A Case of Twenty Nail Dystrophy and Review of Treatment Options

Emily Tongdee, BS,* Shahjahan Shareef, BS,** Tracy Favreau, DO,*** Khasha Toulouei, DO****

*Medical student, Florida International University Herbert Wertheim College of Medicine, Miami, FL
**Medical student, Nova Southeastern University College of Osteopathic Medicine, Ft. Lauderdale, FL
***Dermatologist, Nova Southeastern University College of Osteopathic Medicine, Ft. Lauderdale, FL
****Dermatology resident, 3rd year, Nova Southeastern University College of Osteopathic Medicine, Ft. Lauderdale, FL

Disclosures: None
Correspondence: Shahjahan Shareef; shahjahansen@gmail.com

Abstract

Twenty nail dystrophy (TND), also known as trachyonychia, is an abnormality of the proximal nail matrix. It presents as a homogenous roughness, giving the nail a sandpaper-like appearance.1–3 Idiopathic trachyonychia most commonly presents with spongiotic inflammation with exocytosis of inflammatory cells.1,3–5 This disease has been shown to spontaneously resolve within about five to six years, but that may be too long for some patients, as this nail disorder can be cosmetically disfiguring, further impacting quality of life.6,7 We present a case of idiopathic TND and provide an updated review of the literature and the various treatments that have been utilized to treat the condition. Griseofulvin injections, PUVA, systemic steroids, oral retinoids, cyclosporine A, and nail plate dressings have shown to be highly successful further impacting quality of life.6,7 We present a case of idiopathic TND and provide an updated review of the literature and the various treatments that have been utilized to treat this condition. Griseofulvin injections, PUVA, systemic steroids, oral retinoids, cyclosporine A, and nail plate dressings have shown to be highly successful.

Introduction

Twenty nail dystrophy (TND), also known as trachyonychia, was first mentioned by Hazelrigg et al. in 1977 to describe nail dystrophy occurring in all 20 nails of six children.6 However, since not all 20 nails are always affected in this condition, it has since been termed “trachyonychia.”7 Trachyonychia is a specific type of proximal matrix abnormality that is marked by diffuse homogenous roughness.2 In several case studies, it has been reported as an autosomal-dominant condition of idiopathic origin.6–12 TND has also been described in monozygotic twins. Girls and boys are affected equally, and the condition can occur in adults as well.13–16 Often, the disease presents at birth and evolves slowly, or it may present in infancy or childhood and progress from there.15,17 The peak age range is from 3 to 12 years old.8 It is more commonly seen in males than females when associated with alopecia areata.9

Most commonly, TND presents as nail dystrophy with excessive longitudinal ridging and striations that give the nails a rough or broken appearance. It can involve any number of nails in the upper or lower limbs.1 Additionally, numerous superficial pits on nail surfaces leave the nails with a shiny appearance. In mild cases, alternating elevations along the nail surface, longitudinal ridging and/or pitting are almost always observed.15 In severe cases, TND may present with a sandpaper-like, opaque appearance. Severity of disease may vary between nails, but generally, the nails evolve over time into a muddy, white-grayish discoloration.15,18

Case Report

A 34-year-old Filipino female presented to the clinic with nail disease in all 20 of her nails, starting with the thumb and progressing one by one beginning about two years earlier. She had seen previous dermatologists who said fungal cultures were negative and was empirically treated with oral antifungal medications for months with no improvement. She denied any significant past medical history, surgical history, and hospitalizations. Her family history was significant for type 1 diabetes mellitus and hypertension in her father and mother, respectively. She denied any use of alcohol, tobacco or illicit drugs and said she was not taking any prescription medications, herbal supplements, or vitamins. She alsodenied any allergies or recent travel.

