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Neonatal sublingual traumatic ulceration – case report & review of the literature

REVIEW ARTICLE

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Abstract – Traumatic ulceration of the ventral surface of the tongue is a rare condition observed in neonates with natal or neonatal teeth. These lesions are initiated and exacerbated by the continuous contact between the tongue and natal/neonatal teeth during physiologic activities such as suckling and swallowing, which in turn may interfere with proper feeding, putting the neonate at risk for nutritional deficiencies. Sublingual traumatic ulcers may also be the presenting sign of an underlying developmental or neurologic disorder. This case report details the management of a sublingual traumatic ulcer in a 20-day-old infant, along with a review of the treatment modalities available for management of this condition, especially in neonates.

Neonatal sublingual ulceration was of great concern in Europe, especially Italy, during 19th century, as it was frequently observed in malnourished infants and was considered a cause of death in these children (1). Caldarelli (1857) first described this condition in a cachectic infant, which was later referred to by several Italian authors as *afta cachettica* ('afta' meaning ulcer). This condition was associated with general wasting in the infant patient, which eventually led to death (*'exitus lethalis'*) (1). Antonio Riga, in 1881, described the '*localized membrane formation'* and '*hard tumor'*, associated with this condition that drew the attention of the medical community (1). Francesco Fede, in 1890, published the first histologic studies and reported 20 cases of this oral condition (2).

Ever since these initial reports, various terms have been used to denote these lesions and there appears to be no consensus over the nomenclature. Conventionally, the term Riga-Fede disease (RFD) was used to denote ulcerative lingual lesions that appear in young infants shortly after the eruption of lower incisors (2). In infants, it has also been called Riga's disease, sublingual ulcer, sublingual granuloma, and reparative lesion of the tongue. However, similar lesions were also observed in adults and older children, and were denoted as ulcerated granuloma eosinophilicum diutinum, traumatic granuloma of tongue, eosinophilic granuloma of tongue, eosinophilic ulcer of tongue, traumatic eosinophilic granuloma, and ulcerative eosinophilic granuloma of the tongue. Elzay (3) suggested that the term 'traumatic ulcerative granuloma with stromal eosinophilia' (TUGSE) be used to describe the sublingual ulcerative traumatic lesions observed in both children and adults, due to the increased eosinophilic infiltration observed in the connective tissue beneath these lesions. Even though the term 'granuloma' may be misguiding considering the fact that the pathogenesis of TUGSE do not involve a granulomatous inflammation, it has found wide usage in the names used to refer to these lesions. Goho (4) used the term 'neonatal sublingual traumatic ulceration' (NSTU) to specifically denote the sublingual lesions that occur in neonates due to traumatic raking of tongue over natal/neonatal teeth and this term is preferred as the traumatic lesions in neonates must be considered as a separate entity, due to the special considerations that need to be observed before management of these lesions in neonatal patients. Domingues-Cruz et al. classified riga-fede disease and used the term 'precocious riga-fede disease' to denote those lingual lesions associated with natal or neonatal teeth in the first 6 months of life (5). The rarity of these ulcers can be ascertained from the fact that these lesions have been observed only in 6–10% of cases of natal/neonatal teeth (6).

Early efforts to link sublingual traumatic ulcers to specific infections such as pertussis or cachexia were unsuccessful. Moncrieff (7) reviewed the suggested etiologies of RFD as early as 1933 and concluded that the *'inflammatory granuloma'* associated with this condition

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was a result of mechanical trauma and secondary non-specific infection. The effect of trauma was verified by Bhaskar and Lilly on tongues of rats that were subjected to chronic trauma. They found that the lesions so produced were histologically similar to those encountered in humans (8). However, these conclusions were refuted by many workers (9, 10). Tang et al. (10) suggested that trauma may only be a contributing factor, which exposes the sub-epithelial tissues and capillary endothelial cells to viral and toxic agents that initiate an inflammatory response leading to tissue damage. However, electron microscopic investigations of these sublingual lesions by the same workers failed to reveal viral particles or cellular changes suggestive of viral infection. Later, immunohistochemical studies of these ulcers by Regezi et al. (11) and El-Mofty et al. (12) suggested a T-cell mediated immune response to be responsible for the tissue destruction observed in these conditions. In summary, the etiology of sublingual traumatic ulcers is yet to be deciphered, although chronic trauma appears to be a significant contributory factor.

