Trachyonychia

5

Mohamed EL-Komy

Introduction

In 1977, *Hazelrigg* et al. [1] described six children with an acquired nail dystrophy characterized by excessive longitudinal striations and loss of nail luster. They termed the condition twenty-nail dystrophy (TND) and suggested it to be an idiopathic self-limited abnormality that resolves slowly with age [1]. Earlier, *Alkiewicz* in 1950 used the term "trachyonchie" to describe roughness and grayish opacity of nails that consequently develop brittleness with terminal splitting [2]. The two terms TND and trachyonychia had been used interchangeably by dermatologists for decades. Several authors advocate that the term TND has no specific significance or specificity and should be abandoned [3–5]. Trachyonychia meaning rough nails seems to be a better descriptive term for the clinical picture of the affected nails [6, 7]. Confusingly, some authors consider that TND may spare one or more nails [8]. Essentially, TND is a mere trachyonychia of the 20 nails.

Epidemiology

Trachyonychia is an insidious disease that most commonly affects children, although no age is exempt [4, 5, 9]. Available data suggests that trachyonychia is either idiopathic, especially in children [5], or a manifestation/association with other cutaneous and non-cutaneous systemic diseases [4, 5]. In children, it is very common for trachyonychia to affect the 20 nails [5], thus the name "TND of childhood."

Baran and *Dupre* [10] observed that several cases of trachyonychia are in fact an expression of alopecia areata (AA). Indeed, later on, *Tosti* et al. [11] reported that the prevalence of trachyonychia among patients with alopecia areata is 3.65%. Other dermatosis that has also been reported with trachyonychia includes eczema, lichen planus, psoriasis, Sezary syndrome, and vitiligo [12–14]. There are also several reports of trachyonychia in association with systemic diseases like sarcoidosis, thyroid disorders, IgA deficiency, amyloidosis, and hemolytic abnormalities (Table 5.1) [4, 12, 14, 15]. However, *Jacobson* and *Tosti* [5] pointed out that several reports regarding trachyonychia and disease associations did not actually correspond clinically to the classic nail changes of trachyonychia.

M. EL-Komy (✉)
Department of Dermatology, Kasr AL-Aini, Faculty of Medicine, Cairo University, Cairo, Egypt
e-mail: m_elkomy@kasralainy.edu.eg

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Table 5.1 Associations reported with trachyonychia

Associations
Dermatologic diseases
Reported more than once
Alopecia areata [[9–11]
Eczema [9, 16]
Lichen planus [7, 17]
Psoriasis [18, 19]
Pemphigus vulgaris [20, 21]
Vitiligo [22, 23]
Reported once
Darier's disease [24]
Ichthyosis vulgaris [25]
Incontinentia pigmenti [26]
Systemic disorders
Reported more than once
Hematologic abnormalities [9, 27]
Reported once
Allergic rhinitis and bronchial asthma [8]
APECED syndrome [28]
Behcet's disease [9]
IPEX syndrome [29]
Pityriasis rubra pilaris [9]
Reflex sympathetic dystrophy [30]
Selective IgA deficiency [31]
Sarcoidosis [15]
Sezary syndrome [13]
Thyroid disease [9]

Clinical Picture

Trachyonychia was originally described as excess ridging of the nails [6]. Any number of nails can be affected from a single nail to all 20 nails with fingernails being more frequently affected than toenails [32]. The affected nail(s) may show closely spaced longitudinal striations (onychorrhexis) and a dull, rough nail surface, and numerous, small superficial pits may be evident [12, 33]. Two different subtypes of trachyonychia are described by Baran et al. [3, 10], namely, [1] *the opaque trachyonychia* or vertical striated sandpaper 20-nail dystrophy, which is more frequently seen, more severe, and associated with thickened nails, prominent ridging, surface scaling, and sandpaper appearance (Figs. 5.1 and 5.2) [2], and *the shiny trachyonychia* where the nails are uniform, opalescent, and shiny with several reflective shallow pits (Fig. 5.3). Both subtypes may occasionally be seen in the same patient [32]. Koilonychia,

