Diagnosis and Management of Natal Tooth Secondary to Beckwith–Wiedemann Syndrome in a 25-day-old Infant: A Rare Case Report

Abstract

Beckwith–Wiedemann syndrome (BWS) is a rare genetic disorder characterized by somatic overgrowth and an increased predisposition to various medical conditions, including neoplasms. This case report presents a 25-day-old male infant with BWS exhibiting macroglossia, hepatosplenomegaly, hyperinsulinemic hypoglycemia, gum hypertrophy, and cystic lesions in the maxillary arch, prompting a multidisciplinary collaboration of a pediatrician, pedodontist, oral surgeon, and an oral pathologist. The patient also presented with Hebling's Class III natal tooth. Following all the precautions, the natal tooth was extracted under local anesthesia. This case report stands out for documenting the presence of a natal tooth in a diagnosed case of BWS for the first time, contributing to the understanding of oral manifestations in rare genetic syndromes and underscoring the need for timely intervention and comprehensive medical care in managing this complex genetic syndrome.

Keywords: *Beckwith–Wiedemann syndrome, genetic disorder, macroglossia, natal tooth*

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Introduction

Beckwith–Wiedemann syndrome (BWS) is a rare genetic disorder characterized by lateralized overgrowth, including macrosomia at birth and throughout childhood, macroglossia, abdominal wall defects such as omphalocele, organomegaly, hemihypertrophy, or hemihyperplasia, and ear creases/ear pits in the posterior region. Individuals with BWS have an increased risk of developing childhood tumors, such as Wilms tumor, hepatoblastoma, and neuroblastoma.[1,2] The estimated prevalence of BWS is one affected child in 10,340 live births.[3]

The condition is associated with genetic and epigenetic abnormalities on chromosome 11p15 and can occur sporadically or be inherited in an autosomal dominant manner.[4] To provide comprehensive care, a multidisciplinary approach is employed, comprising pediatricians, geneticists, surgeons, and other specialists. Management involves monitoring and addressing specific symptoms, regular medical checkups, and screenings for tumor detection. Genetic counseling may be recommended for families with a history of BWS to understand the risk of inheritance. Early detection and intervention are crucial for optimal management of the syndrome. This case report explains the diagnosis and management of a 25-day-old male infant with natal teeth.

Case Report

A 25-day-old male infant was referred to the department of pediatric and preventive dentistry for an opinion regarding the dental status of the baby.

An examination of the patient's medical records revealed that he had been diagnosed with BWS. The patient's mother gave a history of difficulty during breastfeeding and refusal of feeding from paladai. The medical status of the patient showed syndromic features such as macrosomia, macroorchidism, ear creases, hepatosplenomegaly, hyperinsulinemic hypoglycemia, and meningitis. The baby measured 50 cm in length and weighed 4.15 kg at birth with a head circumference of 35 cm [Figure 1]. The baby was delivered through cesarean given perinatal asphyxia with meconium aspiration syndrome. The antenatal and natal history revealed polyhydramnios, and the postnatal

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history revealed respiratory distress, multiple episodes of hypoglycemia, and sepsis.

On intraoral examination, the patient had macroglossia [Figure 2], gum hypertrophy, natal teeth in the mandibular left anterior region [Figure 3], and cystic changes seen in the maxillary and mandibular arches anterior and posterior region [Figure 4]. A deep palatal vault with petechiae and adhesions on the buccal surface was also observed [Figure 5]. Ulcerations were noted in the anterior region of the maxillary left side. Extraorally, there was noticeable facial hypertrophy and enlarged incompetent lips. A broad and low nasal bridge was noted [Figure 6].

Figure 1: Syndromic feature of macrosomia

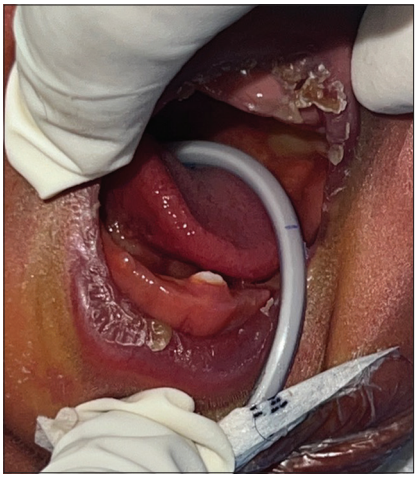

Figure 3: Natal teeth in the mandibular left anterior region

In addition, the patient's physique appeared larger than expected for his age, coupled with a comparatively shorter neck and a diagnosis of natal tooth associated with BWS was established.

