



Oro-Dental Management of a Recessive Case of Thévenard Disease in Childhood: A Case Report

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Abstract

Introduction: Thévenard disease, or hereditary sensory neuropathy type 1, is a rare genetic disorder characterized by sensory loss, recurrent ulcerations, and progressive osteoarticular destruction. While systemic manifestations are well described, oral involvement—especially in recessive forms—remains poorly understood. This article aims to report the oral features of a child with recessive Thévenard disease and to outline specific challenges in dental management.

Case Presentation: A 7-year-old female with recessive Thévenard disease was referred for management of a dental infection. Medical history revealed early-onset febrile episodes with neuropathic pain, chronic cutaneous ulcers, and osteoarticular deformities. Oral examination identified mandibular deviation, incisor-molar hypomineralization (MIH), severe tooth wear, and necrosis of mandibular molars associated with cellulitis. Radiographs showed delayed root maturation and multiple pulp and periapical lesions. Dental management included extractions, preventive, and restorative treatments, pulpotomy, and selective occlusal adjustments. A removable partial denture was fabricated to restore occlusion, and an anterior crossbite was later corrected using a composite inclined plane. The patient was followed for one year, with stable outcomes and improved oral function. **Discussion:** This case highlights the potential oral complications of Thévenard disease and the need for individualized, multidisciplinary care. Dental management must integrate preventive strategies, behavior guidance, and careful monitoring due to altered pain perception, infection risk, and psychological factors. Early recognition of oral manifestations may improve quality of life and reduce morbidity in affected patients.

Subject Areas

Dentistry, Pediatrics

Keywords

Thévenard Disease, Hereditary Sensory Neuropathy, Oral Manifestations,

1. Introduction

Thévenard disease, also known as hereditary sensory neuropathy type 1, is a rare autosomal disorder primarily affecting the feet, with or without involvement of the hands. It is characterized by the development of cutaneous ulcers that slowly progress to bone deformities and osteoarticular destruction. These complications result from distal thermoalgesic anesthesia and can severely compromise mobility, thus affecting patients' quality of life [1] [2].

Although the general clinical manifestations of this disease are well described in the literature, data on its oral repercussions remain extremely limited. To date, few publications have documented oral complications, particularly in the recessive form of the disease [3] [4].

This report presents a case of a young female patient with Thévenard disease managed at the Pediatric Dentistry Department of the Casablanca Dental Consultation and Treatment Center (DCTC) in Casablanca. The aim is to describe the observed oral manifestations, highlight specific management considerations and precautions, and propose clinical recommendations for this rare condition.

2. Case Report

A 7-year-old female patient was referred to the Pediatric Dentistry Department of Casablanca Dental Consultation and Treatment Center for a dental infection involving teeth 74 and 75. The medical history revealed a diagnosis of hereditary sensory neuropathy type 1. Parental consanguinity was noted, with no similar family history. The diagnosis of the recessive form was supported by the absence of any clinical manifestations in both parents, suggesting a non-dominant mode of inheritance.

Her medical history included febrile episodes associated with severe neuropathic pain beginning at six months of age. At nine months, she developed her first ulcer on the thumb, which became infected and healed poorly. Similar lesions later appeared on other fingers and toes. The diagnosis of Thévenard disease was established at the age of two years. The patient is currently on lifelong intermittent antibiotic therapy (penicillin) and receives opioid analgesics during acute pain crises. Behavioral history reported by the mother includes nail-biting and bruxism. The patient also exhibits moderate sensory deficits, making local anesthesia sometimes ineffective, and receives psychiatric care for associated learning difficulties.

General examination showed cutaneous ulcers and osteoarticular deformities affecting the hands and feet (**Figure 1**). Facial examination revealed pronounced dark circles and oral dryness (**Figure 2**). Intraoral examination showed mandibular deviation and an inflammatory swelling adjacent to teeth 74 and 75 (**Figure 3**).



Figure 1. Cutaneous ulcers and osteoarticular destruction of the hands and feet.