Physical exam revealed thickened dystrophic nails with pitting, longitudinal ridging, and onycholysis (Figure 1). The patient was diagnosed with idiopathic twenty nail dystrophy, as all twenty nails were involved and no associations were seen. We presented the patient with many different treatment options, including intralesional steroid injections. Our patient elected to take 20 mg of biotin daily. She was also given reassurance.

Discussion

The etiology of TND is controversial. It can either present as its own entity or as a manifestation of another condition, including lichen planus (4% to 18.5% of cases), alopecia areata (45% to 83% of cases), or psoriasis (13% to 26% of cases).1,3–10,12 TND has also been associated with incontinentia pigmenti, vitiligo, ichthyosis vulgaris, ichthyosis, immunoglobulin IgA deficiency, hemolytic abnormalities, koilonychias, eczema, primary biliary cirrhosis, alopecia universalis and sarcoidosis.6,8,11–13,15,19–21 Alopecia areata is the most common disease associated with trachyonychia.2,22

Trachyonychia has a pathognomonic presentation of thin, brittle nails with longitudinal ridging and striations that give the nails a rough or broken appearance. It can involve any number of nails in the upper or lower limbs.1 Additionally, numerous superficial pits on nail surfaces leave the nails with a shiny appearance. In mild cases, alternating elevations along the nail surface, longitudinal ridging and/or pitting are almost always observed.15 In severe cases, TND may present with a sandpaper-like, opaque appearance. Severity of disease may vary between nails, but generally, the nails evolve over time into a muddy, white-grayish discoloration.15,18

Figure 1
multiple small puncate depressions that are spread in a geometric pattern within parallel lines.2
Since the disease affects the nail matrix, a nail matrix punch biopsy or longitudinal nail biopsy may be performed, but this is not recommended; the risk/benefit ratio is not advantageous, as the procedure is invasive and the condition can be diagnosed clinically.18 In idiopathic TND and TND associated with alopecia areata, histology of the nail will most commonly demonstrate spongiform inflammation of the nail matrix and exocytosis of inflammatory cells (lymphocytes) into the nail epithelia.1,3-5
When the disease is caused by psoriasis or lichen planus, histology will display findings typically associated with both of these conditions. Histology consistent with nail lichen planus will reveal hyperkeratosis, hypergranulosis, and saw-tooth acanthosis containing a band of lymphocytic infiltrate with vacuolar degeneration of basal keratinocytes.15 Histology consistent with psoriasis will reveal acanthosis and focal parakeratosis affecting the proximal nail fold and nail matrix.16 Also, polymorphonuclear leukocytes along the whole length of the dorsal nail plate are commonly observed in TND histologically consistent with psoriasis.14 Focal areas of nail hypergranulosis can be seen in idiopathic trachyonychia, nail lichen planus and nail psoriasis. Inflammation in the matrix can affect keratinization, causing an increase of keratohyalin granules.5 Here, histology may reveal a mild to moderate lymphocytic infiltrate in the superficial dermis of the proximal nail fold and matrix.15 The ventral portion of the nail fold sometimes demonstrates hyperkeratosis of the cuticle. Other possible findings include longitudinal clefts containing zones of eosinophilic onychocytes and parakeratotic cells that may reside in the nail plates. The proximal nail matrix and the ventral proximal nail fold are typically affected more than the distal matrix. This displays clinically in the dorsal nail plate. Furthermore, nail plate abnormalities are more present in the dorsal layer.18
Clinically, trachyonychia secondary to lichen planus or psoriasis appears identical to trachyonychia due to spongiform features.1 However, due to the distinctive presentation of this nail plate abnormality and the risk of permanent nail damage from biopsy, diagnosis will usually be made clinically. Trachyonychia can often be misdiagnosed as onychomycosis, so laboratory studies are recommended before initiating antifungal therapy for the treatment of onychomycosis.4 Due to the many dermatological associations, a thorough skin examination looking