This report details the management of a NSTU along with a review of the treatment modalities available for management of this condition, especially in neonates.

Case report

A 20-day-old male baby of Indian origin was referred to the Department of Pediatric Dentistry, CSM Medical University, India by his pediatrician for management of a rapidly progressing sublingual ulcer. The parents first observed a tooth emerging from the lower jaw in their child's mouth, when the child was just 1 week old. Two weeks later, a large whitish lesion was observed on the undersurface of the tongue. Moreover, the parents also observed that the child experienced discomfort during suckling, and did not nurse. The attending pediatrician attributed the infant's discomfort to the sublingual ulcer and expressed significant concerns over the potential for secondary infection of the ulcerated area and associated bacteremia.

On intraoral examination, an ulcer measuring approximately 20 mm × 12 mm was observed on the ventral surface of the tongue (Fig. 1). The edges of the ulcer were raised and everted, and mild induration of the margins was palpable. The floor of the ulcer was covered by a white, necrotic membrane, which could be separated from the base using a betadine soaked sterile cotton swab. Dental examination revealed the presence of a neonatal tooth projecting 2 mm from the mandibular gum pad in the left central incisor position. No mobility was detected in the erupted tooth. The medical history of the patient was reviewed with the parents, and questions pertaining to specific symptoms (Table 1) were raised to exclude the presence of any underlying developmental disorder associated with sublingual traumatic ulcers, and the history was non-contributory. Radiographic examination of the offending tooth revealed it to be a primary central incisor. The weight and head circumference of the baby were within normal limits for his age.

Considering the age of the child, extent of the lesion, difficulty in nursing, and the pediatrician's concern pertaining to the chances of secondary infection, the parents were explained that extraction of the offending neonatal tooth would be the most logical solution. However, the parents were very apprehensive and never provided consent for extraction. They insisted that a 'less traumatic' approach be adopted.

Due to lack of parental consent, it was decided to manage the lesion conservatively. The incisal edge of the neonatal tooth was smoothened to eliminate the sharp traumatizing edges (Fig. 2). The parents were advised to apply oral triamcinolone acetonide in orabase (0.1% triamcinolone acetonide, Kenacort Oral Paste; Sarabhai Piramal Pharmaceuticals Ltd, Vadodara, India) over the ulcerated region using a dry cotton swab. The infant's pediatrician had advised the parents to supplement the patient's diet with infant formulas to compensate for the poor acceptance of breast-fed milk due to the sublingual ulcer. During the follow-up appointment after 1 week, the lesion was found to heal well (Fig. 3).

However, 2 weeks later, the parents once again reported with similar complaints of feeding difficulty. Moreover, review of the child's medical report revealed that the child had also failed to gain weight. On examination, it was found that the neonatal tooth had re-erupted and was traumatizing the healing ulcer (Fig. 4). To avoid repetitive trauma, it was decided to extract the neonatal tooth. The parents were once again advised that extraction would be the logical solution considering the intraoral and systemic status of their ward. The risk of repetitive trauma to the non-healing ulcer and its persistence far outweighed the benefits of retaining the tooth.

The neonatal tooth was extracted under general anaesthesia followed by gentle curettage of the socket

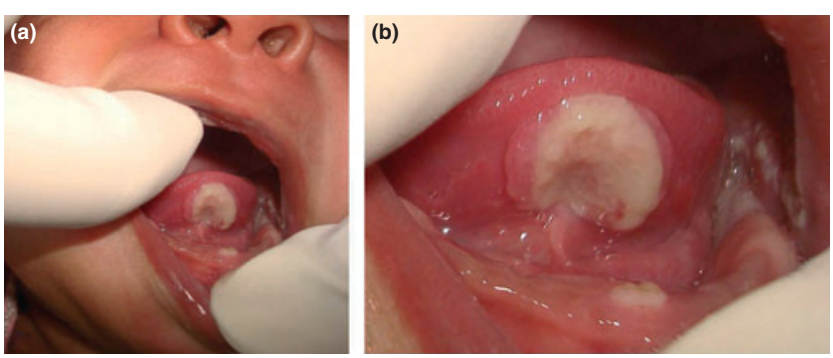

Fig. 1. Intraoral view showing the neonatal tooth and large sublingual ulcer with a white necrotic slough covering the base.

