International Journal of Community Medicine and Public Health
Sulaiman NM et al. Int J Community Med Public Health. 2026 Feb;13(2):965-968
http://www.ijcmph.com
pISSN 2394-6032 | eISSN 2394-6040

DOI: https://dx.doi.org/10.18203/2394-6040.ijcmph20260325

Case Report

Dental management under general anesthesia in a pediatric patient with Netherton syndrome: a case report

Noura M. Sulaiman[1], Mawadh A. Demyati[1], Neda E. Alomar[2], Murtada A. Ahmed[3]*

[1]Department of Pediatric Dentistry, Prince Sultan Military Medical City, Riyadh, KSA
[2]Department of Pediatric Dentistry, Jazan General Hospital (Ministry of Health), Jazan, KSA
[3]Department of Dentistry, Tamam Al-ilaj Medical Complex, Riyadh, KSA

Received: 01 January 2025
Accepted: 21 January 2025

***Correspondence:**
Dr. Murtada A. Ahmed,
E-mail: murtada_elimam@hotmail.com

ABSTRACT

Netherton syndrome (NS) is a rare genodermatosis with autosomal recessive inheritance caused by mutations in the serine peptidase inhibitor Kazal type 5 (SPINK5) gene, characterized by the triad of ichthyosiform erythroderma, hair shaft abnormality, and atopic diathesis. A 5-year-old Saudi boy diagnosed with NS is reported. He presented with many food allergies, mild lymphopenia, trichorrhexis invaginata, eczema-like lesions, elevated immunoglobulin E (IgE), and exfoliative erythroderma at birth. Several carious teeth and widespread non-plaque-induced gingival inflammation were among the intraoral findings. Following preoperative multidisciplinary clearance, full dental rehabilitation under general anesthesia was carried out. Prophylactics, composite restorations, stainless steel crowns, and extractions of non-restorable teeth were included as part of the treatment. The patient's recovery from the procedure went smoothly, and both oral comfort and function improved. In conclusion, this case highlights the critical role of multidisciplinary collaboration in the safe delivery of dental rehabilitation under general anesthesia for patients with NS.

Keywords: Netherton syndrome, SPINK5 gene, Pediatric care, Oral rehabilitation, General anesthesia

INTRODUCTION

Netherton syndrome (NS) is a rare autosomal recessive disorder caused by mutations in the serine peptidase inhibitor Kazal type 5 (SPINK5) gene, leading to hair shaft abnormalities like trichorrhexis invaginata, congenital ichthyosiform erythroderma, or ichthyosis linearis circumflexa, and an atopic diathesis that includes allergies and recurrent infections.[1,2]

About 1 in 200,000 live births are affected by NS, which manifests early in life and frequently causes serious morbidity as a result of compromised nutrition, recurrent infections, and dysfunctional skin barriers.[3] Oral findings are not as commonly discussed as the immunologic and dermatologic aspects of NS. According to the limited reports that are currently available, patients may be more susceptible to oral infections, dental caries, and management challenges as a result of systemic fragility.[4-6]

Interdisciplinary care is crucial when general anesthesia is needed for dental rehabilitation.[7] This case report describes the oral manifestations and dental management of a 5-year-old Saudi boy with genetically confirmed NS.

CASE REPORT

A 5-year-old Saudi male was referred to the pediatric dental residency clinic. A comprehensive review of medical records, clinical history, and genetic testing, including whole exome sequencing, was conducted to confirm the diagnosis of NS. Clinical management involved a multidisciplinary approach that included dermatology, allergy/immunology, and anesthesiology.

Intraoral clinical examinations and intraoperative radiographic evaluations were performed to establish the treatment plan. Dental rehabilitation was carried out under

general anesthesia to manage extensive dental decay and systemic fragility.

Clinical history revealed multiple systemic features of NS. The child exhibited trichorrhexis invaginata, a characteristic hair shaft abnormality, and atopic diathesis, characterized by eczema-like lesions on the face and body (Figure 1).

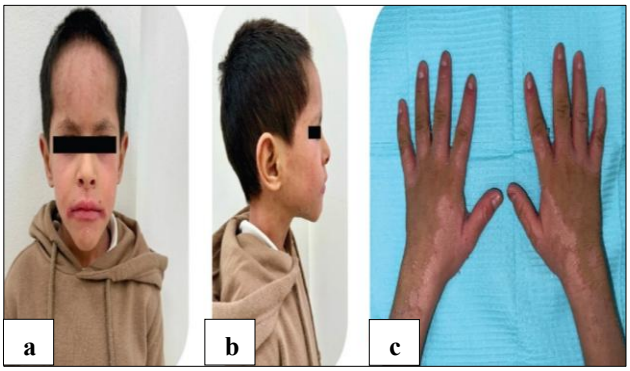

Figure 1: Extra-oral clinical photographs: (a) frontal view showing eczema-like facial lesions, lip and eye swelling, and hair with trichorrhexis invaginata, (b) lateral view highlighting trichorrhexis invaginata, and (c) dorsum of hands showing cutaneous manifestations of exfoliative erythroderma.