Table 1. Treatments used for TND

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Route</th>
<th>Dose</th>
<th>Time</th>
<th>Note</th>
</tr>
</thead>
<tbody>
<tr>
<td>Steroids (general)</td>
<td>Topical</td>
<td>1% ointment</td>
<td>4 months</td>
<td>Relapse, painful, proximal nailfold, need long-term compliance (effective in 4 children)</td>
</tr>
<tr>
<td>Triamcinolone</td>
<td>Intramuscular</td>
<td>0.5 mg/kg - 1 mg/kg</td>
<td>Bimonthly for 4 months</td>
<td>Proximal and lateral nailfolds</td>
</tr>
<tr>
<td>Prednisone</td>
<td>PO</td>
<td>0.5 mg/kg</td>
<td>Alternate days for 4 weeks</td>
<td>No relapse</td>
</tr>
<tr>
<td>Triamcinolone acetonide</td>
<td>IM</td>
<td>(10 mg/ml)^3</td>
<td>2 times per week for 8 months</td>
<td>Shown to be effective. Usually fewer side effects vs. the daily dose of corticosteroids over weeks and months.</td>
</tr>
<tr>
<td>Betamethasone</td>
<td>PO</td>
<td>4 mg</td>
<td>Mini pulse therapy (2 consecutive days every week for 2 months)</td>
<td>Required 2 courses, with side effects of peeling, erythema on proximal nailfold (showed improvement in 1 patient with alopecia areata)</td>
</tr>
<tr>
<td>Tazeticone</td>
<td>Topical</td>
<td>0.10%</td>
<td>Nightly for 3 months</td>
<td>Psoriatic trachyonychia (improvement in roughness, ridging, pitting, subungual hyperkeratosis)</td>
</tr>
<tr>
<td>Acitretin</td>
<td>PO</td>
<td>0.3 mg/kg</td>
<td>Daily dose for 3 months</td>
<td>Idiopathic trachyonychia (successful in 5 patients).</td>
</tr>
<tr>
<td>Cyclosporine A</td>
<td>PO</td>
<td>3 mg/kg/day^2^-3.5 mg/kg/day^2</td>
<td>Daily for 2.5 months</td>
<td>Psoriatic trachyonychia, general anesthesia used.</td>
</tr>
<tr>
<td>PUVA</td>
<td>n/a</td>
<td>0.7 J/cm²^2 - 1.4 J/cm²^2</td>
<td>3 times a week for 7 months</td>
<td>All treated nails showed significant improvement, untreated remained dystrophic.</td>
</tr>
<tr>
<td>5-fluorouracil</td>
<td>Topical</td>
<td>5%</td>
<td>Every 2-4 days for 16 weeks</td>
<td>Psoriatic trachyonychia; periungual irritation limits the drug’s use.</td>
</tr>
<tr>
<td>Griseofulvin/steroid</td>
<td>PO/intramatrix</td>
<td>10 mg/kg</td>
<td>6 months</td>
<td>Extensive treatment; LP trachyonychia, general anesthesia used.</td>
</tr>
<tr>
<td>Biotin</td>
<td>PO</td>
<td>20 mg</td>
<td>Daily</td>
<td>Primary biliary cirrhosis patient.</td>
</tr>
<tr>
<td>Petrolatum</td>
<td>Topical</td>
<td>Not known</td>
<td>Not known</td>
<td>Partial resolution seen.</td>
</tr>
<tr>
<td>Nail plate dressings</td>
<td>Topical</td>
<td>Once a week</td>
<td>6 months</td>
<td>Significant improvement at 3 months; near complete resolution at 6 months</td>
</tr>
<tr>
<td>Vitamin supplement</td>
<td>PO</td>
<td>Not known</td>
<td>Not known</td>
<td>Partial resolution seen.</td>
</tr>
</tbody>
</table>
for psoriatic features, dermatosis, and/or hair loss should be performed.

