

Case Report on Familial Trachyonychia

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Abstract

Trachyonychia may be idiopathic, familial, or occur in association with other dermatologic conditions. It manifests in two distinct forms: opaque (more severe) and shiny trachyonychia, or as a combination of both. The condition typically affects children, with peak prevalence between ages 3 and 12, but it may occur at any age and in both sexes. It can be inherited in an autosomal dominant pattern and has been documented in monozygotic twins. While frequently idiopathic, trachyonychia may be associated with alopecia areata, eczema, lichen planus, psoriasis, or vitiligo. Diagnosis can be challenging when no other clinical features are present. A thorough dermatologic evaluation is essential to identify treatable underlying conditions. There is no universally accepted treatment, and the disease is often self-limiting, with reassurance being a critical part of management.

Keywords: Trachyonychia, Twenty-nail Dystrophy (TND), Nail Dystrophy, Rough Nails, Longitudinal Ridging, Nail Matrix Disorder

Interdiction

Twenty-nail dystrophy, also known as trachyonychia, is a proximal nail matrix disorder characterized by diffuse, homogeneous roughness of the nails (resembling sandpaper), thin, brittle nails and longitudinal ridging. Less commonly, it may present as opalescent nails with pitting, affecting all 20 nails [1, 2]. First described by Alkiewicz in 1950, the condition was later named "Twenty Nail Dystrophy (TND) of Childhood" by Hazelrigg et al. in 1977 [3].

Trachyonychia can be idiopathic, familial, or present in association with other dermatologic conditions. The disorder may manifest in two distinct forms: opaque (the more severe presentation) shiny trachyonychia, or occasionally as a combination of both [4, 5]. This condition predominantly affects children, with an insidious onset and peak prevalence between the ages of 3 and 12. However, it can also occur at any age and affect both males and females [6, 7]. Trachyonychia is reported to be transmitted in an autosomal dominant fashion in some families and there are reports of monozygotic twins affected by TND [8]. However, these likely represent an association between TND and alopecia areata, which may occur in twins and several members of a family [9-11] Although TND is often idiopathic,

a few case reports have shown TND and skin diseases such as alopecia areata, eczema, psoriasis, lichen planus, vitiligo, etc. Determining the cause of trachyonychia when other clinical features are not present can be challenging [12, 13]. A thorough systemic dermatological examination is required to identify possible associated treatable diseases. There are no universally accepted treatment options. The condition is shown to be self-limiting over years, even in the presence of associated disease therefore, reassurance of the patient is essential [14].

Case Report

A 32-year-old female civil servant was incidentally discovered in a control group of a study, having dystrophy of all twenty nails for the past 20 years. Her siblings had similar symptoms. She denied any form of rash on her skin or mucosal surfaces, no hair loss, nor allergies. She is not known for any chronic medical condition. Dermatologic examination revealed all twenty nails as dystrophic, opaque, lusterless, brittle, longitudinal ridging and rough surface (figure 1 and 2). Other examination findings were unremarkable. Based on the clinical presentation and familial pattern, a diagnosis of familial twenty-nail dystrophy was made. Fungal studies were negative. She declined further evaluation and any form of therapy as she had lived with it for 20 years without any concern.



Figure 1



Figure 2



Figure 3

Discussion

Twenty-nail dystrophy (TND), also known as trachyonychia is a proximal nail matrix disorder that commonly affects all 20 nails. It presents as an idiopathic, familial or in association with other dermatologic conditions such as alopecia areata, eczema, lichen planus, psoriasis, vitiligo, etc. TND was first described in 1950 by Alkiewicz and termed TND of childhood in 1977 by Hazelrigg et al [15]. Trachyonychia has a pathognomonic presentation of thin brittle nails with longitudinal ridging, and superficial striations seen in a regular, parallel pattern, giving it a sandpaper-like appearance. Other possible findings include elevation/pitting, and splitting.

Less commonly, pitting and a shiny colour. achyonychia is reported to be transmitted in an autosomal dominant fashion in some families and there are reports of monozygotic twins affected by TND. However, these likely represent an association

between TND and alopecia areata which may occur in twins and several members of a family [16, 17]. TND has been described occasionally in adults but commonly affects children aged 3 to 12 years. The condition has a slow progression with an equal predilection for males and females.

The strong association of TND with dermatologic conditions that have an autoimmune aetiology has raised the suspicion that the nail changes could be immunologically mediated. Tosti et al, 40 of 1,095 with alopecia areata had trachyonychia. They found trachyonychia in 3% of adults. They noted nail changes may precede or follow the onset of alopecia areata or the two may arise simultaneously [7, 18]. Alopecia areata is one common abnormality associated with trachyonychia [11, 19]. Trachyonychia is seen in 10% of patients affected by nail lichen planus. Oral lichen planus is the most common type of lichen planus associated with trachyonychia.

Table 1: Causes of Trachyonychia

Dermatologic	Non-dermatologic
Alopecia areata	Autoimmune hemolytic anaemia
Vitiligo	Idiopathic thrombocytopenic purpura
Eczema	Trauma
Psoriasis	Immunoglobulin A deficiency
Lichen planus	
Ichthyosis vulgaris	
Graft versus host disease	
Darrier's	
Pemphigus vulgaris	
Amyloidosis	
sarcoidosis	

Clinical Features

Rough nails with excessive longitudinal ridging are typically seen. Nail plates may be thickened or thin, cuticles are usually thickened and ragged [1, 20]. Baran in 1981 categorized two appearances; opaque or shiny trachyonychia. Opaque trachyonychia is more severe, with rough nails appearing rubbed with sandpaper. Shiny variant is opalescent nail with numerous pits [4, 5, 21].

