

Dental management of oral self-mutilation in neurological patients: A case of congenital insensitivity to pain with anhidrosis

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Abstract

Hereditary sensory and autonomic neuropathy type IV is a rare disease characterized by fever episodes, mental retardation of different intensity, recurrent episodes of fever secondary to anhidrosis, little or no perspiration and congenital insensitivity to pain. Oral self-mutilation is also a characteristic sign.

In this article, we present the case of an infant, aged 22 months, who showed these clinical characteristics and was treated with a dental device to prevent the patient from injuring her tongue. This device consisted of two acrylic splints joined at the back in the posterior sector, it provided an anterior open bite and allowed the infant to breathe through her mouth.

The lesions of the patient had improved after using the device but the patient died due to the medical problem.

Neuropathies treatment is a great challenge for medical teams. Dentists should form part of these teams because of the bucal implications that may appear. Different appliances can be designed in order to solve the special problems each case may present.

Key words: *Self Mutilation, neurological patients, insensitivity to pain, anhidrosis, hereditary sensory autonomic neuropathy type IV.*

Introduction

Hereditary sensory and autonomic neuropathies are rare diseases characterized by a decrease of pain perception. Hereditary sensory and autonomic neuropathy type IV of the Dyck classification (1), also known as congenital insensitivity to pain with anhidrosis, is inherited with a recessive autosomic pattern. This disease is characterized by mental retardation, recurrent episodes of fever secondary to anhidrosis if exposed to high temperatures, little or no perspiration, and alteration of the pain and thermal sensitivity. (2)

Without pain perception, there appear frequent traumatic injuries and large self-injuries.(3) Patients normally injure themselves using their teeth.(4) Frequently, we observe finger biting, lacerations and ulcerations of the tongue,

lips and other areas of the oral mucosa. Furthermore, it is normal to find dental luxations and severe attrition. (2) The fact that the self-injurious behavior continually occurs implies a delay of wound cicatrization and facilitates overinfection. (4)

Case Report

A 22 month old infant was admitted as an emergency to '12 de Octubre' hospital in Madrid [Spain] having a fever for the last 12 hours and tongue biting due to convulsive crises.

- Medical History

When the infant was a few months old, the parents realized that she did not sweat and suffered from hyperthermia episodes related to temperature which decreased thanks to physical measures.

At the time of admission, the infant showed extreme hyperthermia ($> 42^\circ\text{C}$), CF 230 beats/min, central cyanosis, inadequate peripheral perfusion and respiratory distress. Orotracheal intubation and connection to mechanical ventilation were carried out. AP reached 75/70 with 170 beats/min CF thanks to physiological saline solution and inotropic drugs (dopamine + dobutamine). Physical and pharmacological measures were adopted to control the hyperthermia.

The first hemogram was normal but, in subsequent hours, it presented leukopenia (3,950 leukocytes) with left shift (47% segmented, 16% band), thrombopenia (47,000 platelets), PCR reached level 4 and coagulopathy (55% prothrombin activity, 20.8 prothrombin time, 61.4 TTPA, 139 fibrinogen) with PDF > 20 . The biochemical study showed an increase of transaminases (316 GOT, 157 GPT, 34 GGT), LDH (1,146) and CPK (1,190).

The cranial CT scan carried out the day of admission and the cranial MR of the following day were normal.

The lumbar puncture showed an opening pressure of H₂O of 20 cm and a normal cytochemistry (1 leukocyte, 71.8 mg/dl glucose, 0.15 g/l protein).

The patient was treated with cefotaxime, vancomycin, acyclovir and antiseizure medication.

In subsequent hours and days, a good hemodynamic and respiratory control was achieved. The EEG showed a marked slowing down of the activity, that did not react to any stimulus. No epileptiform activity was observed. Cranial CT scan was repeated and showed a general cerebral edema. Intracranial pressure was monitored using a Camino catheter.

Once sedation was removed and extubation carried out, the infant became hemodynamically stable and breathed properly, although a serious neurological affection still remained.

The patient recovered the sleep-wake cycles in subsequent days and opened her eyes spontaneously. However, the neurological examination showed a comatose condition. The patient frequently showed agitation episodes, hyperventilation and an opisthotonus condition that could remain for hours if sedation and muscle relaxants were not used. This condition was associated with hyperthermia not with perspiration. A new cranial CAT scan confirmed the serious residual cerebral affection with signs of generalized ischemia. The EEG still showed a serious slowing and toning down of cerebral activity, with no reaction.

