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Congenital insensitivity - To - pain with anhidrosis (CIPA): A case report

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Abstract
Congenital insensitivity to pain with anhidrosis (CIPA) is a rare disease characterized by recurrent fever episodes, mental retardation of different intensity, anhidrosis, little or no perspiration and congenital insensitivity to pain. Oral self-mutilation is also a characteristic sign. In this article, we present a case who showed these clinical characteristics and was treated with a dental device to prevent the patient from injuring his tongue. Neuropathies treatment is a great challenge for medical teams. Pediatric dentist should be a part of these teams because of the oral implications that may appear. The recognition and awareness of this rare clinical condition may be helpful in reaching a correct diagnosis and offering symptomatic treatment, to avoid potential tissue damage at an earlier stage of the disease.

Keywords: HSAN 4, self-mutilating, dental management

Introduction
Congenital insensitivity to pain with anhidrosis (CIPA) is characterized by recurrent episodes of unexplained fever, failure to thrive, absence or decreased perspiration, insensitivity to pain, self-mutilation and mental retardation [1]. Congenital insensitivity to pain with anhidrosis (CIPA) or Hereditary sensory autonomic neuropathy Type IV is an autosomal recessive disorder due to lack of maturation of small myelinated and unmyelinated fibers of peripheral nerves, which convey sensation of pain and temperature, therefore, resulting in self-mutilation [2]. There is anhidrosis due to lack of innervation of normal sweat glands resulting in recurrent episodes of hyperpyrexia [2]. The classification of various types of HSAN (Hereditary sensory autonomic neuropathy) is based on the inheritance pattern, clinical features, and systems of neurons predominantly affected [3].
Pain perception is important as it leads to a protective response. Absence of pain leads to traumatic and self-inflicted injuries. Biting of oral structures, hands, fingers leads to serious self-mutilating damage [4]. Recurrent episodes of oral ulcerations and healing leads to cicatrisation and limited mouth opening [5].
Presently fewer than 60 cases have been reported in medical literature. We report a case of CIPA with self-mutilation. The aim is to present this rarely encountered disease, and information related to its diagnosis and treatment.

Case Description
A 19-month-old boy was brought to department of Pedodontics and Preventive Dentistry by his parents for treatment of oral ulcers due to self-mutilation. The child started biting his tongue and fingers after his teeth erupted at the age of seven months. According to the mother, the child had no sweating and there was absence of crying after painful stimuli. Parents noticed multiple episodes of fever and absence of sweating since the child was 2 months old. None of these episodes of hyperpyrexia had any localization for fever. The child was unable to tolerate the high summer temperatures. On exposure to sun he would become extremely hot. His developmental milestones were delayed.
On physical examination the child had mild pallor with bites marks over the tongue and tips of the fingers. Multiple ulcers on ear, elbow, hand and scalp (Fig1 A, B, C). On Oral examination his tongue and lower lip had multiple ulcers and tongue was mutilated (Fig 1 D).

