Congenital Insensitivity to Pain – Boon or Bane??? A Case Report with Dental Implications

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Abstract

Pain is an unpleasant sensory and emotional experience associated with actual or potential tissue damage. It interferes with the daily activity of a person's life. Congenital insensitivity to pain with anhidrosis (CIPA) is a rare, hereditary, autosomal recessive disorder resulting from the mutation of the neurotrophic tyrosine receptor kinase 1 gene (NTRK1). Insensitivity to pain results from loss of nociceptive afferents, while anhidrosis is caused by loss of innervation to the sweat glands. Insensitivity to pain and mental retardation lead to self-inflicted injuries to fingers, lips, and tongue, corneal lacerations, painless bony fractures, joint deformities with consequent chronic osteomyelitis and septic arthritis. Early diagnosis and specific dental management of patients with congenital insensitivity to pain are important for prevention of the characteristic oral and dental problems accompanying this disorder.

Keywords: Congenital Insensitivity, Neuropathy, Dental Implications

1. INTRODUCTION

Pain is a reflexive mechanism of body protection. Tissue damage is indicated in the form of pain and painful stimuli cause reflex withdrawal from the source of such damage[1]. Most of the diseases are manifested in the form of pain and are often the motivation for patients to seek medical or dental treatment. Insensitivity to pain is a symptom in certain disorders which may be congenital or acquired. The Congenital types are present at birth, and the majority are diagnosed in early childhood and are collectively termed as Hereditary Sensory and Autonomic Neuropathies (HSAN- Table 1) [2]. Congenital insensitivity to pain is a rare disorder, first described in 1932 by Dearborn as Congenital pure analgesia[3]. Congenital insensitivity to pain with anhidrosis (CIPA) is an autosomal recessive type of HSAN. Usually it is diagnosed early in infancy, as the patient has high fever because of the inability to perspire. Self-mutilation is an almost invariable feature of this disorder, most often involving the teeth, lips, tongue, ears, eyes, nose, and fingers[1, 4, 5]

The incidence of this disorder has been estimated to be 1 in 25,000 population[6]. Here is a rare case of CIPA in a female baby with generalized absence of pain, anhidrosis and its oral and dental implications.

2. CASE DESCRIPTION

A 13 month old baby girl was referred to the department of Pedodontics, M.S. Ramaiah Dental College and Hospital by a well-known neurologist from the academic point of interest. She was born of a consanguineous marriage and having an older sibling who is not affected. Post-natal history revealed that she had pneumonia 3 hours after birth and was kept in NICU. Increased secretions from the eyes were noticed at 3 months of age and when the parents consulted ophthalmologist; corneal ulcerations were diagnosed. After ophthalmic examinations she was referred for neurological opinion. The child was taken to neurologist at the age of 9 months; CT scan was done which revealed diffuse cerebral (predominantly cortical) atrophy (fig.1). ENMG studies at 10 months reported normal conduction. Further the child was diagnosed as having HSAN type 4. On examination, the clinical findings noted were – delayed milestone, scars on right and left fingers, deformation of tip of the tongue (fig2 and fig 3). Primary maxillary central and lateral incisors were present but mandibular incisors were missing. Parents gave the history of exfoliated lower incisors. Exocrine

secretions like tears and salivation found to be normal.

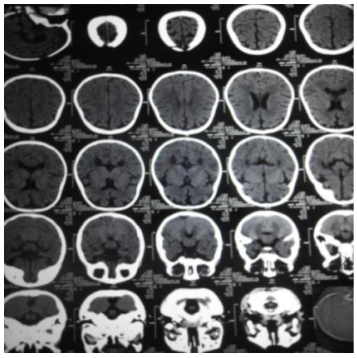

Fig. 1 Showing diffused cerebral atrophy

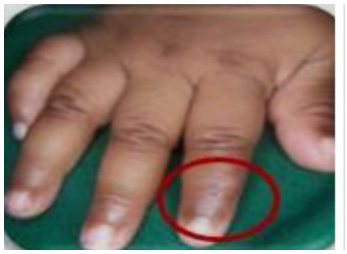

Fig. 2 Showing scar on the index finger

3. DISCUSSION

CIPA is a rare disorder caused by loss of pain sensation, and the teeth represent one means by which these patients can have self-mutilation. As the primary teeth erupt, the oral injuries are noticed. Frequently the tongue and lips are affected, with resultant scarring and deformation[7], as in the present case. Self-mutilative tooth luxation and multiple missing teeth because of self-extraction of the teeth has also been reported[8]. The present case showed missing lower incisors and the parents gave the history of exfoliation; where in it is not very clear whether it had exfoliated or auto extracted.