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Table 1. Developmental disorders associated with neonatal sublingual traumatic ulcer

Syndromes	Clinical features detected during neonatal period
Congenital insensitivity to pain with anhidrosis	Episodes of hyperpyrexia due to lack of perspiration. Nerve biopsy reveals total absence of unmyelinated nerve fibers that convey impulses of pain, temperature and autonomic functions, lack of an axon flare following intradermal histamine
Familial dysautonomia	Poor sucking and swallowing, choking inspirations, pneumonia, red blotching of skin, diminished/no fungiform papillae of the tongue, lack of an axon flare following intradermal histamine. Absence of overflow tears with emotional crying can be detected only after 7 months of age

Fig. 2. Incisal edges of the neonatal tooth smoothened to reduce the traumatic impact over the ulcer.

Fig. 3. Regression of sublingual lesion evidenced after 1 week following incisal grinding.

Fig. 4. Re-erupted neonatal tooth traumatizing the healing ulcer.

so that no remnant of dental papilla or hertwigs epithelial root sheath is left behind (13). The infant was re-examined by the pediatrician to exclude the presence of any neurologic or systemic disorder associated with sublingual neonatal ulcer. However, no sign of developmental or neurologic disorders was observed. One month later, the lesion had resolved completely (Fig. 5), and the child could be fed comfortably.

Discussion

Sublingual traumatic ulcers are observed both in children and adults. The infantile swallow, an essential function in the neonate, is closely associated with suckling, and both are well developed by about the 32nd week of intrauterine life and begin to cease only with the eruption of the primary incisor teeth. Children achieve most features of the mature swallow at 12–15 months of age (14). This compulsive extension of the tongue beyond the gum pads associated with suckling and infantile swallowing places the neonate at a high risk of encountering a traumatic sublingual ulcer in the presence of a natal or neonatal tooth. Kinirons (15) reported a peculiar case wherein a sublingual ulcer was noticed in a neonate immediately after birth in relation to a natal tooth, suggestive of the role of infantile swallowing and suckling in initiation and progression of the NSTU.

Table 2 lists the treatment alternatives that have been adopted for the management of NSTU by various authors (available in English literature). Slayton (16) provided an excellent summary of the treatment alternatives in the management of riga-fede disease. She reported a case of sublingual traumatic ulcer in a 10-month-old child, which was managed conservatively by reshaping the incisal edges, and the feeding pattern of the child was also altered, which involved use of a sippy cup with relatively larger holes so as to avoid vigorous sucking. However, such alterations in the feeding pattern cannot be advised in neonates. In the present case, the pediatrician had advised the parents to provide infant formulas through a spoon, to supplement the infant's nutrition, which was lacking due to difficulty in breast feeding.

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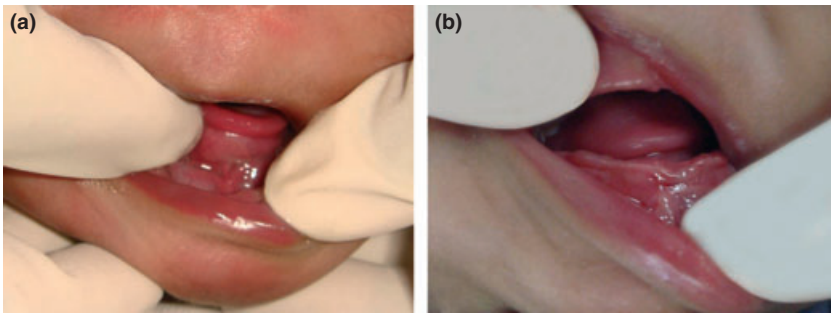

Fig. 5. Healing of sublingual lesion and extraction socket evidenced after 35 days.

Table 2. Sublingual ulcers associated with natal/neonatal teeth and their management

Author	Age	Offending tooth	Treatment	Outcome
Jacobs MH 1956 (18) (part of review)	1 week	Natal mandibular incisor	Extraction	Healing with loss of tip of tongue
Kinirons MJ 1985 (15)	Immediately after birth	Natal tooth in mandibular incisor region	Extraction	'Rapid healing'
Goho C 1996 (two patients) (4)	21 days	Natal tooth in mandibular incisor region	Extraction	Lesion resolved fully in 5 days
	10 days	Two natal mandibular central incisors	Composite resin coverage of the incisal edges	Sublingual abrasions had resolved in 1 week
Buchanan and Jenkins 1997 (21)	4 weeks	Two natal mandibular central incisors.	Stomahesive wafer covering of incisal edges.	Complete healing in 21 days. Fibrous scar was observed 4 months later
Uzamis et al. 1999 (33)	2 months	Two neonatal teeth in mandiblar incisor region	Extraction	Healing of ulcerated area observed within 1 week
Kaur et al. 2003 (34)	4 months	Two neonatal mandibular incisors	Grinding of sharp incisal edges and topical application of 'dologel'	1st week – signs of regression. 2nd week – regression of lesion and hyperplastic area was observed
Hegde RJ 2005 (35)	28 days	Two neonatal teeth in mandibular incisor region	Extraction	Lesion resolved fully in 10 days
Choi et al. 2009 (20)	2 months	Natal tooth in mandibular incisor region	Grinding of incisal edges and topical corticosteroids in orabase	Complete healing in 1 month