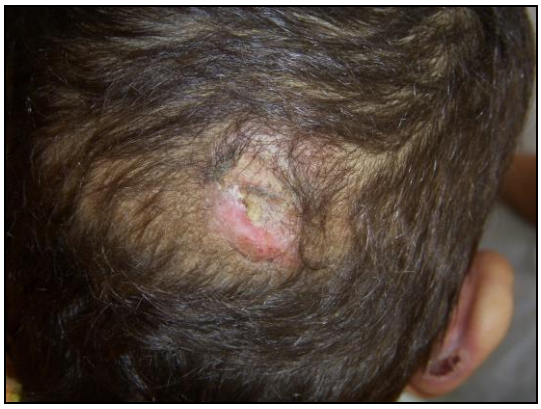

Fig 1(A): Ulcers over scalp and ear

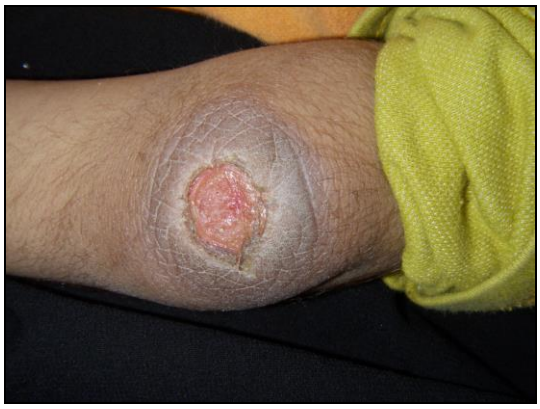

Fig 1(B): Ulcers over elbow

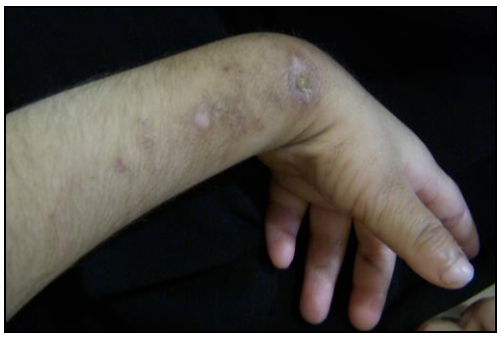

Fig 1(C): Bite marks over wrist

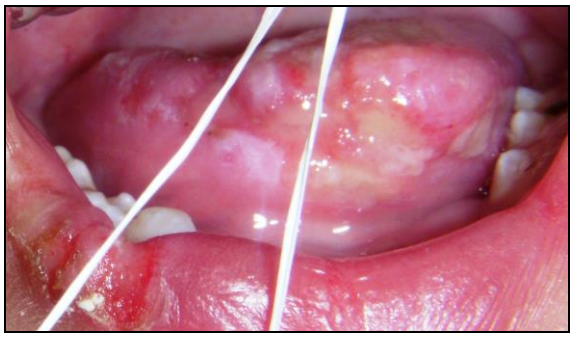

Fig 1(D): Extensive ulceration over tongue and lip with fitted mouth guard

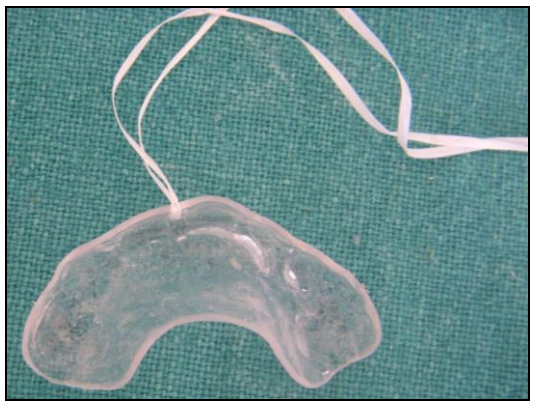

Fig 2: Maxillary mouth guard

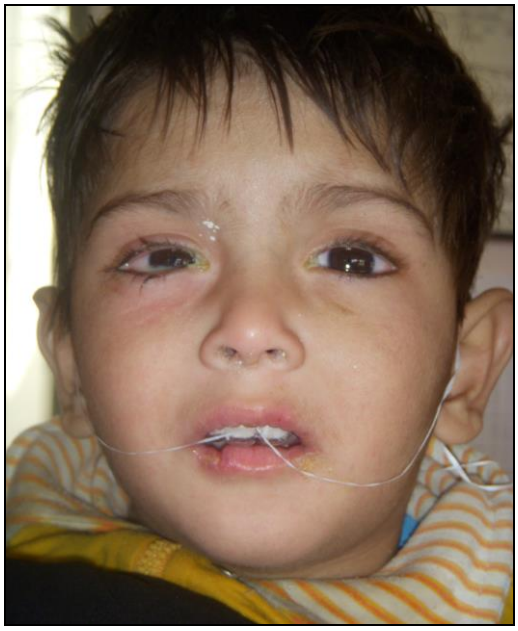

Fig 3: Mouth guard anchored on ear by floss

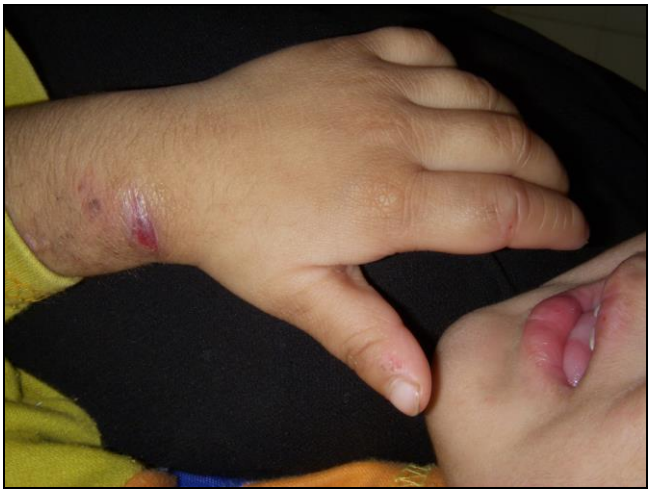

Fig 4: Two weeks after therapy - Healed ulcers over wrist and tongue

His all four lower incisors were missing. Mother gives history of extraction of same teeth by a general dentist. The touch and pressure sensation were normal while pain sensation was absent. On neurological examination, there was hypotonia with normal deep tendon reflexes. There was no family history of paraesthesias, leprosy or tuberculosis. There was history of consanguinity with parents being first cousins, but parents and a six-month old female sibling were normal. Hereditary sensory autonomic neuropathy was suspected with the present case.

Child was previously referred to a general dentist by his pediatrician who advised extraction of all erupted teeth, however his parents sought advice for alternative treatment. Full mouth extraction is an extremely radical treatment and should be the last resort. To avoid psychological and functional implications of tooth loss a decision to preserve the teeth and protect the soft tissues from any further trauma was taken. Several attempts were made to grind the sharp cusps but patient continued to bite. A mouth guard like appliance was planned. A rubber base impression was made and a full coverage maxillary mouth guard was fabricated on model of dental stone. A Biocryl acrylic sheet 1.5 x 12.5 mm thick (Duran Schev - Dental GmbH Germany) was heated and drawn over the cast under vaccum pressure (Ministar). After clear splint was removed from the cast and trimmed to gingival level. It was fitted and retained with dental undercuts and luting cement (Fig 2). A dental floss was incorporated in the mouth guard and anchored to patient's ear to prevent accidental aspiration (Fig 3).