Treatment of trachyonychia is aimed primarily at the underlying cause, if found. TND usually remits spontaneously over several years, as it is a self-limited condition. Therefore, counseling and reassurance for parents and patients is important, as is explaining to them that the disease is self-resolving. In about 50% of patients, nail changes will reveal significant improvement or resolve within five to six years. Waiting this long can be hard for some patients, as this condition can negatively affect quality of life. Currently, no specific treatment exists for the condition, and treatment is done only for cosmetic reasons. The disease does not leave any scars.

The basis of treatment involves regulating the differentiation of keratinocytes and/or decreasing the inflammation in the nail fold or nail matrix. Treating TND associated with psoriasis or lichen planus with systemic therapies may involve corticosteroids, oral retinoids, or cyclosporine (Table 1, p. 44).

There have been several reports of treatment success. One article reported success with griseofulvin in an intra-matrix steroid injection in a 6-year-old boy diagnosed with lichen planus-related TND under general anesthesia over a period of six months. This method has been quite successful for TND with associated lichen planus. Relapse has been seen with intraligamental injection of corticosteroid, and compliance is reduced due to pain. Tosti et al. treated a lichen planus trachyonychia patient with oral prednisone 0.5 mg/kg on alternate days for four weeks. Four months after stopping the treatment, the nails demonstrated only mild longitudinal ridging, with no relapse over three years. They also attempted intramuscular triamcinolone acetonide 0.5 mg/kg to 1 mg/kg per month in 15 children with typical nail lichen planus with successful results. Silverman et al. treated a 9-year-old girl with lichen planus-related TND using potent steroids for six months, which did not result in any improvement.

In psoriatic TND, recurrence of nail changes is usually seen when treatment is stopped. Still, a study using an oral retinoid (acitretin) 0.3 mg/kg daily for three months showed improvement in roughness, ridging, pitting and subungual hyperkeratosis. Topical 0.10% retinoid (tazarotene) did not achieve the same effect, but resulted in peeling and erythema on the proximal nail folds; however, mild improvement was seen in one patient with alopecia areata. Two studies also showed success with oral cyclosporine A at 3 mg/kg/day: Pierard et al. administered it daily for two and half months with success in five patients with psoriatic trachyonychia, and a case series of 15 patients with idiopathic trachyonychia saw an 87% improvement after six months of therapy. Good results were seen with PUVA three times a week for seven months at strengths of 0.7 J/cm2 to 1.4 J/cm2. Topical 5-fluorouracil, on the other hand, was not successful and resulted in periungual irritation that limited its use. Arias-Santiago et al. used nail plate dressings that consisted of an ultra-thin adhesive layer containing lactic acid, silicon dioxide, aluminium acrylate, vinyl copolymer, and azelac acid that were applied once a week for six months. This particular method showed significant improvement at three months and close to complete resolution at six months. Mittal et al. took the unique approach of administering PO 4 mg betamethasone two consecutive days every week for two months. This mini pulse therapy was shown to be effective, with fewer side effects when compared to daily oral steroids for weeks or months. Petrolatum and vitamin supplementation showed only partial resolution. Biotin was used successfully to treat TND in two patients with primary biliary cirrhosis.

Conclusion

TND, also known as trachyonychia, is a disease that can present independently, idiopathically, or in association with other conditions. Nail matrix biopsies are usually not recommended, as they risk leaving the patient with permanent nail damage and the diagnosis can usually be made on a clinical basis. To date, no particular treatment has been universally accepted. Positive results have been achieved with griseofulvin injections, PUVA, systemic steroids, oral retinoids, cyclosporine A, and nail plate dressings. But in a large majority of cases, trachyonychia is a self-limiting disease and, as a result, treatment should only be given if absolutely essential, such as when the patient’s quality of life is detrimentally affected. In this population, there are treatment options that can significantly shorten the duration of the disease. Patients should be aware of the treatment choices and the associated risks. Patients should also be reassured that abstaining from therapy is also an acceptable and safe option due to the benign nature of the disease.
References