Abstract
Hereditary sensory and autonomic neuropathy is a rare syndrome which is seen in early childhood. Five different types have been described. Absence of pain and self-mutilation are characteristic findings of this syndrome. This report describes one female and two male children with the syndrome. The most severe oral consequence of their disorder was damage to the oral tissues and tongue. The primary aim in management was to monitor the eruption of the permanent teeth.

Key words
Hereditary sensory and autonomic neuropathy · Insensitivity-to-pain syndrome · Congenital analgia

Introduction

Congenital analgia syndrome is characterized by the absence of sensitivity to subjective pain, which is a protective mechanism for the body. Absence of pain sensation is a symptom in several neuropathies. Sensory neuropathies may be grouped into two main categories: congenital and acquired. As the name implies, the congenital types are present at birth, or at least are diagnosed in early childhood; they are not caused by disease or trauma.

A genetic background is suggested for most of the hereditary sensory neuropathies, but the mode of transmission is still unclear. Autosomal dominant transmission has been suggested, but in cases in which the condition has been documented in several members of a family, autosomal recessivity seems best to explain the mode of transmission. According to Leiber and Olbrich, it is, in all probability, a simple recessive congenital disease.

Several types of inherited sensory neuropathies may be distinguished (Table 1), based on whether the neuropathy is hereditary, sensory, and autonomic (HSAN), or hereditary non-neuropathic analgesia (HNNA). The differential diagnosis is made according to the presence or absence of additional features such as mental retardation, oral lesions, loss of touch and temperature sensation, deep tendon reflexes, axon reflexes, production of tears and sweat, and abnormal nerve histology. In addition, secondary skeletal disorders, such as fractures, epiphysiolysis, and osteomyelitis, may appear, because the patients are more exposed to trauma. A conspicuous feature is also the many self-inflicted traumas that most often involve the teeth, lips, tongue, ears, eyes, nose, and fingers.

With the exception of HSAN type III, the hereditary sensory syndromes seem to be extremely rare. Presentations are therefore often single cases, and few publications deal with the oral manifestations. In this article, three cases are reported, which best fit the description of congenital sensory neuropathy and anhidrosis (HSAN type IV).

Case 1

The patient was first seen by the authors when she was 2 years of age, and was followed until the age of 5 years. She was the only child of her parents. It was found in her family history that her parents were first-degree relatives, and hers was the only case in the members of the extended family. The patient’s medical records through early childhood showed that pregnancy and birth had been uneventful. Her mental development was retarded, and a diagnosis of
HSAN type IV was made at age 1 year by the physicians at the Department of Pediatric Health and Diseases of the Istanbul University Faculty of Medicine. Pain, touch, and temperature sensation, and proprioceptive, perception, seemed to be affected. The production of tears and sweat was reduced.

The main medical problems of case 1 had been keratitis of the right eye, fissured lips, and some scars and wounds on her tongue and lips (Fig. 1). The periodontal tissues were intact. Visible injuries, caused by the erupted teeth, were found on the underside and the edges of the tongue (Fig. 2). Some of the open sores were necrotic or covered with fibrin. The maxillary alveolar process and the hard palate were damaged at this age. A soft acrylic splint was made to fit the upper and lower arch (Fig. 3). An open space was left over and around the erupting teeth to allow free vertical movement. The splint prevented the teeth being touched by the patient, protected soft and hard oral tissues from further injuries, and discouraged tongue thrusting. After a few weeks the wounds had healed, and the condi-

Table 1. Hereditary sensory syndromes

<table>
<thead>
<tr>
<th>Condition</th>
<th>Inheritance</th>
<th>Mental retardation</th>
<th>Oral lesions</th>
<th>Touch sensation</th>
<th>Temperature sensation</th>
<th>Tears</th>
<th>Sweat</th>
</tr>
</thead>
<tbody>
<tr>
<td>HSAN type I; hereditary radicular neuropathy</td>
<td>AD</td>
<td>No</td>
<td>No</td>
<td>Reduced+</td>
<td>Reduced++</td>
<td>Normal</td>
<td>Distal loss</td>
</tr>
<tr>
<td>HSAN type II; congenital sensory neuropathy</td>
<td>AR</td>
<td>No</td>
<td>Yes</td>
<td>Reduced++</td>
<td>Reduced+</td>
<td>Diminished</td>
<td>Distal loss</td>
</tr>
<tr>
<td>HSAN type III; familial dysautonomia (Riley Day)</td>
<td>AR</td>
<td>Could be retarded</td>
<td>Yes</td>
<td>Normal</td>
<td>Reduced+/-</td>
<td>Absent</td>
<td>Erratic</td>
</tr>
<tr>
<td>HSAN type IV; congenital sensory neuropathy and anhidrosis</td>
<td>AR</td>
<td>Could be retarded</td>
<td>Yes</td>
<td>Reduced+/-</td>
<td>Reduced+/-</td>
<td>Normal</td>
<td>Absent/ reduced</td>
</tr>
<tr>
<td>HSAN type V</td>
<td>AR</td>
<td>No</td>
<td>Yes</td>
<td>Reduced++</td>
<td>Reduced+/-</td>
<td>Normal</td>
<td>Absent/ reduced</td>
</tr>
<tr>
<td>Hereditary non-neuropathic analgesia (HNNA)</td>
<td>AR</td>
<td>Normal or retarded</td>
<td>Yes</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
</tbody>
</table>

Summarized from Gorlin et al.®

HSAN, Hereditary sensory and autonomic neuropathy; AD, autosomal dominant; AR, autosomal recessive

Fig. 1. Case one at 2 years of age

Fig. 2. Labial and oral deformity due to biting

Fig. 3. Application of splint on upper and lower arch