Decubital ulcer on the ventral surface of the tongue was found to be a typical and common feature in the infantile patients with congenital insensitivity to pain was. Oral scalding could be an evidence of a lack of oral thermal sensation[9]. Absence of fungiform and circumvallate papillae is an important clinical finding in HSAN type IV.There is a report that dental pulp infection resulting from dental caries caused osteomyelitis that led to mandibular bone fracture in a patient with HSAN type IV[10].

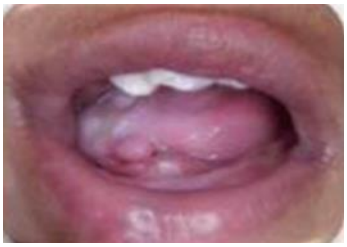

Fig. 3 Showing tongue deformity

Differential diagnosis of the above case could be:

- Congenital insensitivity to pain – HSAN type V
- Lesch Nyhan syndrome
- Ectodermal dysplacia
- Leprosy

At present, there is no specific treatment for HSAN. Some reports in the literature describe a poor prognosis for these disorders[11]. Due to painless injuries, the bones, joints and soft tissues of the extremities as well as the orbits, nasal bones and oral cavity undergo mutilating effects. Bar-On et al have described preventive measures for orthopedic complications such as use of special shoe ware, periods of non-weight bearing, surgical wide debridement and curative osteotomy for deformity[12]. Elimination of sharp surfaces of the teeth by grinding or addition of composites, the use of mouth guards and other appliances and extraction of teeth have been suggested for prevention of dental injuries. The use of intraoral appliance is often difficult because the mutilation may begin with the eruption of primary incisors[9]. However, in case of severe mutilation; extraction is unavoidable[13]. Early loss of primary teeth for any reason is known to lead a significant increase in need for orthodontic treatment.

Late presentations of systemic and dental illnesses put these patients at higher risk. Hence prevention of dental disease is very important in them, as dental caries can progress to pulpal involvement without causing pain and may lead to infection and tooth loss or even osteomyelitis. The dental team should be prepared to manage these patients as soon as the diagnosis is made. Also, careful monitoring should continue throughout the life time of the patient.

4. CONCLUSION

Since pain is the precursor of many pathological conditions; absence of pain for any reason may cause a number of adverse conditions and result in injury. As dental practitioners; it is very important for us to be aware of the potential complications caused by HSAN so that the appropriate treatment can be provided promptly. 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 untoward complications. Moreover, careful monitoring should continue throughout the patient's life time, along with comprehensive dental care to maintain the patient's social, psychological and behavioural rehabilitation.

REFERENCES

1. Rasmussen P. The congenial insensitivity-to-pain syndrome (analgesia congenital): report of a case. Int J Paediatr Dent 1996; 6:117-22.
2. Littlewood SJ, Mitchell L. The dental problems and management of a patient suffering from congenital insensitivity to pain. Int J Paediatr Dent 1998; 8:47-50.
3. Dearborn G. A case of congenital pure analgesia. J Nerv Ment Dis 1932; 75:612-5.
4. Hall KH. Paediatric Orofacial Medicine and Pathology. 3rd ed. London: Chapman & Hall Medical; 1994. pp. 330–5.
5. Narayanan V. Oral and maxillofacial manifestations of hereditary sensory neuropathy. Br J Oral Maxillofac Surg. 1996; 34:446–9.
6. Dave N, Sonawane A, Chanolkar S. Hereditary sensory autonomic neuropathy and anaesthesia - a case report.Indian J Anaesth. 2007; 51(6):528–30.
7. Bolgul BS, Hamamci N, Agackiran E et al. HK J Paediatr 2010; 15: 234-37.
8. Thomson CC, Park RI, Prescott GH. Oral manifestations of the congenital insensitivity to pain syndrome. Oral Surg Oral Med Oral Pathol 1980; 50: 220-5.
9. Amano A, Akiyama S, Ikeba M et al. Oral manifestations of hereditary sensory and autonomic neuropathy type IV. Oral Surg Oral Med Oral Path Oral Radiol Endod 1998; 86: 425-31.
10. Kubo T, Kubota K, Banba S et al. A case report: mandibular osteomyelitis and morbid fracture of mandible caused by dental caries in patient with congenital insensitivity to pain with anhydrosis. Japanese Journal of Dental Handicaps 1997; 18:96.
11. Erdem TL, Ozcan I, Ilguy D et al. hereditary sensory and autonomic neuropathy: review and a case report with dental implications. J Oral Rehab 2000; 27:180-3.
12. Bar-On E, Weigl D, Parvari R et al. congenital insensitivity to pain. J Bone Joint Surg 2002; 84: 252-4.
13. Butler J, Fleming P, Webb D et al. Congenital insensitivity to pain – review and report of a case with dental implications. Oral Surg Oral Med Oral Path Oral Radiol Endod 2006; 101: 58-62