All microbiological studies were negative.

No autonomic study was carried out because the patient's condition prevented her from being taken to the neurophysiology laboratory.

The patient died of intercurrent respiratory process. Necropsy was not carried out.

- *Oral Exploration*

The infant was in temporal dentition. Right central and lateral lower incisors were attrited due to serious

bruxism and left central and lateral lower incisors were self-luxated.

An initial exploration showed a serious laceration and inflammation of the tongue. In some areas, we could even observe her tooth marks. (Figure 1)

- *Dental Management*

A device consisting of two acrylic splints joined at the back in the posterior sector was made. The acrylic was thicker at the joint to provide an anterior open bite. This prevented further self-injurious biting, and allowed the infant to breathe through her mouth. (Figures 2 and 3).



Fig. 1. Oral injuries at the time of admission.



Fig. 2. Tongue biting prevention device on casts.



Fig. 3. Device into the mouth.

The advantage of this appliance was that as it covered both, the upper and the lower arch, retention was higher and it could not be forced out of the mouth by tongue movements.

The parents and the nursing staff were trained in the insertion and care of the appliance and it was worn 24 hours a day.

The girl died on the 8th day of placing the dental device. During this time, the tongue started to heal up; she did not bite herself or bleed.

Discussion

The symptoms for which the patient was admitted to the ICU (extreme hyperthermia, encephalopathy, shock, coagulopathy) were consistent with a heat stroke once a systemic infectious process, such as a septic shock or an infection of the central nervous system was rejected. These symptoms were associated with cerebral edema with highly serious neurological consequences, which finally caused the death of the patient, derived from an intercurrent respiratory process.

Clinical manifestations indicated a hereditary sensory and autonomic neuropathy type IV (congenital insensitivity to pain with anhidrosis). In the absence of genetic confirmation, this was the suggested diagnosis as the neurophysiologic study of the autonomous nervous system and the histology of the peripheral nerve could not be carried out.

20% of the patients suffering from congenital insensitivity to pain with anhidrosis die of hyperthermia at early stages of life.(5)

There are different neurological syndromes associated with oral self-mutilation, such as the Lesch-Nyhan syndrome (6 - 8), the Cornelia de Lange syndrome and the Tourette syndrome (9). Congenital insensitivity to pain with anhidrosis frequently appears together with this problem. Oral manifestations of this syndrome are characteristic and can help to obtain an early diagnosis.

The most frequent oral finding is self-mutilation of the tongue, followed by lip and oral mucosa self-injuries (10). Furthermore, these types of patients commonly lose their lower temporal incisors due to self-extraction or luxation derived from the continuous oral self-trauma. (2)

There are diverse possibilities of dental treatment in patients with self-injurious behavior. Occasionally, teeth extraction has been recommended, in other occasions to grind the sharp edges of the teeth, or even the use of dental devices to prevent the self-trauma of oral tissues (10).

Numerous devices have been proposed to prevent oral self-injuries. Willette (11) in a case of self-inflicted lip injury, describes a lip shield to separate the lips and cheeks from the teeth, but in our case, this appliance would not have prevented self-inflicted tongue injury. Fabiano (12) placed a mandibular occlusal bite plane in a patient with multiple sclerosis, but there are two important differences

in relation to our patient, first his patient could collaborate, and second our patient had temporal dentition and she had lost lower incisors which made retention of a mandibular bite plane very difficult.

A maxillary occlusal splint like the one used by Walker (13) or a maxillary mouthguard like the one used by Littlewood (3) would have posed the same problems of collaboration and retention. A tongue protector, like the one described by Kozai (14) could not be used because lower incisors were lost, and besides the child used not only the incisors, but all her teeth to injure her tongue. An acrylic tongue stent as described by Peters (15) could have been used, but we believe that the wire fixation he uses would have produced more disturbances to such a young patient as the child we present.

In this case, we chose a device that provided an anterior open bite, let the tongue cicatrize and prevented the infant from self-injury. We chose this appliance because as we have explained before it was difficult that it was forced out of the mouth by tongue movements. All the appliances that we have mentioned above are less retentive than the one we used. Moreover as she was fed thanks to a nasogastric catheter, the anterior open space left by the appliance let the infant breathe through her mouth.

The patient died of an intercurrent respiratory process on the 8th day of placing the dental device. We confirmed that it was effective as the patient stopped biting her tongue and it started to cicatrize.

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