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Case Note

Congenital insensitivity to pain: A rare encounter in Orthodontics

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Abstract

Congenital insensitivity to pain (CIP) is a very rare disease which is not commonly described in books or literature. The patients suffering from this disease have autonomic neuropathy with hereditary component, described as Hereditary Sensory Autonomic Neuropathy (HSAN). At present five types of HSAN have been identified. Here we present a case of HSAN type IV. The main goal of our treatment was to assist the healing of grossly mutilated tongue and to prevent further damage, which was achieved by fabrication of a removable orthodontic appliance.

Key words: Congenital insensitivity to pain, Hereditary Sensory Autonomic Neuropathy, Tongue mutilation, Thomas criteria.

Introduction

Congenital insensitivity to pain (CIP) is a medical condition in which patient seems not to perceive sensation of pain[1]. It means they have a markedly impaired ability to perceive the type, intensity and quality of painful stimuli. The disease is very rare and hardly ever described in textbooks[2,3].

Generally patients affected by this condition often die in early childhood as they fail to notice injuries and illnesses. Pain is a typical sensory experience that may be described as the unpleasant awareness of a noxious stimulus or bodily harm. It is highly protective phenomenon developed by nature. The sensation of pain protects human and other species from the tissue damaging effects of dangerous stimuli and appears to be critical for survival of the organism.

Case report

A six year old male patient came to the Department of Orthodontics, Zhengzhou University, China, with the chief complaints of ulceration on the tongue and lack of pain sensation. On further interviewing, patient's mother stated that the boy never complained of pain. He also had recurrent fever.

On examination the tongue was grossly mutilated (Fig. 1). There were also multiple scar marks, bruises and cuts on the fingers, face and scalp area. The patient had no excessive sweating. Patient had no pain while pinching or pricking with pointed object on the skin. He also had loss of heat sensation. However he could feel pressure and light touch on the skin. Clinical and radiological examinations revealed absence of multiple deciduous teeth and several erupting permanent teeth (Fig. 2). The patient's mother could not recall whether the teeth were removed by the patient himself or due to any other reason.

The diagnosis of congenital insensitivity to pain is mostly clinical. Criteria that excluded individuals with acquired lesions were proposed for CIP[1]. The majority of the patients have a type of Hereditary Sensory and Autonomic Neuropathy (HSAN). Nagasako et al[3] defined the criteria for five different types of HSAN based on Thomas' findings[4]. Depending on the criteria of Nagasako et al our patient was diagnosed to be a case of HSAN type IV.

Though some reports in literatures state that some sensation in the extremities may develop as the child

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grows, our main aim was to protect the child from further self induced and accidental injuries. So our treatment options were:

1. Just symptomatic treatment
2. Make the patient fully aware of the disease and provide proper counselling
3. Fabrication of the removable intraoral appliance to help reduce and heal tongue mutilation.

We adopted the second and third treatment options after discussing with the patient's parents. Intraoral removable appliance was constructed with self cure acrylic resin covering the palatal mucosa (Fig. 3). The patient was followed up after three weeks and also after one month. There was no further mutilation of the tongue and the wound was healing normally.

