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Case Report

Management of Odonto-onycho-dermal dysplasia: the role of botulinum toxin

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Abstract

Various methods exist for measuring wound dimensions, including photography, comparison, ruler, or graph techniques1. With the advancement of current-generation smartphones equipped with high-quality built-in cameras, accessibility to such technology has become more widespread and affordable. Leveraging specific applications developed for these smartphones has provided numerous advantages in the medical field. One such software, Imitomeasure, has been designed to measure wound dimensions without direct contact with the wound site. Unlike traditional clinical measurements involving rulers or tape, Imitomeasure offers a non-contact approach, reducing the risk of infection transmission to the patient. The efficacy of Imitomeasure has been evaluated, revealing its effectiveness in wound dimension assessment. In this investigation, the dimensions of bedsores located over bilateral greater trochanteric regions and presacral region of a patient who was bed-ridden due to Parkinson disease were assessed using the Imitomeasure application on a smartphone.

Keywords: imitomeasure application; wound dimension; measurement

Introduction

Odonto-onycho-dermal dysplasia (OODD) is a rare autosomal recessive condition that phenotypically involves the presence of oligodontia, palmoplantar hyperkeratosis as well as hyperhidrosis of the palms and soles. There is limited literature on the management of this condition, the aim of which is often symptom-directed. This case report aims to describe the novel use of botulinum toxin for the treatment of odonto-onycho-dermal dysplasia.

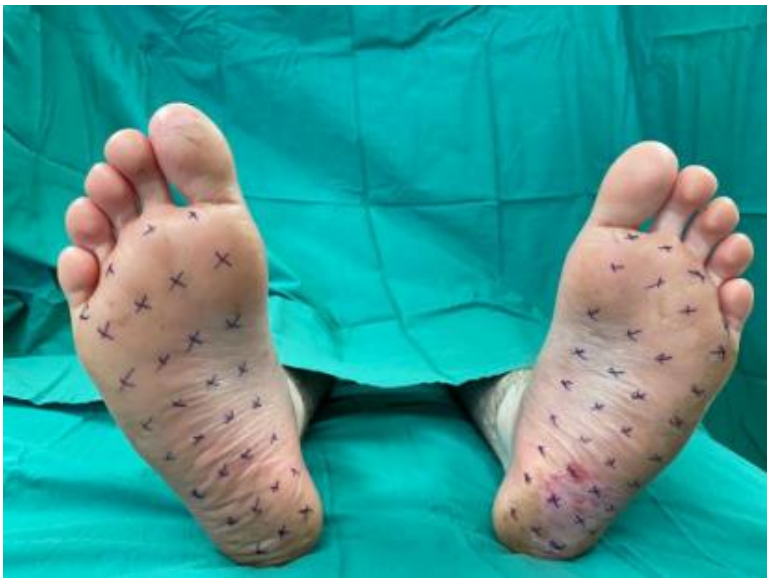
Case Report

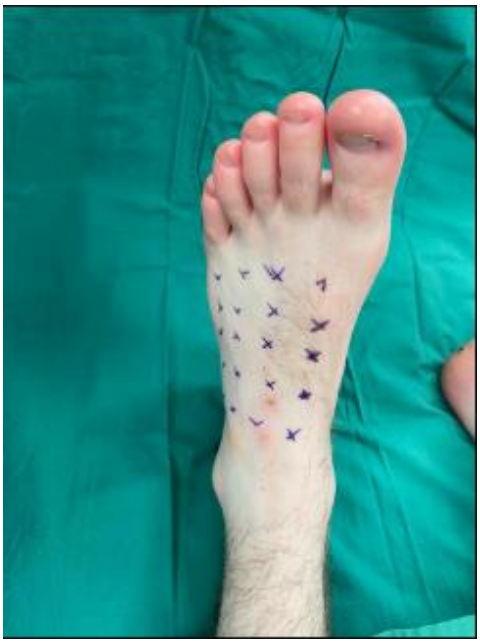
The now early adolescent male patient initially presented to our health service with excessively dry skin, associated palmoplantar keratoderma, heat intolerance and dysregulated sweating. Other associated symptoms of note included conical teeth and hypodontia, as well as the presence of thin and brittle nails. Given the myriad of his symptoms, a genomic DNA test was performed which confirmed the presence of a compound heterozygous WNT10A variant, providing the service with a unifying diagnosis of Odonto-onycho-dermal dysplasia. This is on a background of eczema, autism spectrum disorder, anxiety and depression.