Diagnosis

Diagnosis is made both with clinical features and nail matrix punch or longitudinal nail biopsy for pathologic diagnosis. However, a pathologic diagnosis is not required as the disease has a benign outcome even when cause by lichen planus [1, 13, 14].

The most common histopathologic features with trachyonychia are spongiosis and exocytosis of the inflammatory cells into the nail epithelia. Hypergranulosis can be seen in idiopathic, lichen planus and psoriasis-associated TND [1, 14, 17].

The risk/benefit ratio of performing a nail biopsy to identify the

pathologic cause of trachyonychia dictates that a nail matrix biopsy should not be a part of standard procedure [14, 22, 23].

Differentials.

- Alopecia areata: Often difficult to make a distinction between geometric superficial pitting similar to trachyonychia
- Brittle nails: Longitudinal ridging and superficial pitting, no roughness.
- Lichen planus: Longitudinal fissures and pterygium.
- Psoriasis: Pitting, oil spot, nailbed discolouration, onycholysis, subungual hyperkeratosis and splinter haemorrhage
- Senile nails: Mild longitudinal ridging that does not usually involve the entire nail plate as in trachyonychia

Treatment

There is no universally accepted treatment regimen for trachyonychia. Hazelrigg et al stated that trachyonychia is self-limiting and self-resolving in children hence reassurance of children and parents is crucial [14, 15]. Treatment is usually for cosmetic reasons and patients may often improve without treatment. While TND is not harmful, nail disease has been

shown to hurt patient quality of life. For patients with underlying disease, treatment of the associated disease may improve the appearance of the nails.

Treatment options include: 1. Observation/active non-intervention, 2. Emollient and camouflage nail polish, 3. Topical corticosteroids, tazarotene gel, 5 fluorouracil, 4. Nail plate

dressing (ultra-thin adhesive bandage applied once a week with lactic acid, silicon dioxide, aluminium acetylacetone, copolymers of vinyl acetate with acrylic acid, and azelaic acid, 5. Nail unit steroid injections of triamcinolone into proximal nail fold, 6. Topical psoralen UVA, 7. Systemic agents (Biotin, Cyclosporin, Retinoid and Corticosteroids).

Table 2. Treatment Options

Treatment	Route	Dose	Time	Note
Steroids (general)	Topical	1% ointment	4 months	
Triamcinolone	Intralesional	0.5 mg/kg - 1 mg/kg	Bimonthly for 4 months	Relapse, painful, proximal nail fold, needs long-term compliance (effective in 4 children) ¹⁴
Prednisone	PO	0.5 mg/kg	Alternate days for 4 weeks	No relapse ¹³
Triamcinolone acetonide	I'M	(10mg/ml) ³	2 times per week for 8 months	Proximal and lateral nail folds ²³
Betamethasone	PO	4 mg	Mini pulse therapy (2 consecutive days every week for 2 months)	Shown to be effective. Fewer side effects vs. the daily dose of corticosteroids over weeks and months. ²⁴
Tazoretene	Topical	0.10%	Nightly for 3 months	Required 2 courses, with side effects of peeling, and erythema on proximal nail fold (showed improvement in 1 patient with alopecia areata) ²³
Acitretin	PO	0.3 mg/kg	Daily dose for 3 months	Psoriatic trachyonychia (improvement in roughness, ridging, pitting, subungual hyperkeratosis) ¹⁴
Cyclosporine A	PO	3mg/kg/day	Daily for 2.5 months	Psoriatic trachyonychia (successful in 5 patients)
Cyclosporine A	PO	2 mg/kg/day -3.5 mg/kg/day ⁶⁵	6 months	Idiopathic trachyonychia, in case series of 15 patients 87% showed significant improvement after 6 months of therapy. ¹⁹
PUVA	Topical	0.7 J/cm ² - 1.4 J/cm ²	3 times a week for 7 months	All treated nails showed significant improvement, untreated remained dystrophic. ⁵
5-fluorouracil	Topical	5%	Every 2-4 days for 16 weeks	Psoriatic trachyonychia; periungual irritation limits the drug's use. ¹³
Griseofulvin/steroid	PO/intra-matrix	10 mg/kg	6 months	LP, trachyonychia, general anesthesia used ⁵
Biotin	PO	20 mg	Daily	Primary biliary cirrhosis patient. ²⁵
Petrolatum	Topical	Not known	Not known	Partial resolution seen
Nail plate dressings (ultra-thin adhesive layer with lactic acid, silicon dioxide, aluminium acetylacetone, vinyl copolymer, and azelaic acid)	Topical	Once a week	6 months	Significant improvement at 3 months; near complete resolution at 6 months ^{9,12,23}
Vitamin supplement	PO	Not known	Not known	Partial resolution seen

Conclusion

Twenty-nail dystrophy, or trachyonychia, is a disorder of the proximal nail matrix affecting all twenty nails. The aetiology may be idiopathic, familial, or associated with other dermatologic conditions. The hallmark features include diffuse, homogeneous roughness (resembling sandpaper), thin, brittle nails with longitudinal ridging (opaque), or, less frequently, opalescent nails with pitting (shiny), involving all nails. Diagnosis is primarily clinical and based on characteristic findings [24, 25].

The condition is typically self-limiting in children. While treatment is generally sought for cosmetic reasons, there is no universally accepted therapeutic approach unless an underlying dermatologic condition is identified.

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