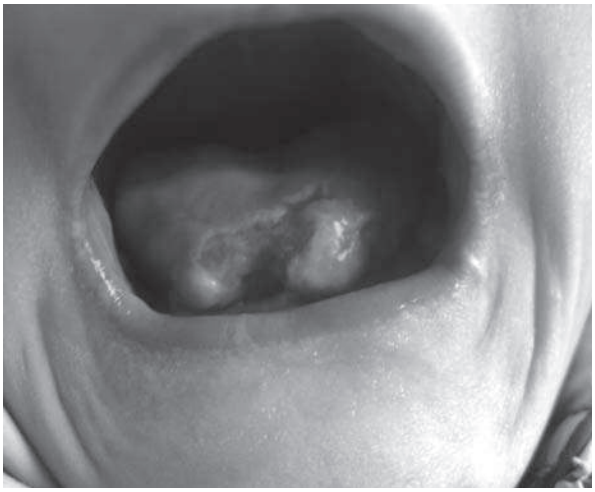

Fig. 1: Self induced ulceration on the tongue

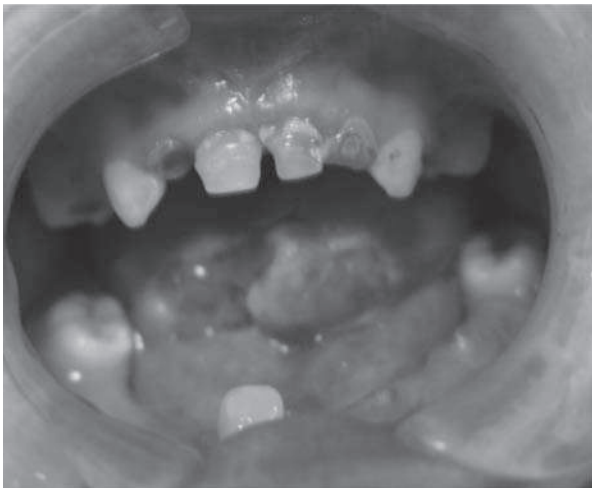

Fig. 2: Absence of deciduous teeth seen clinically

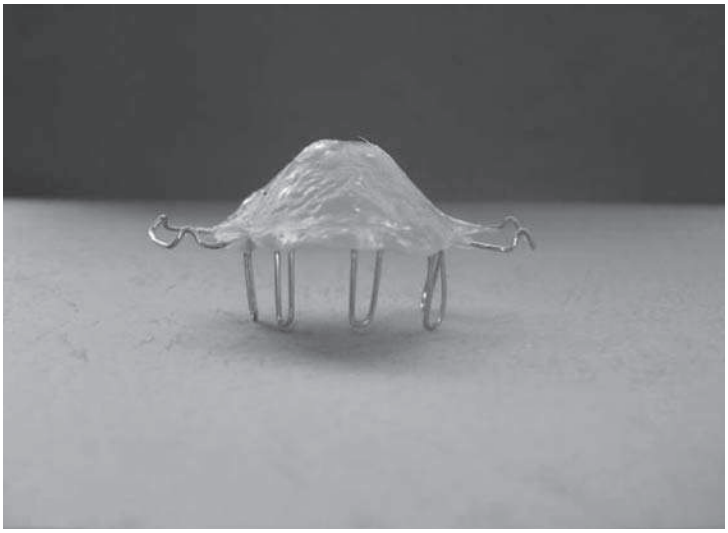

Fig. 3: Removable appliance to protect the tongue from mutilation

Discussion

Reports of the individuals who seemed insensitive to pain from birth onwards have long history but it was not until 1930 that this condition attracted medical attention[3]. Various terms were used in the past to describe those patients including Congenital pure analgesia[5], Congenital universal indifference to pain[6,7] and Congenital absence of pain[8]. Jewesbury used the term Congenital Insensitivity to pain[2].

The present case was referred to us due to two reasons, firstly due to absence of several teeth and secondly due to tongue mutilation. Congenital indifference to pain associated with oligodontia is rarely mentioned in the literature[9,10,11]. The differential diagnosis of the disease were Riley-Day syndrome[12,13], Hereditary sensory radicular neuropathy[14,15] and HSAN IV. In Riley-Day syndrome and Hereditary sensory radicular neuropathy, the sweat gland is not affected. In this particular case the clinical findings best fits the features of HSAN IV[16].

Patients with CIP usually go to the pediatricians with early sign of elevated body temperature[4].

CIP associated with self extraction was reported by Sato Fumihiko et al[9]. He reported that, because of the absence of pain, the patient extracted his teeth prematurely and caused decubitus ulcer of buccal mucosa due to displaced eruption of third molar. The eruption of the teeth may be crucial point in the development of self mutilation. The incisors are the child's sharp weapons which the child can use to damage tongue, cheeks, lips and fingers[17,18,10].

Conclusion

The rare disease Congenital indifference to pain may be associated with dental anomalies.

Patient with CIP may present to dental surgeons seeking treatment of mutilated tongue and oral structures.

Proper counselling and intraoral removable appliance to disrupt the self mutilation may be helpful for healing and to prevent further damage to the tongue.

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