For this patient, the most concerning clinical manifestation of this condition is the severe bilateral plantar foot hyperhidrosis, which results in skin maceration that often progressed to cracking, ulcerations and cellulitis. This desquamation of both feet was often accompanied by severe pain, the treatment of which often required a prolonged hospital admission for intravenous analgesia directed by the acute service pain team. Previous hyperhidrosis treatments that had been trialled by the patient included:

- Iontophoresis with glycopyrrolate solution
- Topical aluminium chloride
- Topical glycopyrrolate
- Corn starch and Talcum powder
- Oral anticholinergic medication

However, the vast majority of these treatments were ceased due to either being ineffective or intolerable adverse side effects by the patient (i.e. pain from iontophoresis, dry mouth from anticholinergic medication, etc.). Regular Botulinum toxin injections administered under a general anaesthetic at regular 4-monthly intervals has been the only intervention that has demonstrated success in achieving remission from his symptoms and the associated pain episodes. These injections involve the use of 150units of Botulinum Toxin injected into both the plantar and dorsal surfaces in the locations outlines in Figure 1 and Figure 2.





While this intervention does not completely treat all the patient's hyperhidrosis associated symptoms, it does alleviate the frequency and intensity of episodes. As such, it is performed in conjunction with regular oral analgesia as directed by the chronic pain services team as well as a prescribed regimen of medicated ointments including corticosteroid ointments, tacrolimus and thicker emollients such as urea cream.

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Discussion

OODD is an extremely rare autosomal recessive form of ectodermal dysplasia. First described in 1983, Fadhil et al. described five individuals displaying the phenotypic characteristics of this disorder from a consanguineous Lebanese family (1). These features have been subsequently delineated in a number of other case series. All patients shared a number of key features including; Altered dentition (often

conical shaped incisors), hyperhidrosis of the palms and soles, onychodysplasia, hyperkeratosis of the palms and soles, hypotrichosis, dry skin (2). Diagnostic confirmation of this condition occurs via genomic DNA testing and identification of a mutation in the WNT10A gene (3).

Treatment strategies for the management of OODD are not well documented in the literature. Symptom based treatment including topical keratolytic agents and corticosteroid creams have been vaguely suggested as appropriate interventions (4). From our institution's experience, early diagnosis as well as a multi-disciplinary approach to the management of OODD is required. Key stakeholders in the patient's management should include:

Acute and chronic pain service team – Management of chronic plantar foot pain as well as flares of acute pain

- Psychiatry – Sensor-modulation strategies to manage altered sensations and distress
- Dermatology – Treatment of dry skin and palmoplantar keratoderma with topical therapy regimes
- Plastic Surgery – Management of hyperhidrosis with regular botulinum toxin injections
- Disability liaison coordinator – Coordination of services and teams required
- General practitioner/family medicine practitioner – The primary prescriber and liaison for the patient while in the community

Botulinum toxin in the management of hyperhidrosis and its sequelae for the OODD patients represents a novel use of this medication. Its use has been well-described in the literature for treating focal hyperhidrosis, often of the axillary region (5, 6). The mechanism action involves inhibition of the release of acetylcholine at the eccrine sweat receptor neuromuscular junction, preventing the known hyperstimulation that leads to excess sweat production. Under a general anaesthetic, our institution's protocol involves injecting 150unit of botulinum toxin to the plantar and dorsal surface of a single foot. A 30-gauge needle is utilised and the plantar surface is injected into the dermis at 31 evenly distributed sites and into the dorsal dermal surface at 20 evenly distributed sites. This is repeated on a 4-monthly basis. In this particular case, is has demonstrated a significant reduction in hospital admission as a result of decrease in the frequency of painful hyperhidrotic 'flare-ups'.

Conclusion

Treatment of OODD is complex and not well-described in the literature. Close involvements between a multi-disciplinary set of teams is essential to optimise outcomes. Additionally, we describe the novel use of botulinum toxin as an adjunct to the long-term management of OODD.

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