Figure 2. Facial examination showing pronounced dark circles and oral dryness.



Figure 3. Endobuccal views revealing. (a) Mandibular deviation; (b) multiple caries; (c) swelling near the 74, 75.



Figure 4. Panoramic radiograph showing complete dentition (except third molars), generalized tooth wear, periapical radiolucencies, and delayed root development.

Panoramic radiography (**Figure 4**) revealed a complete dentition except for the absence of third molar germs. Extensive tooth wear was evident on all teeth. Radiolucencies affecting the pulp, periapical radiolucent areas encompassing the

roots of teeth 74 and 75, and furcation involvement were observed, along with delayed root maturation of permanent teeth relative to the patient's chronological age (**Figure 4**).

Based on clinical and radiographic findings, the diagnosis included left-sided lower facial cellulitis related to necrosis of teeth 74 and 75, superficial to moderately deep amelodentinal lesions on the first four permanent molars and tooth 84, a juxta-pulpal lesion on tooth 54, and incisor-molar hypomineralization (MIH).

Before dental treatment, a letter was sent to the treating physician, who confirmed that dental management could proceed, considering the patient's psychological profile and allowing progressive adjustment of anesthetic doses as needed. Emergency management began with extraction of teeth 74 and 75. Behavioral management was based on the Tell-Show-Do technique, with specific adaptations to accommodate the patient's sensory deficits and learning difficulties. Explanations were simplified and repeated using clear visual demonstrations, as her altered pain perception limited the effectiveness of sensory-based guidance. The clinical environment was kept calm, and instruments were introduced gradually to reduce anxiety. Frequent positive reinforcement and a slower procedural pace were implemented to support cooperation and ensure a predictable and reassuring experience for the child. The mother and patient were motivated to maintain good oral hygiene and dietary habits.

The choice of dental procedures was based on the clinical condition of each tooth and the patient's overall functional needs. A pulpotomy was performed on tooth 54 because the tooth was vital, showed no signs of radicular infection, and preserving it was essential for maintaining arch space in a young child. Selective grinding was carried out to correct a functional mandibular deviation caused by occlusal interferences; this conservative approach allowed for immediate improvement in mandibular centering and stabilization of the occlusion without the need for additional orthodontic appliances.

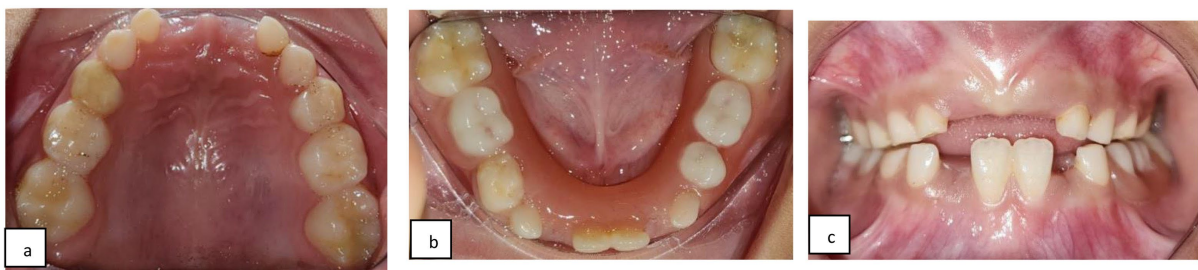


Figure 5. Endobuccal views after oral cavity management. (a) Maxillary; (b) mandibular; (c) improving occlusal vertical dimension.

Subsequently, dental treatment was carried out quadrant-wise, following strict hygiene and asepsis protocols. The procedure began gradually with prophylactic brushing, followed by preventive restorations on teeth 16 and 36, composite res-

tortions on teeth 26, 46, and 84, and a pulpotomy on tooth 54, before selective grinding of teeth 73 and 83. A removable partial denture replacing teeth 74, 75, and 84 was fabricated without clasps to prevent gingival injury. Improved vertical dimension of occlusion was observed after placement of the dental prosthesis (**Figure 5(b)**). Regular follow-up was scheduled. Three months later, eruption of teeth 11, 21, 32, and 42 was observed, along with the development of an anterior crossbite, corrected using a composite inclined plane. The patient was subsequently monitored for one year (**Figure 6**).