Immunological examinations revealed elevated IgE levels, which manifested as multiple food allergies, primarily to tree nuts, peanuts, kiwi, and oats, characterized by lip and eye swelling. Additionally, a banana allergy was identified, which was associated with worsening eczema.

A lymphocyte subset analysis performed in 2017 revealed mild lymphopenia. Based on these features, diagnostic considerations included congenital ichthyosiform erythroderma and peeling skin syndrome. Whole exome sequencing subsequently demonstrated a homozygous variant in the SPINK5 gene, confirming the genetic diagnosis of NS.

Management of NS at the dermatological level included topical therapy with emollients recommended at least twice daily and preferably after bathing. In areas of inflamed or eroded skin, emollients without urea are preferred to reduce inflammation.

Intraoral examination revealed generalized, non-plaque-induced moderate gingival inflammation, characterized by soft, edematous, erythematous gingiva with loss of stippling and bleeding on probing. Hard tissue examination revealed multiple carious teeth requiring restoration to prevent further deterioration (Figure 2).

Due to the patient's age and behavior, radiographs (Figure 3) were not taken at this time and were instead scheduled to be taken in the operating room. Prior to this, the patient was placed on a three-month oral hygiene maintenance follow-up to minimize superimposed gingival inflammation.

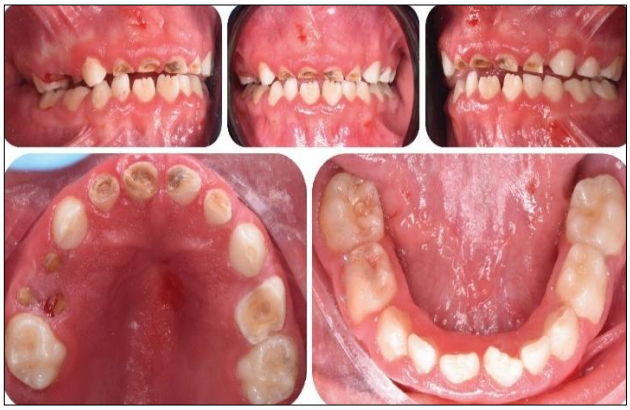

Figure 2: Pre-operative intra-oral photographs: showing carious teeth and moderate gingival inflammation.

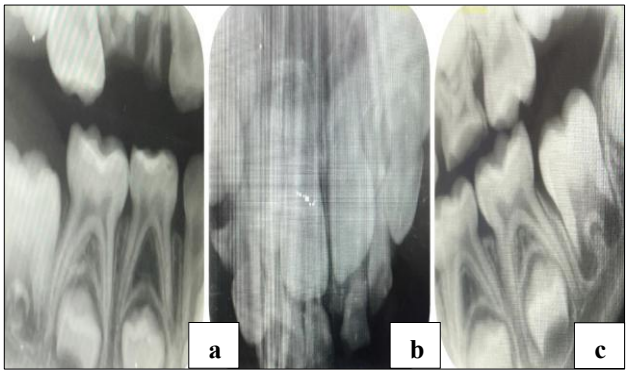

Figure 3 (a-c): Pre-operative intra-oral radiographs: Arrows highlighting areas of dental caries.

Because of the extensive carious lesions, behavioral challenges, and systemic fragility, conventional dental treatment was not feasible. After multidisciplinary clearance from dermatology, allergy/immunology, and anesthesiology, dental management was performed under general anaesthesia.

The procedure included full-mouth prophylaxis, composite restorations for teeth with manageable decay, stainless steel crowns for teeth with multisurface lesions, and extraction of severely decayed non-restorable teeth. Postoperatively, the patient was closely monitored for delayed hypersensitivity reactions, skin hydration was maintained, and analgesics were prescribed for pain management.

The child was discharged the following day after meeting hospital discharge criteria, and follow-up was arranged. A six-month postoperative follow-up demonstrated improved oral health, characterized by the resolution of gingival inflammation (Figure 4).

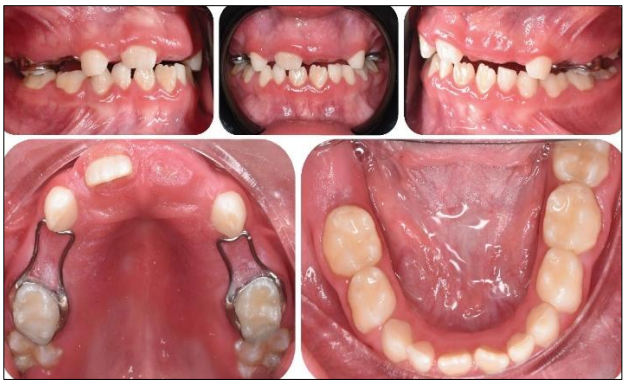

Figure 4: Six months post-operative follow-up: Demonstrating healing of gingival tissues and successful restorations.