Treatment

Addressing the chief complaint of the patient's parent, and through telecommunication with the oral and maxillofacial surgery and oral pathology departments, it was decided to extract the natal tooth under local anesthesia. The patient's medical fitness certificate was obtained from the treating doctor. Vitamin K injection (0.5 mg) was given to the patient intramuscularly the previous night of the proposed treatment.[5] The patient was already on broad-spectrum antibiotic coverage. The tooth was engaged with anterior extraction forceps and was pulled out using buccal and rotational force. The socket was very carefully curetted using a spoon excavator to remove any HERS or pulpal tissue left behind. The socket was then compressed, and hemostasis was achieved using gentle pressure [Figure 7]. The tooth was stored in 10% formalin and sent for further investigations.

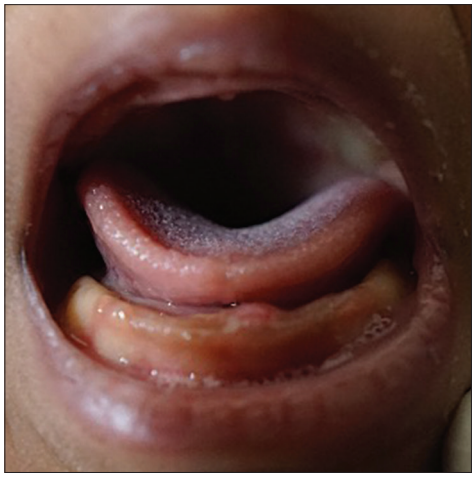

Figure 2: Macroglossia

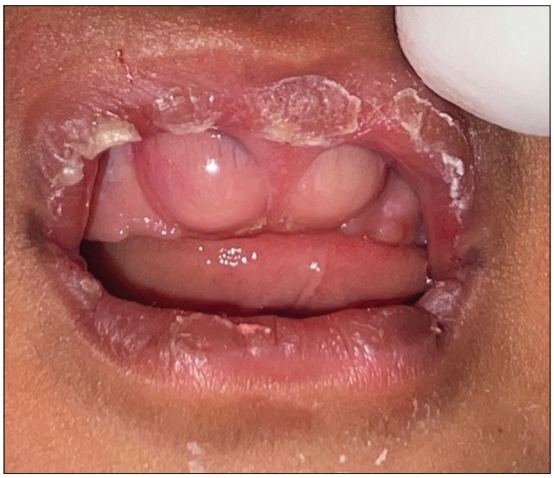

Figure 4: Cystic lesions of the maxillary arch in the anterior and posterior region

The gross examination of the tooth revealed a Class III natal tooth,[6] secondary to BWS.

Follow-up done after 2 months showed uneventful healing of the extraction site, and the mother reported no challenges with breastfeeding.

Findings of the ground section

The histological features included hypoplastic enamel of varying thickness, and the enamel structures were not evident. The coronal dentin was predominantly tubular, the DEJ was flat, and the radicular dentin was partly present. A wide pulp space was also noted [Figure 8a and b]. The ground section of the tooth was also viewed under polarized microscope [Figure 9].

Discussion

BWS is a rare genetic disorder characterized by overgrowth, certain physical abnormalities, and an increased risk of certain childhood cancers. It was first described by Dr. J. Bruce Beckwith and Dr. Hans-Rudolf Wiedemann in the 1960s. Individuals with BWS often exhibit excessive growth before and after birth, with manifestations including macroglossia, abdominal wall defects such as omphalocele or umbilical hernia, ear creases or pits, organomegaly, and hemihyperplasia. Neonatal hypoglycemia is common due to excess insulin production. BWS is typically caused by alterations in genes on chromosome 11, with most cases occurring sporadically. Early diagnosis and appropriate interventions are crucial for improving outcomes and quality of life in BWS patients.