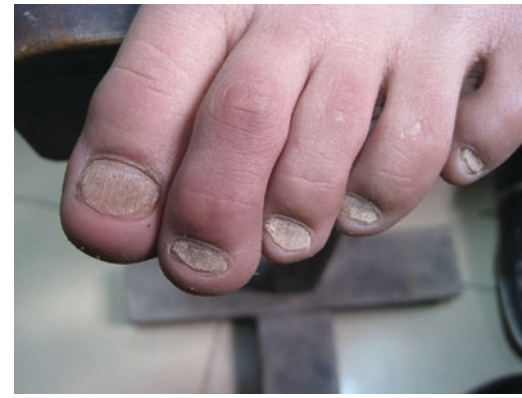

Fig. 5.1 Opaque trachyonychia with sandpaper-like nail plate and koilonychia

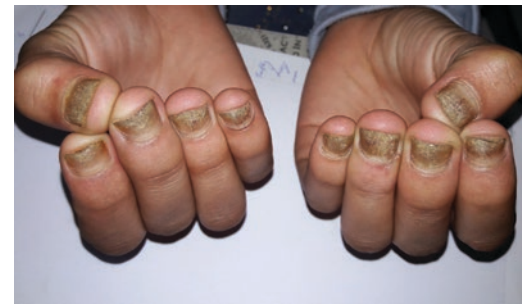

Fig. 5.2 Opaque trachyonychia with nail plate thickening

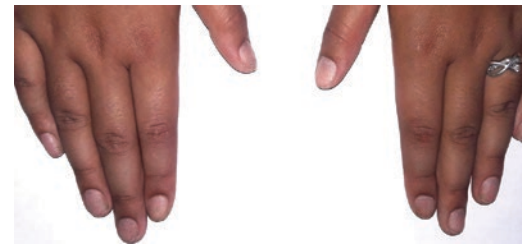

Fig. 5.3 Shiny trachyonychia with numerous shallow pits

nail thinning, fragility, and cuticle hyperkeratosis have also been described with TND/trachyonychia (Figs. 5.4, 5.5, and 5.6) [14, 31]. The nail affection is symptomless [32] although some patients may complain of difficulty in performing fine work especially with the opaque subtype.

When trachyonychia/TND is suspected clinically, it is mandatory for the dermatologist to

Fig. 5.4 Shiny trachyonychia with cuticle hyperkeratosis, spooning, and nail thickening

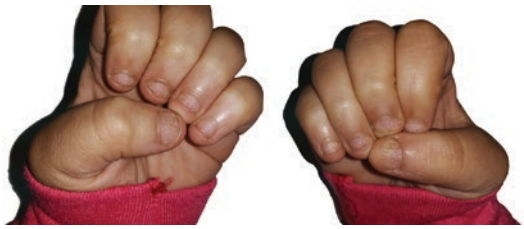

Fig. 5.5 Trachyonychia with nail thinning and distal fragility

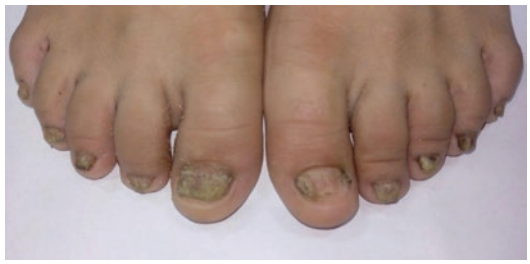

Fig. 5.6 Trachyonychia of toes in teenager football player pronounced over dominant foot

examine for known associations especially AA, psoriasis, and lichen planus (Fig. 5.7). Nevertheless, trachyonychia may precede the classic cutaneous manifestations of these dermatoses. Trachyonychia had been classified clinically into two groups depending on the absence (idiopathic trachyonychia) or presence of associated dermatologic diseases [7].

Etiology and Pathogenesis

As discussed earlier, trachyonychia is believed to be either a manifestation of other dermatologic and systemic diseases or an isolated or idiopathic disorder [12].

Familial cases of trachyonychia/TND had been reported with an autosomal dominant pattern [34]. Balci et al. reported a genetic locus in the form of a balanced translocation XX, t(6q13;10p13) in a mother and daughter with trachyonychia [35]; however, both of their patients also showed anonychia and hypoplasia in several nails. *Gordon* et al. argue that familial cases are likely a representation of the association between trachyonychia and alopecia areata as the latter may occur in a familial manner [12]; moreover, nail changes may follow or precede the onset of alopecia areata by months or years [11].

