Bifid tongue in hereditary sensory and autonomic neuropathy type IV

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Abstract
Hereditary sensory and autonomic neuropathy type IV or Congenital insensitivity to pain with anhydrosis is a rare autosomal recessive disorder. It is characterized by absence of small myelinated and unmyelinated sympathetic nerves resulting in pain and temperature insensitivity. We report a case of a 15 year old boy with hereditary sensory and autonomic neuropathy type IV associated with bifid tongue which is an extremely rare oral presentation and not often reported in literature.

Keywords: Hereditary sensory and autonomic neuropathy type IV, Bifid tongue, Self mutilation.

Introduction
Hereditary sensory and autonomic neuropathy (HSAN) is a rare syndrome characterized by congenital insensitivity to pain, temperature changes and autonomic nerve formation disorders. It is classified into five types namely: sensory radicular neuropathy (HSAN I), congenital sensory neuropathy (HSAN II), familial dysautonomia or Riley Day Syndrome (HSAN III), congenital insensitivity to pain with anhidrosis (HSAN IV) and congenital indifference to pain (HSAN V).[1]

Congenital insensitivity to pain is a rare disorder and was first described in 1932 by Dearborn as ''Congenital pure analgesia''.[2] HSAN type IV is inherited as an autosomal recessive trait and is characterized by recurrent episodes of unexplained fever, failure to thrive, absence or decreased perspiration (despite normal sweat glands), insensitivity to pain, self mutilation and mental retardation.[3] Presently, fewer than 60 cases have been reported in the medical literature.[4] The most common oral manifestations are signs of self-mutilation in the form of ulcerations of the lower lip, tongue, and oral mucosa. Self-extraction of the teeth has also been reported.[5,6] The presentation of bifid tongue is a rare entity and only few cases are reported in literature till date.[7]

Case Report
A 15-year-old male patient reported to the Department of Oral medicine and Radiology with the complaint of decayed tooth in the lower right back region since 1 week. His medical history revealed that he was diagnosed with hereditary sensory autonomic neuropathy type IV and is epileptic since birth. Past dental history showed uneventful extractions a year back. There was no relevant family history reported. On general examination, patient was co-operative with mild mental retardation, slurred speech and was physically challenged. Bulbous terminal phalanges of upper limbs with dystrophic nails were noticed (Fig. 1). Patient was under treatment by pediatrician for osteomyelitis of left ankle with charcot's arthropathy at the time of dental visit (Fig. 2). Extra-oral examination showed convex facial profile due to mandibular deficiency (Fig. 3). Scar was noticed on the bridge of the nose and on the cutaneous surface of the lower lip. There was decreased vision of left eye. Submental and bilateral submandibular lymph nodes were palpable and tender. On intra oral examination, soft tissue lacerations were noticed on the lower lip due to repeated biting of the lip. Bifid tongue was also noticed (Fig. 4). The patient's oral hygiene was poor with multiple root stumps, decayed teeth and multiple missing teeth in all the quadrants. Based on the history and clinical examination a provisional diagnosis of chronic irreversible pulpitis with respect to 46, and 47 was given. The patient was referred to Department of Oral Surgery for extraction of the root stumps. Restoration of the decayed teeth with oral rehabilitation was also done. Patient along with his parents were advised to follow the strict oral hygiene measures to prevent the further dental problems.

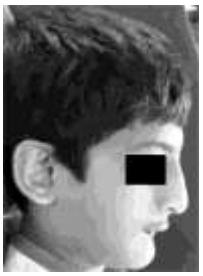

Fig. 1: Showing scars on the dorsal surface of hands with bulbous terminal phalanges of upper limbs with dystrophic nails.

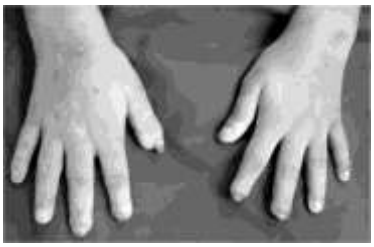

Fig. 2: Showing scars with the swelling in the foot region.