Several factors, such as tooth prognosis, the potential for aspiration, challenges in breastfeeding, the risk of hemorrhage, and ulcerations, influence the management of natal/neonatal teeth. Most natal and neonatal teeth predominantly belong to the primary dentition and are not supernumerary. Typically situated in the lower incisor region, they are present in pairs in 61% of cases and align with the normal primary dentition in 95% of cases, with only 5% classified as supernumerary.[7,8] As per Bodenhoff and Gorlin's classification, 85% of affected teeth are lower incisors, 11% are upper incisors, 3% involve lower canines and molars, and merely 1% pertain to upper canines and molars.[7] These considerations should be taken into account when planning treatment. Histological examinations generally reveal hypoplastic enamel covering the majority of natal and neonatal tooth crowns, showcasing varying degrees of severity. Characteristics such as the absence of root formation, wide extensive and vascularized pulp, irregular dentin formation, and the absence of cementum formation are commonly observed.[9] Howkins' (1932)

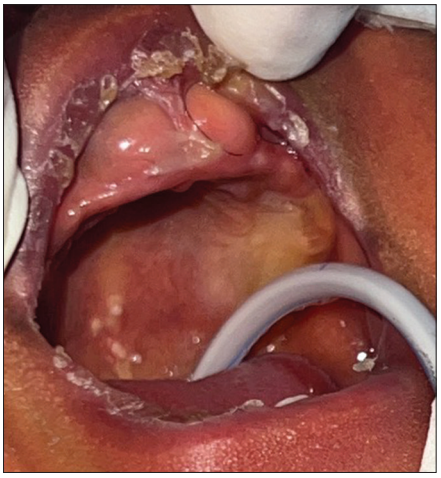

Figure 5: A deep palatal vault with petechiae and adhesions on the buccal surface