If the ulcerated area is large and denuded, even a smoothened incisal edge may still come in contact with and traumatize the tongue during suckling and delay healing (4). Moreover delayed healing of traumatic tongue ulcers has been reported to result in cleft deformities of tongue on healing (17, 18). Goho (4) reported two cases of neonatal sublingual traumatic ulcer, which were associated with neonatal teeth. Extraction was the preferred treatment in an infant who had a large ulcer and was refusing to nurse, resulting in lack of weight gain, whereas a conservative treatment alternative (in the form of composite resin coverage of incisal edges) was adopted in a healthy infant with minimal ulceration of the tongue. In the present case, presence of a large ulcer coupled with failure to gain weight, difficulty in breast feeding, and the increased risk of septicemia favored extraction as the treatment of choice. In infants less than 10 days of age, who are candidates for extraction of natal/neonatal teeth, it is advisable to consider the need for vitamin K supplementation (0.5–1.0 mg) with the pediatrician if vitamin K was not administered immediately after birth.

According to Zhu and King (19), RFD by itself is not an indication for extraction. Conservative treatment options can be adopted in case of smaller ulcers that are associated with firm or less mobile neonatal/natal teeth (Grade I mobility). Reshaping or composite resin coverage of the sharp incisal edges of natal/neonatal teeth (20) is the common conservative treatment option recommended for management of neonatal sublingual ulcer. Often, only the sharp incisal edges of the natal/neonatal teeth would have erupted providing very less tooth structure for composite resin coverage, which are better managed by smoothening the sharp edges. Buchanan and Jenkins (21) reported a case of neonatal sublingual ulcer that was managed conservatively by covering the incisal edges of the neonatal teeth by stomahesive wafers. However, the parent and patient compliance for such a procedure that involves placing a foreign object within the oral cavity throughout the day (until the infantile swallowing pattern ceases) is questionable.

More often, the systemic status of the patient should be given due consideration before exercising conservative treatment strategies. Neonates presenting with associated signs or symptoms of dehydration (18) or complaints of lack of weight gain are better candidates for extraction, due to the definitive and rapid healing that can be expected in these cases. Moreover, persistent dehydration or malnutrition during the neonatal period may

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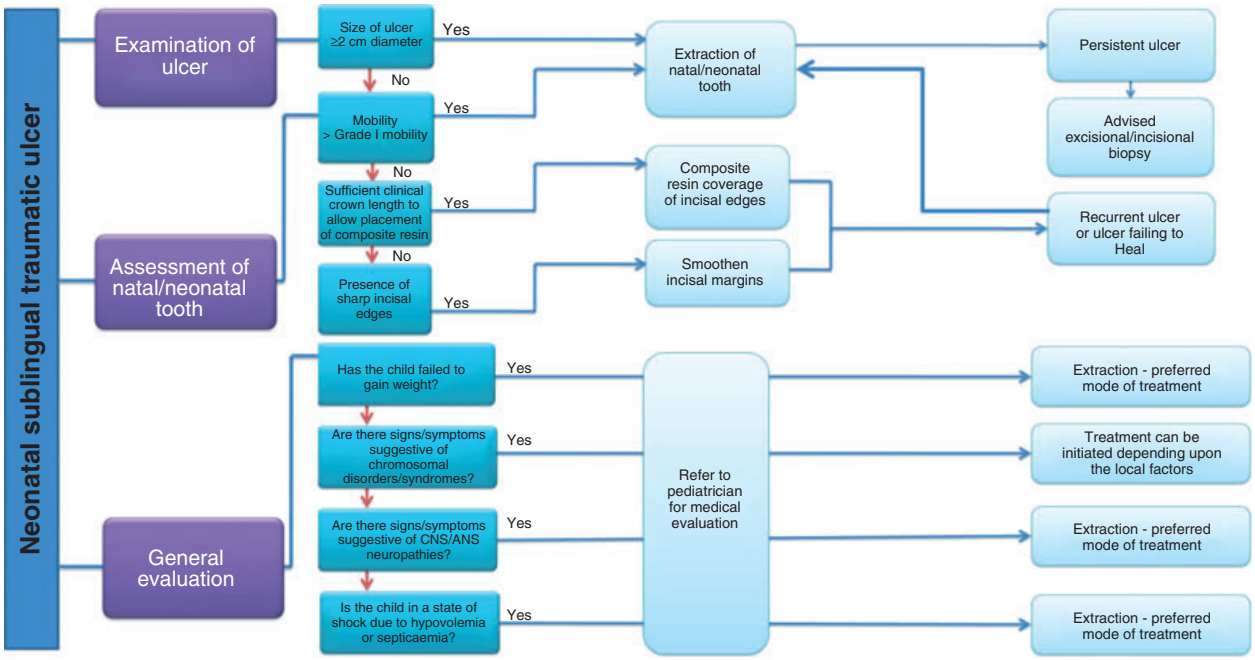