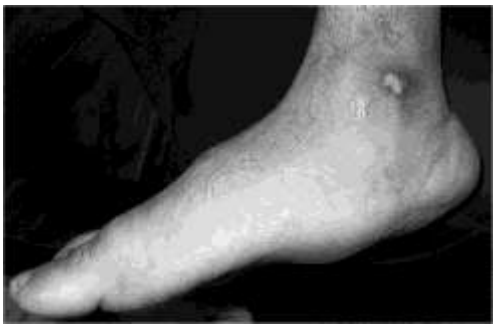

Fig. 3: Shows convex facial profile with mandibular deficiency.

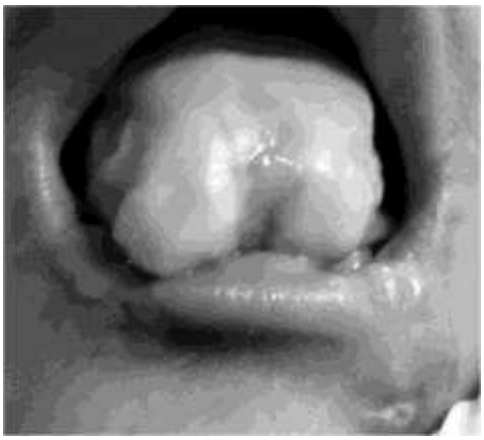

Fig. 4: Showing scars and laceration of the lower lip with bifid tongue.

Discussion

HSAN type IV is the second most common HSAN and like the other recessively transmitted HSAN, its onset is in infancy.[8-10] It is a sensory syndrome in which the patient does not respond to painful stimuli. Recent researches point towards gene mutations as the cause of this syndrome. The mutations are in the neurotrophic tyrosine receptor kinase 1 gene, which is the receptor for nerve growth factor.[11]

HSAN type IV is characterized by anhidrosis, absent or markedly decreased sweating.[8-10,12] It is anhidrosis that causes episodic fevers and extreme hyperpyrexia which is usually the earliest sign of this disorder and can cause recurrent febrile convulsions secondary to high environmental temperature.[8,12]

Individuals with HSAN IV have definite problems in healing of ectodermal structures – skin and bone. Fractures are slow to heal and large weight bearing joints appear particularly susceptible to repeated trauma and frequently go on to the development of Charcot joints and osteomyelitis. The latter was seen in our case. Speech is usually clear, but in the present case the speech was slurred. Although hypotonia and delayed developmental milestones are frequent in the early years, strength and tone normalize with age. However, there can be severe learning problems. Hyperactivity and emotional liability are common.[8,10,12]

Congenital insensitivity to pain is a rare disorder in which oral manifestations may be the presenting complaint. Self mutilation of the tongue and lips are frequently seen with resultant scarring and deformation.[1,13,14] Similar findings were noticed in the case presented here. These injuries often begin as the primary dentition erupts. Tooth luxation and severe dental attrition have also been observed.[13,14] The oral manifestations are especially characteristic in HSAN; however, the dental features have been described in only a few reported cases, as this disorder is exceedingly rare.[1] In the present case, slurred speech was noticed probably due to presence of bifid tongue. Till date only one case of bifid tongue associated with HSAN IV in a 9-year-old girl reported in the literature.[15]

Presently, there is no specific treatment for congenital insensitivity to pain or the other hereditary sensory and autonomic neuropathies. Several methods for prevention of these injuries have been suggested, including elimination of sharp surfaces of the teeth by grinding or addition of composite, the use of mouth guards and other appliances and extraction of teeth. In the present case, extraction of the root stumps was done and instructions to maintain oral hygiene were advised. The use of intraoral appliances is often difficult or impossible to implement because the mutilation may begin in infancy with the eruption of the primary incisors.[14] Thus, extractions may be unavoidable in cases in which the mutilation is particularly severe.[16,17] There is increase in need for orthodontic treatment due to early loss of primary teeth. Loss of the entire primary dentition at such an early stage may result in loss of arch length in both the maxilla and mandible with marked mesial tipping of the permanent first molars, particularly in the mandible.[18]

Conclusion

The dental team plays very important role in the management of patients with HSAN IV as soon as a diagnosis is made. Along with comprehensive dental care to maintain the patient's social, psychological, and behavioral rehabilitation, careful monitoring should continue throughout the patient's lifetime. Training the

patient as well as family members regarding oral hygiene maintenance should be included in the management protocol. It is important for clinicians to be aware of the potential complications caused by HSAN so that the appropriate treatment can be provided promptly following early diagnosis, thus preventing the development of untoward complications.

Conflict of Interest: None.

Source of Support: None.

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