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Case Report

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Oral Management of Hereditary Sensory and Autonomic Neuropathy Type V: A Rare Case Report

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Abstract

Hereditary sensory and autonomic neuropathies (HSAN) are rare diseases. Five different types are described. Hereditary sensory and autonomic neuropathy type V, also known as congenital insensitivity to pain, is a rare autosomal recessive disease seen in early childhood. Self-mutilation is an invariable feature of this disorder involving the teeth, tongue, lips, and fingers.

This report described the case of a 2-year-old baby boy who had self-mutilating injuries to her tongue and hands, caused by biting. Protective splints were given to the patient to prevent further self-mutilation.

Keywords: Autonomic Neuropathy, Hereditary Sensory, Insensitivity to pain, Oral Management, Self-mutilation

Introduction

Hereditary sensory and autonomic neuropathies (HSAN) are rare diseases [1,2]. These disorders affect the number and distribution of small myelinated and non-myelinated nerve fibers. They are characterized by loss of sensitivity to pain, touches and other sensory or autonomic abnormalities [3,4]. Five different types are described [5].

Hereditary sensory and autonomic neuropathy type V, also known as congenital insensitivity to pain, is a rare autosomal recessive disease, which was first described in 1932 by Dearborn as "Congenital pure analgesia [2].

This disease is characterized by insensitivity to pain with onset at birth and impaired temperature sensitivity, associated with normal tactile and vibratory sensations and a severe reduction in non-myelinated fiber number. Motor and tendon reflexes are intact in most cases [2,6,7].

Self-mutilation is an invariable feature of this disorder, involving the teeth, tongue, lips, fingers, ears, eyes, and nose [4].

This disease is caused by a mutation in NGFB (nerve growth factor- Beta) gene responsible for pain sensation [3,6,8].

Early diagnosis and specific dental management of these patients is important for prevention of oral and dental problems accompanying this disorder [7,9].

This case report describes the different diagnostic elements and oral management, of a 2 years old boy suffering from HSAN type V.

Case Description

A 2 years old baby boy was referred by his pediatrician to the Pediatric Odontology Department at the Monastir Dental Clinic (Tunisia) complaining of tissue loss on the tongue and hands due to biting. The patient was diagnosed with hereditary sensory and autonomic neuropathy type V.

Family history showed that the patient was born of a non-consanguineous marriage and had an older sibling who was also affected by HSAN. The parents were healthy without any specific pathology. Pregnancy and birth were uneventful.

General examination revealed the presence of normal reactions to light touch, tickling, and pressure and normal deep tendon reflexes. Sensory examination showed complete loss of pain and impaired temperature sensitivity.

His physical appearance was normal, with no dysmorphic features. The patient's height and weight were appropriate for his age with normal posture.

Physical examination of his extremities showed that his fingers were short with stubby blunt ends and tissue loss with ulceration on both hands due to biting and chewing (Figure1).

Intraoral examination revealed a bitten tongue aspect with a loss of tissue and ulceration on the ventral surface of the tongue.

The patient presented a complete primary dentition, except for the second deciduous molars maxillary and mandibular, which were not erupted.

Intraoral hard-tissue examination demonstrates no carious lesions (Figure 2).

To prevent self-mutilations to the tongue, lips and extremities, acrylic protector splint were indicated. First silicone impressions were taken from both jaws. Acrylic protector splints were constructed to cover the incisal edges to prevent damage by the teeth. (Figure3).

The splints were bonded to the patient's teeth with glass ionomer cement (Figure4). The patient's parents were then instructed regarding oral hygiene.