Fig. 6. Schematic illustration depicting the factors, which need to be considered while managing a neonatal sublingual traumatic ulcer.

negatively influence the physical and mental growth of the child.

Sublingual traumatic ulcers are often oral manifestations of an underlying neurologic or developmental disorder such as congenital insensitivity to pain with anhidrosis (CIPA) [hereditary sensory and autonomic neuropathy(HSAN) – type IV] (22, 23) and familial dysautonomia (FD) or Riley-Day syndrome [HSAN type III] (24, 25). CIPA presents more frequently in boys than girls, and infants with this condition present commonly with episodes of high fever related to warm environmental temperatures as they do not perspire (23, 26). FD is expressed in neonatal life by poor sucking and swallowing, and feeding difficulties remain a major symptom throughout childhood (25, 26). Even though such conditions have been reported only in older infants with RFD in association with primary incisors, their presence should also be evaluated in neonates (27) with sublingual traumatic ulcers. Correlation of clinical signs and symptoms with appropriate genetic tests can enable early diagnosis and intervention in both these conditions (Table 1). Even though cases of conservatively managed traumatic tongue lesions have been reported in individuals with these neuropathies (28), extraction of the offending tooth appears to be the better option with good long-term prognosis (23). Disorders characterized by self-mutilation, such as Lesch-Nyhan disease (LND), also can result in similar oral manifestations (29). However, the onset of self-injury may not occur during the neonatal period in LND and symptoms such as hypotonia, recurrent vomiting and difficulty with secretions may only be evidenced around the age of 4 months (26). Even though numerous reports have suggested the use of biopsy prior to management of RFD (25, 30), such procedures are rarely required and need to be reserved only for lesions that fail to heal even after extractions. Figure 6 provides a schematic illustration of the factors that need to be considered while managing a neonate with sublingual traumatic ulcer.

The diet of neonates who have difficulty in accepting breast-fed milk due to the sublingual traumatic ulcer must be adequately supplemented with infant formulas. Use of fluoridated water for reconstituting powdered formulas should be avoided (31). Ready-to-use formulas are preferred in these infants due to their low fluoride content. However, restoration of breast feeding should be the primary objective while treating a neonate with sublingual traumatic ulcers and the parents need to be motivated for the same.

Parent counseling is an indispensable component for successful management of neonatal sublingual ulcer. The pediatric dentist should be empathetic while addressing the parental concerns keeping in mind the distress that the parents may be experiencing due to their infant's condition. Informed consent must be obtained after the parents are properly informed about all treatment aspects involved in this situation (32). The importance of periodic follow-up and home dental hygiene also needs to be emphasized.

In those patients who were managed by means of extraction, long-term follow up may be required to evaluate the patients for space-related problems and abnormalities in the development of permanent successors in the future, even though such incidences are rare following extraction of natal/neonatal teeth (6).

Conclusion

Successful management of neonatal sublingual traumatic ulcers involves obtaining a detailed history, proper

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general and local examination, effective parent counseling, and prompt treatment. Failure to diagnose or treat these lesions promptly may result in mutilation or permanent deformities of the tongue, or poor nutritional intake resulting in dehydration and growth retardation.

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