DISCUSSION

A rare genetic condition known as NS is caused by mutations in the SPINK5 gene, which impair the function of the lympho-epithelial Kazal-type-related inhibitor, a serine protease inhibitor crucial to the integrity of the skin barrier.

Hair shaft defects such as trichorrhexis invaginata (bamboo hair), congenital ichthyosiform erythroderma or ichthyosis linearis circumflexa, and an atopic diathesis involving elevated IgE, multiple allergies, and recurrent infections constitute the classic clinical trial.

The NS symptoms in this case, including systemic allergic reactions and a genetically confirmed homozygous mutation in SPINK5, correspond with this recognized phenotype. The findings in this case are consistent with previous reports describing the same manifestations, though there may be some differences.

One study reported a case of a 3-year-old boy with a SPINK5 gene mutation who, since the neonatal period, presented with desquamative erythroderma, ichthyosis, hypotrichia, recurrent infections, food-induced anaphylaxis, growth delay, and elevated IgE/IgA levels, leading to a diagnosis of NS.[8] Oral manifestations of NS are less commonly described; however, dental considerations in NS have been discussed.

The present case is consistent with the case report in a similar study, which described a 3-year-old male with NS presenting with generalized moderate plaque accumulation, bulbous erythematous gingiva, and moderate gingivitis, but no carious lesions. Due to the child's age and behavior, radiographs were not taken at that time. Prophylaxis was performed, and the child was scheduled for 3-month oral hygiene maintenance with multidisciplinary follow-up to reduce gingival inflammation.[5]

The present case adds to this body of evidence, as it aligns with reports from other special healthcare needs populations indicating that extensive dental caries often necessitates full-mouth rehabilitation under general anaesthesia.

Research on dental procedures performed under general anaesthesia shows that it provides safe, comprehensive care for children with systemic comorbidities and severe oral disease, reduces the risk of procedural complications, and often improves quality of life and postoperative outcomes. An interesting meta-analysis study found evidence that the oral health-related quality of life of children receiving dental treatment under general anaesthesia improves with a large effect size in the short-term following treatment.[9]

A prospective cohort study reported on dental procedures performed under general anaesthesia in children with early childhood caries, revealing that most procedures – pulpotomies, stainless steel crowns, and restorations – could be successfully completed in a single general anaesthesia session with good outcomes.[10]

Two case reports on the clinical features of NS and the investigation of targeted biologic therapy describe patients with severe eczema, scaling, high IgE levels, and allergic comorbidities, highlighting the negative effects on both skin and overall health.[11] Although these reports do not provide details about dental rehabilitation, they demonstrate the medical fragility of patients with NS and support the view that multidisciplinary approaches are often required for dental interventions.

Dental rehabilitation under general anaesthesia was an appropriate and essential decision in the treatment of the current patient due to systemic sensitivity, the need for extensive dental work in a single session, and the likely reduced cooperation caused by discomfort or skin disease.[12] This approach reduces the risk of infection, minimizes recurrent stress, and can improve feeding and nutritional status, all of which are crucial for children with NS who may experience failure to thrive or digestive difficulties due to multiple food allergies.[13]

The present case is consistent with published reports emphasizing the importance of multidisciplinary care in NS. Dermatologists, immunologists, pediatricians, and dentists must work together to provide individualized and safe treatment plans.

An international multi-stakeholder e-Delphi consensus highlights that NS affects not only the skin but is a disease requiring a comprehensive multidisciplinary approach in clinical care and research.[7] Standard follow-up is necessary to detect problems at an early stage.[14,15] Although long-term dental outcomes were not available, postoperative improvement in oral function and comfort was achieved.

CONCLUSION

This case demonstrates how uncommon dermatological conditions can affect oral health systemically and highlights the importance of interdisciplinary approaches to ensure optimal outcomes. Early dental intervention is necessary to prevent and manage oral complications in children with NS.

To reduce the risk of gingival and dental disease, regular recalls, tailored preventive care, and minimally invasive procedures are essential. Careful planning and close coordination between dental and medical professionals enable safe completion of treatment under general anaesthesia when extensive rehabilitation is required.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Sulaiman NM, Demyati MA, Alomar NE, Ahmed MA. Dental management under general anesthesia in a pediatric patient with Netherton syndrome: a case report. Int J Community Med Public Health 2026;13:965-8.