Hereditary sensory autonomic neuropathy presenting as recurrent traumatic injury to the tongue

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ABSTRACT

Hereditary sensory autonomic neuropathy (HSAN) is a group of rare genetic disorders in which affected patients have a diminished capacity to feel pain. This case describes a five-month-old female who initially presented with extensive trauma to her tongue. The repetitive injury to the tongue caused it to heal with a bifid deformity. This case highlights the importance of recognition of extensive oral trauma as one of the early signs of HSAN that should provoke a timely referral for neurological assessment.

INTRODUCTION

HSAN Type 4 is characterized by the inability to feel pain or thermal stimuli as well as anhidrosis and intellectual disability. Patients with HSAN are susceptible to a variety of physical injuries that can lead to permanent disability and premature death. The clinical manifestations of HSAN generally appear in infancy and include self-mutilating injuries to the oral cavity, eyes, and hands. As a result of complications related to injuries and fractures, patients with more severe presentations of HSAN rarely survive to adulthood.

CASE DESCRIPTION

A 5-month female patient presented with a chief complaint of bleeding from the tongue. Upon clinical exam, the mandibular primary central incisors were partially erupted in the oral cavity. The ventral surface of the tongue had a large ulceration with significant tissue loss at the midline (Fig 1).

Four weeks after the initial emergency room visit, the parents consented to the extraction of the mandibular primary central incisors (Fig 2). The patient is currently being monitored and re-evaluated at an older age to correct the clefting defect (Fig 3)

CASE REPORT

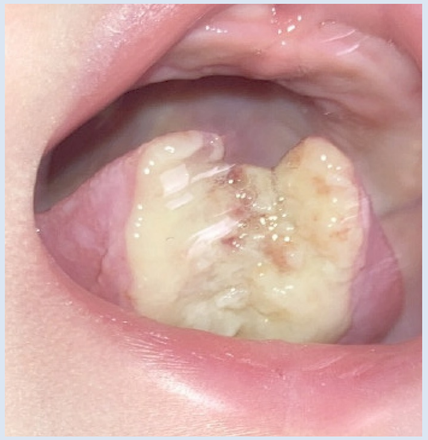

Figure 1: Initial presentation of the midline tongue ulceration following eruption of lower primary incisors

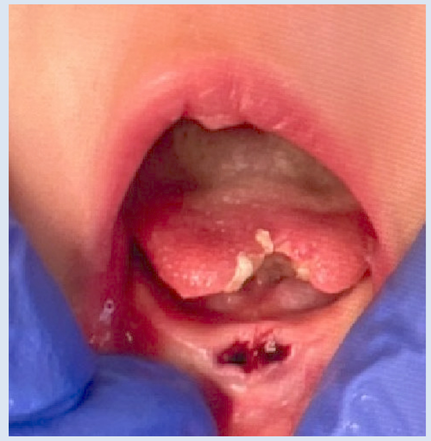

Figure 2: Immediately after extraction of 7.1 and 8.1

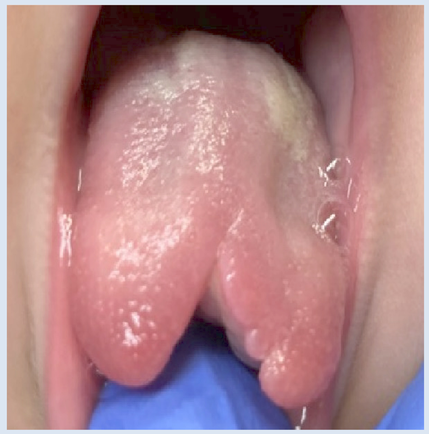

Figure 3: Intraoral photo of the tongue completely healed with a midline clefting defect

DISCUSSION

The strength of this current case report lies in the relatively early diagnosis of this rare syndrome. The diagnosis of HSAN at ten months of age prompted routine follow up visits with dentistry, ophthalmology, and occupational therapy for counselling and injury surveillance, with the hope of preventing long term complications of the disease. It is plausible that earlier extraction of the mandibular primary central incisors may have mitigated the damage to the tongue. There is no single consensus on the dental management best practices for patients with HSAN. The limited pool of patients with HSAN creates an obstacle for clinicians to offer a consensus on the most suitable treatment options.

CONCLUSION

Oral trauma is an early presenting sign of HSAN. Pediatric dentists may be one of the first healthcare providers to assess the condition. Early diagnosis of HSAN may potentially allow for interventions that mitigate more extensive injuries.

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