Trachyonychia is considered a nail aberration which may be caused by several inflammatory diseases that disturb the nail matrix

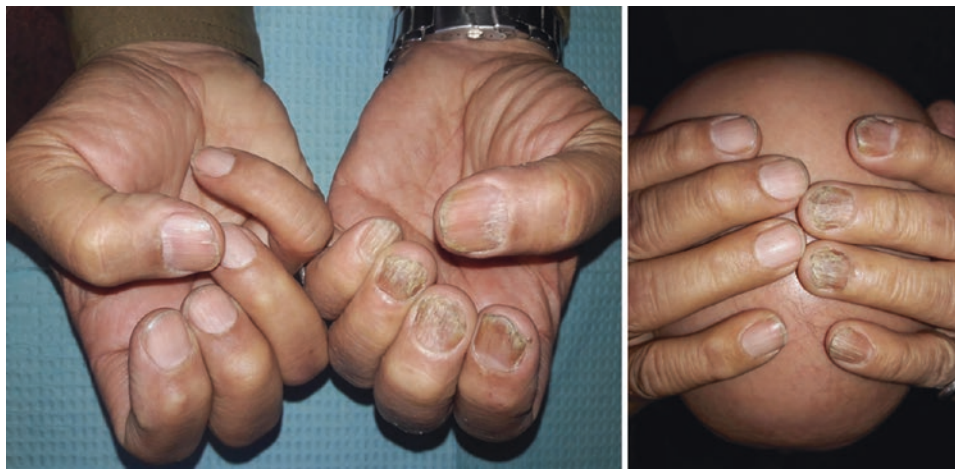

Fig. 5.7 Alopecia totalis and trachyonychia in a middle-aged male

keratinization, but not its mitotic activity [17, 32]. Regardless of the cause, nail matrix onychocytes instead of maturing to form a compact layer of tightly adherent flat cells produce a stratum corneum-like layer that easily desquamates [32]. The course and extent of the matrix inflammatory process and consequently its keratinization are possibly responsible for the different clinical presentations of trachyonychia [7]. A remittent, waxing, and waning inflammatory insult to the nail matrix that never ceases may be responsible for the opaque type of trachyonychia, while an intermittent, focal, and regularly recurrent inflammatory insult to the matrix that is separated by periods of normal matrix function results in the shiny type [36].

Based on the predominant histopathological findings, namely, spongiosis and lymphocytic infiltration, in the majority of patients with idiopathic trachyonychia, it is suggested that this entity may represent a subgroup of endogenous eczema/dermatitis or an autoimmune response confined to the nail matrix [14]. Nonetheless, trachyonychia associated with alopecia areata is suggestive of abnormal keratinization of the proximal matrix with spongiotic changes and lymphocytic infiltration comparable to the idiopathic type of trachyonychia [11, 37].

Pathology

The histopathological changes observed in trachyonychia are usually most severe in the proximal nail matrix in comparison to distal matrix and nail bed [11, 14, 32, 36].

In idiopathic trachyonychia, the majority of patients as reported by Tosti et al. revealed spongiotic changes in the nail apparatus with mild-to-moderate lymphocytic infiltrate in the superficial dermis of the proximal nail fold and nail matrix [7]. The degree of spongiosis ranges from mild to severe changes with intraepidermal microvesicle formation [8]. Dorsal matrix focal hypergranulosis with abnormal eosinophilic appearance of overlying nail plate was also observed.

The nail plates show longitudinal clefts (clinically seen as longitudinal ridging), zones of eosinophilic onychocytes, and zones of parakeratosis (which desquamate and, hence, the ragged appearance observed clinically) [7].

Peculiarly, few patients with idiopathic trachyonychia may show histopathological features of lichen planus or psoriasis without evidence of cutaneous or mucosal affection. Even more bizarre, histopathological features of psoriasis or lichen planus may occasionally be seen in cases of trachyonychia associated with alopecia areata and vitiligo [7, 22, 32, 33].

In trachyonychia with other dermatologic disease, the histopathology of the nail apparatus may coincide with the associated cutaneous dermatosis. Nail matrix lichen planus will reveal an extensive hyperkeratosis, hypergranulosis, lichenoid interface lymphohistiocytic infiltrate, and vacuolar degeneration of basal keratinocytes. Psoriasis would usually involve the proximal nail fold and the nail matrix with hypergranulosis, acanthosis, and focal parakeratosis [14].

As trachyonychia almost never results in permanent nail damage, biopsy from the nail apparatus has been considered unnecessary except in severe, recalcitrant, or uncertain cases [4, 5]. *Haber* et al. suggested implementation of a systemic therapy when histopathological evidence of psoriasis or lichen planus is evident as a primary cause of the trachyonychia [4].