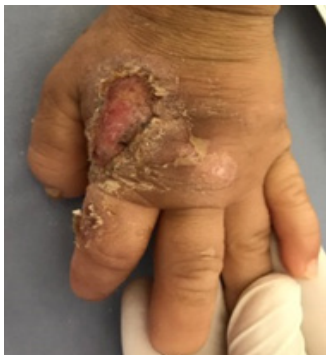

Figure 1: Physical examination

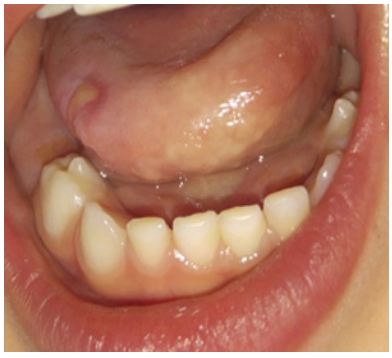

Figure 2: Intraoral examination

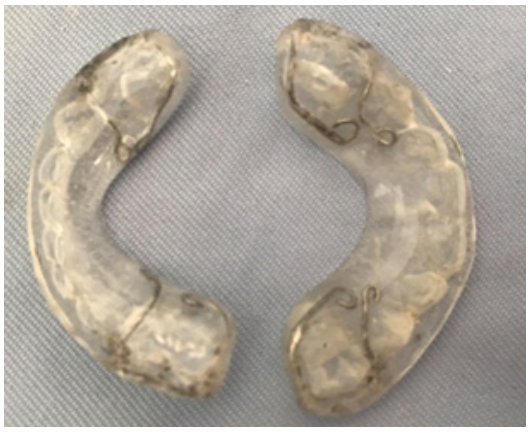

Figure 3: Acrylic protector splints

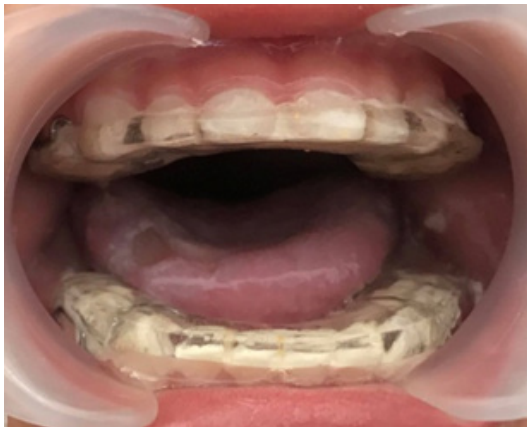

Figure 4: Acrylic protector splints bonded to the patient's teeth

Discussion

Congenital insensitivity to pain is a rare disorder in which oral manifestations can lead to its diagnosis.
It is caused by a mutation in the neurotrophic tryosine kinase receptor, type 1 (NTRK1) gene located on chromosome 1 (1q21-q22) coding for nerve growth factor beta (NGFB) gene. NGFB is the gene encoding the NGF protein and responsible for pain sensation [3,6,8].

Owing to the lack of pain sensation, patients often use their teeth to harm themselves, which may result in severe injuries to oral tissues, including laceration and ulceration of tongue, lips, and oral mucosa. In most cases, these bite injuries start as the primary dentition erupts [2,6,10].

Others traumatic lesions, such as, bites, and digital amputations, and self-tooth extraction can be reported [10,11]. Auto extraction

may be more frequent among subjects with mixed or permanent dentition, as both erupting and shedding teeth are mobile and can easily be removed [7].

The child with hereditary sensory and autonomic neuropathies needs specialized care and a multidisciplinary team approach comprising the neurologist, psychologist, family physician, and pediatric dentist to provide him the holistic care approach. The dental team should be actively involved in the management of these patients as soon as the diagnosis is made, thus preventing the development of complications [4,9,10].

Presently, there is no specific treatment for hereditary sensory and autonomic neuropathies and there are no standard techniques to prevent or treat oro-facial self-inflicted injuries. The treatment plan is predicated according to the circumstances of each individual case [10].

Some methods for prevention of these injuries have been suggested including elimination of sharp surfaces of the teeth by grinding or addition of composite [4,7,9,10]. The use of protective splints or intraoral appliance can also be used but it is often difficult because the mutilation may begin with the eruption of primary incisors. This alternative treatment plan is only possible when the deciduous molars are erupted and able to retain the splints [4,9,12].

This protective splint is used to cap mandibular or maxillary teeth or both in an attempt to break the lip and tongue biting habit [11]. In our case, we considered the use of an oral appliance is the best option.
Training of the family regarding dental-gingival hygiene is also important.

In case of severe mutilation, teeth extraction can be indicated. It is an extremely radical treatment that should be avoided as it will have a psychological impact on children [4,9]. Preventive dental care such as pit and fissure sealants, oral prophylaxis and professional topical fluoride application must be done. In fact, caries can progress to pulpal involvement without causing pain and may lead to infection and tooth loss [9,10].

Moreover, careful monitoring should continue throughout the patient's lifetime, along with comprehensive dental care to maintain the patient's social, psychological and behavioral rehabilitation [7].

Conclusion

Children with hereditary sensory and autonomic neuropathies need specialized care and a multidisciplinary team approach. The dental team should be actively involved in the management of these patients as soon as the diagnosis is made, thus preventing the development of complications.

References

1. Romero M, Simón R, Garcia-Recuero JI, Romance A (2008) Dental management of oral self-mutilation in neurological patients: A case of congenital insensitivity to pain with anhidrosis. Med Oral Patol Oral Cir Bucal 13: E644-647.
2. Jonathan B, Padraig F, Webb D (2006) Congenital insensitivity to pain - review and report of a case with dental implications. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 101: 58-62.
3. Haga N, Kubota M, Miwa Z (2013) Epidemiology of hereditary sensory and autonomic neuropathy type IV and V in Japan. Am J Med Genet Part A 161: 871-874.
4. B SEZG N BOLGÜL, N HAMAMCI, EA AçKIRAN, S ÇELENK, B AYNA (2010) Congenital Insensitivity to Pain: A Case Report with Dental Implications. HK J Paediatr 15: 234-237.
5. TL Erdem, I Ozcan, D Ilguy, S Irin (2000) Hereditary sensory and autonomic neuropathy: review and a case report with dental implications. Journal of Oral Rehabilitation 27: 179-182.
6. Yozou A, Haga N, Funto T, Owaki D, Chiba R, et al. (2015) Hereditary sensory and autonomic neuropathy types 4 and 5:Review and proposal of a new rehabilitation method. Neuroscience Research 104: 105-111.
7. Singla S, Marwah N, Dutta S (2008) Congenital intensitivity to pain. Journal of Dentistry for Children 75: 2.
8. Kalaskar R, Kalaskar A (2015) Hereditary sensory and autonomic neuropathy type V: Report of a rare case. Contemp Clin Dent 6: 103-106.
9. Vidya K, Latha A, Prakash CS (2018) Congenital Insensitivity to Pain-Boon or Bane???: A Case Report with Dental Implications. Journal of Dental & Oro-facial Research 14: 67-69.
10. Neves BG, Roza RT, Castro GF (2009) Traumatic lesions from congenital insensitivity to pain with anhidrosis in a pediatric patient: dental management. Dent Traumatol 25: 545-549.
11. Abdullah N, Kausar SF, Samsudin AR (2015) Congenital Insensitivity to Pain without Anhidrosis: Orodental Problems and Management. Hindawi Publishing Corporation Case Reports in Dentistry 2015:179892.
12. Amano A, Akiyama S, Morisaki I (1998) Oral manifestations of hereditary sensory and autonomic neuropathy type IV Congenital insensitivity to pain with anhidrosis. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 86: 425-431.