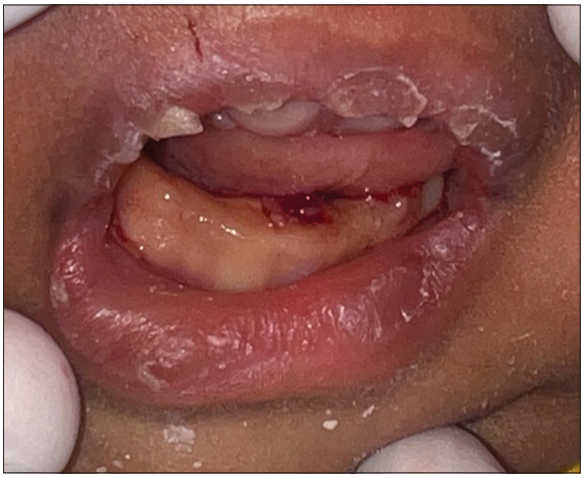

Figure 7: Postextraction hemostasis achieved

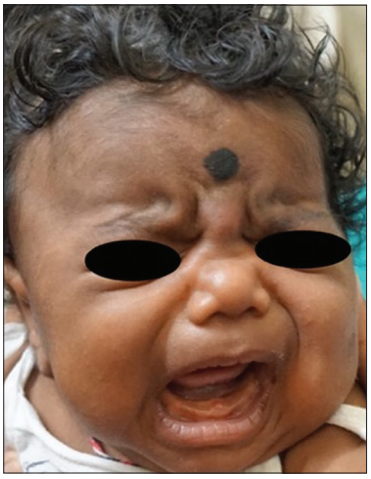

Figure 6: Broad and low nasal bridge

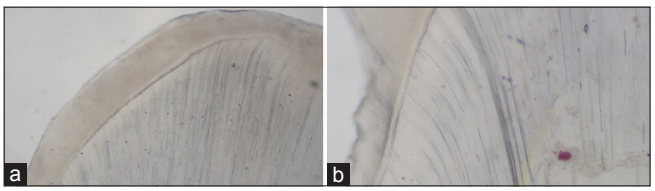

Figure 8: (a and b) Ground section of the natal tooth

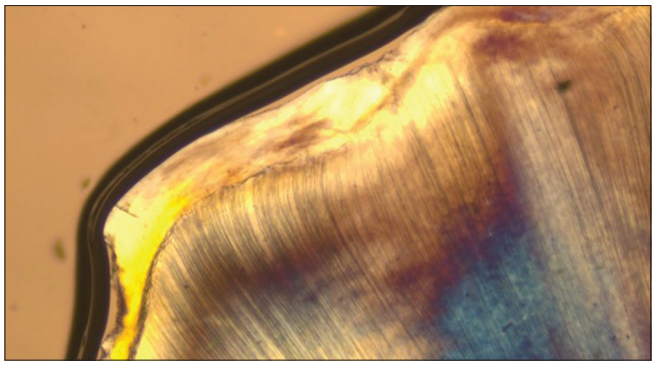

Figure 9: Natal tooth under polarized microscope

microscopic analysis of natal tooth sections indicates generally normal dentin, except for irregular spaces near the amelodentinal union and an expanded pulp chamber. Microscopic findings include irregular interglobular areas resembling osteodentin, atypical dentinal tubule arrangement, and a gradual decrease in the number of dentinal tubules from the crown to the cervical region.[10] These identified features align with observations in the current patient.

The decision to extract natal teeth is influenced by potential risks such as dislocation, aspiration, and traumatic injury to the infant's tongue or maternal breast.[11,12] In this case, the baby's suckling difficulties, refusal of feed from a paladai, ulcerations on the corresponding maxillary arch, and macroglossia were pivotal factors leading to the decision for natal tooth extraction. Numerous researchers have highlighted the potential risk of aspiration associated with natal teeth. One such instance of spontaneous tooth exfoliation has been reported by Bigeard *et al*.[13] based on the account provided by the parents of a 28-day-old infant, reporting the sudden disappearance of a natal tooth leading to suspicion that the tooth might have been swallowed, suggesting the possibility of aspiration.

In a unique case described by Kinirons,[14] a baby was born with a natal tooth, and there was the concurrent observation of a sublingual ulcer immediately after birth. According to the author, this ulcer likely stemmed from suction forces experienced during intrauterine life.

Rahul *et al*.,[15] discussed six cases of management of natal tooth in their report, which showed mobility was a common factor, posing a subsequent risk of aspiration. Consequently, extraction emerged as the sole treatment option for these cases. However, in one instance, a conservative approach was employed, managing the situation by grinding the incisal edges of the implicated tooth. If the chosen treatment involves extraction, the procedure is anticipated to be uncomplicated, as these teeth can be readily extracted using forceps or even manually with the fingers, posing minimal difficulty.[9] However, precautions must be taken, including avoiding extraction within the first 10 days of life to prevent hemorrhage, assessing the need for Vitamin K administration before extraction, considering the overall health of the infant, preventing unnecessary gingival injury, and remaining vigilant against the risk of aspiration during the removal process.[15-17] There have been reports of an association between premature teeth in newborns and specific syndromes such as Down's syndrome, Riga–Fede disease, Ellis–Van Creveld syndrome, Hallermann–Streiff syndrome, Rubinstein–Taybi syndrome, Pierre Robin syndrome, cleft lip and palate, ectodermal dysplasia, and craniofacial dysostosis.[18]

Conclusion

BWS is a rare genetic disorder characterized by somatic overgrowth and an increased predisposition to various medical conditions. Individuals affected typically manifest gigantism, visceromegaly, abdominal wall defects, and heightened susceptibility to specific neoplasms, particularly Wilms tumor and hepatoblastoma. The etiology of the syndrome is often sporadic, attributed to genetic and epigenetic anomalies, although familial inheritance is also documented. Clinical management necessitates regular medical surveillance, targeted intervention for specific health issues, and vigilant monitoring for neoplastic developments. Genetic counseling assumes a critical role in elucidating the risk of recurrence in subsequent pregnancies for affected families. Timely and comprehensive intervention, guided by a multidisciplinary approach, contributes to favorable prognoses for individuals with BWS. According to a literature search and my current knowledge, this case report is pioneering in documenting the occurrence of a natal tooth in a diagnosed case of BWS.

Declaration of patient consent

This is to certify that we have obtained all appropriate patient consent forms. In the form, the patient's parents have given their consent for the child's images and other clinical information to be reported in the journal. The patient's parents understand that their and the child's names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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