISCUSSION

The most impending complication of natal and neonatal teeth is an ulceration caused by rubbing the teeth against the ventral surface of the tongue. This condition is termed as Riga-Fede Disease, if the patient is younger than two years of age.[8] The incidence of Riga-Fede disease in the presence of natal or neonatal teeth has been reported to be 6% to 10% with female predilection.[9] The classic ulcer associated with Riga-Fede disease along with a skeletal abnormality can also indicate the existence of underlying developmental or motor disorders like Pallister Hall Syndrome, Short Rib Polydactyly Syndrome Type II, Odonto Tricho Ungual Digital Palmer Syndrome and many more.[2]

The presence of natal and neonatal teeth may have different line of treatment as per its clinical presentation. The maintenance of these teeth in the mouth is the first treatment option, unless this would cause injury to the baby.[10] The decisive factors in retaining such teeth are the implantation and degree of mobility, inconveniences during suckling, interference with breast feeding, possibility of traumatic injury, and whether the tooth is part of the normal dentition or is supernumerary.[11]

An early accurate diagnosis is mandatory to differentiate Riga-Fede disease from serious neurologic and hereditary disorders which also presents similar oral ulcerations.[12] The disease is associated with infection, dehydration and traumatic ulcer which interferes with proper suckling and feeding which can be a risk factor for nutritional deficiencies. So management of the disease is of prime concern for pedodontists. The treatment of lesion has varied over the years, from conservative to invasive management. The treatment options being addition of composite restoration, recontouring of the sharp incisal edges of natal teeth, extraction of offending teeth and modifying feeding behaviour with feeding appliances.[13]

The risk of dislocation and consequent aspiration, in addition to traumatic injury to the baby's tongue and/or to the maternal breast, have been described as reasons for removal.[14]

According to Allwritht (1958)[15] and Zhu and King (1995)[16], the Riga-Fede disease does not represent, by itself, an indication for extraction since an acute incisal margin can be relieved by smoothing.

If the treatment option is extraction, this procedure should not pose any difficulties since these teeth can be removed with a forceps or even with the fingers.[17] However, Bodenhoff J (1960)[17] emphasized to avoid extraction up to the 10th day of life to prevent hemorrhage, assessing the need to administer vitamin K before extraction, considering the general health condition of the baby, avoiding unnecessary injury to the gingiva, and being alert to the risk of aspiration during removal. This waiting period before performing tooth extraction is due to the need to wait for the commensal flora of the intestine to become established and to produce vitamin K, which is essential for the production of prothrombin in the liver.[18] Thus, it is safer to wait until a child is 10 days old before extracting the tooth. If it is not possible to wait then it is advisable to evaluate the need for administration of vitamin K with a pediatrician, if the newborn was not medicated with vitamin K immediately after birth. Vitamin K (0.5-1.0 mg) is administered intramuscularly to the baby as part of immediate medical care to prevent hemorrhagic disease of the newborn.[15]

Syndactyly, also known as webbed toes is characterized by fusion of two or more digits of the feet. Oligodactyly is the presence of fewer than five toes or fingers on a foot or hand. In our presented case Syndactyly is present on left foot and Oligodactyly is present on right foot. A case of Riga Fede disease along with polydactyly was referred earlier[19], but occurrence of Riga Fede with syndactyly in one foot and Oligodactyly in another is very rare and not reported in literature till date; hence such cases should be looked for further establishment of an association.

CONCLUSION

1. Natal/neonatal tooth with mobility is the only indication for extraction, and in firmly attached teeth other conservative management strategies should be planned.
2. Vitamin K starts forming in body from 10th day. So if extraction is planned before that vitamin K supplements must be given to the infant.
3. Association of Riga-Fede disease and developmental, neurological or motor diseases has been found so one should always rule out for any such condition.

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