Figure 6. Follow up. (a) 3 months after; (b) composite inclined plane correcting anterior crossbite after eruption of the four permanent incisors; (c) one-year follow-up.

3. Discussion

Thévenard disease, or hereditary sensory neuropathy type 1, is an extremely rare genetic disorder manifesting as an ulceromutilating acropathy. It primarily involves distal peripheral sensory neuropathy, resulting in loss of pain and temperature sensation. This insensitivity predisposes patients to chronic cutaneous ulcers that progress to progressive mutilation of fingers and toes, as observed in the present case. The disease is often associated with autonomic dysfunction, which may include hypohidrosis or anhidrosis, heart rate and blood pressure variations. Although classically inherited in an autosomal dominant pattern, isolated recessive forms have also been reported, as in our patient [1] [5].

The disease affects both sexes equally and may present at birth or later in childhood, adolescence, or adulthood. Its progression is typically slow, extending from the second to third decade of life [2] [6]. Clinically, patients exhibit thermoalgesic sensory deficits, exposing them to repeated trauma, particularly at pressure points of the hands and feet. Such trauma often goes unnoticed, leading to painless ulcers, recurrent infections, osteoarthritis, and eventual mutilation. Some patients, however, experience severe pain crises, reflecting complex nerve involvement where insensitivity and pain coexist [5] [6].

Hand involvement is exceptional and usually occurs late in the disease course. Early lesions in the hands, as in this case, indicate a rare, atypical presentation [1]. Oral manifestations of Thévenard disease are scarcely reported. Turk *et al.* described hypodontia in a patient with a recessive form of the disease [7].

The severity of the oral condition at presentation can be attributed in part to the patient's underlying sensory neuropathy. Because pain perception is markedly reduced in Thévenard disease, early symptoms of dental caries likely went unnoticed, delaying care until the infection had progressed to an advanced stage, in-

cluding facial cellulitis. In a typical child, dental pain serves as an early warning sign prompting timely intervention, whereas in this patient, the absence of pain contributed to the late diagnosis and the extensive destruction observed.

The patient's MIH may be related to multiple courses of penicillin during childhood. Some studies suggest a possible link between early antibiotic exposure and MIH, though the evidence remains inconclusive [8] [9].

Dental management challenges in this case are similar to those observed in other sensory neuropathies, such as HSAN1, where reduced pain perception delays the detection of caries and infections. However, the specific pattern of lesions and progression observed here appears unique to Thévenard disease.

Oral management of such patients requires a rigorous psychobehavioral approach to gain the child's trust and cooperation. When necessary, premedication or conscious sedation with nitrous oxide may be employed. Due to prolonged antibiotic exposure, resistance to specific molecules, such as amoxicillin, must be assessed to guide therapy in case of infection. Oral sensory deficits likely contribute to hypoesthesia, complicating early detection of dental lesions and increasing the risk of local or regional infections, as observed in our patient, who presented with cellulitis adjacent to necrotic teeth 74 and 75. Beyond oral manifestations, our patient exhibited psychological challenges, including learning difficulties and bruxism, commonly observed in hereditary sensory neuropathies.

This extremely rare condition, often associated with significant disability, requires meticulous and individualized oral management due to the multiple systemic and oral complications it may cause. The pediatric dentist, as a key factor in the overall care pathway, must work in close collaboration with other medical specialties. Regular follow-up and constant adaptation of dental treatment, in view of the child's specific sensory features, are essential to improve the patient's oral health and, consequently, quality of life.

Although this report describes a single case and its findings cannot be generalized, it provides valuable insights into the oro-dental management of the rare recessive form of Thévenard disease, contributing to the limited literature on this condition.

Conflicts of Interest

The authors declare no conflicts of interest.

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