The child accepted the appliance well, which allowed the oral lesions to heal completely within 10 days (Fig 4). The patient is advised new mouthgards to fit the developing dentition as he grows older till he reaches an age where he could be made to understand his problem and help in breaking the habit. The parent's cooperation is important for the successful interception of habit.

Discussion

HSAN IV or CIPA is a disorder caused by mutations of the TRKA/NGF gene. This gene encodes the receptor tyrosine kinase for nerve growth factor (NGF) [6]. The association between CIPA and the TRKA/NGF system suggests that this system plays a major role in development and function of pain perception and thermos-regulatory sweating [7]. CIPA is associated with bouts of pyrexia, anhidrosis and mental retardation. Nerve biopsy reveals absent unmyelinated fibres[8].

The clinical picture of our patients suggests the diagnosis of CIPA. Impaired sense of pain, absence of sweating, signs of self-mutilation and presence of recurrent fever with onset in infancy supports the diagnosis. Mental retardation in HSAN IV is variable, from severe to mild, and some patients were initially reported to be apparently normal, but later mild retardation was showed by a formal assessment[9]. Impairment of pain sensation and oral mutilation have been reported in some syndromes such as Lesch-Nyhan syndrome, Tourette syndrome and de Lange syndrome [10] Absence of nail and hair abnormalities and presence of normal sweat glands on skin biopsy exclude the anhidrotic ectodermal dysplasia [11].

Managing such cases is challenging for the Pediatric Dentist as this condition do not have any cure. Absence of pain, developmental and psychological problem, functional impairment due to early loss of teeth further making the case difficult to manage.

The aim of management is to prevent self-mutilation of oral structures by teeth. There are different treatment options to achieve this goal. Simple grinding of sharp tooth edges can be tried, but is not always successful. Extraction of all erupted teeth can be done but it is too radical procedure and is a last alternative [4].

Mouth guard is an effective appliance to treat such cases. They are easy to build, wear and clean with minimum chances of aspiration. Mouth guards can effectively prevent trauma to all structures by teeth. Parents Co-operation, and regular changing of mouth guards will be required till the patient is old enough to appreciate and avoid self-mutilating behaviour.

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