Differential Diagnosis

The unique picture and settings of trachyonychia are usually enough to make the correct clinical diagnosis; however, it is useful to recognize the difference between trachyonychia and the conditions listed in Fig. 5.8 [4, 5, 38–42].

Treatment

A wide range of treatments had been suggested for trachyonychia ranging from topical and systemic medications to ultraviolet therapy and nail

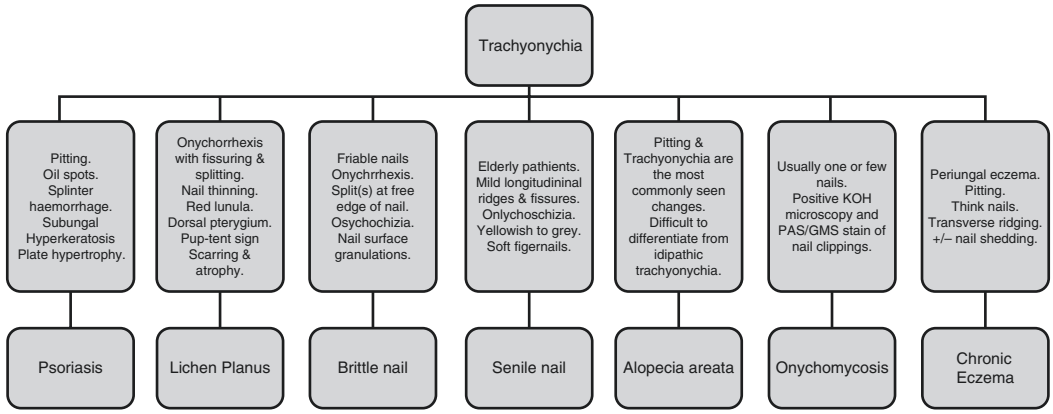

Fig. 5.8 Differential diagnosis of trachyonychia

plate dressings but no standard evidence-based approaches [4, 5, 12]. Unfortunately, most of the available data are from case reports and small cohorts of patients. On the other hand, trachyonychia is mostly a self-limiting disease especially in children who are the most commonly presenting age group of this abnormality, most probably due to an anxious parent(s) [14]. Thus, it is not unwise to consider a nonintervention approach and reassurance for childhood cases with idiopathic trachyonychia.

Topical Therapies

- *Emollients and moisturizers.* One out of three patients with idiopathic trachyonychia showed marked improvement after 2 months of applying a 6% urea cream, while another pediatric patient showed only partial improvement after petrolatum use for 22 months [19, 43].
- *Topical corticosteroids.* Potent topical steroids have been used for the treatment of trachyonychia whether idiopathic or disease associated, but its efficacy is not well documented. Variable responses were reported among six patients with idiopathic and disease-associated trachyonychia [19]. Similarly, the use of topical fluocinonide in eight pediatric patients was not associated with consistent results [43]. Clobetasol propionate under occlusion showed a substantial improvement in one patient [44], and betamethasone dipropionate combination with calcipotriol showed mostly partial response in 39 patients [45].
- *Tazarotene.* Topical un-occluded tazarotene 0.1% gel cleared trachyonychia associated with AA in one patient after 3 months of therapy [46].
- *5-Fluorouracil.* This had been reported successful in one patient with psoriasis-associated trachyonychia [47].
- *Tacrolimus.* One patient with idiopathic trachyonychia showed decreased roughness in all nails after 4 month of 0.1% tacrolimus ointment twice daily [48]. The author had found minor response in several patients with the use of tacrolimus 0.03% ointment under occlusion in combination with weekend clobetasol propionate in idiopathic trachyonychia.
- *Topical PUVA.* This had been reported to be effective in one patient after 7 months of therapy [49].

Intralesional Triamcinolone Acetonide For trachyonychia, intralesional triamcinolone matrix injections are a well-tolerated and effective technique in improving nail texture and functionality [50]. One injection to the proximal nail fold in four pediatric patients showed a 42% decrease in

pitting after 4 months [51]. Eleven of 19 patients (58%) treated for 6 months with intralesional triamcinolone at 4-week intervals showed improvement between 75% and 100% in their trachyonychia [52].

Systemic Therapies

- *Acitretin.* After 7 months of 0.3 mg/kg/day, acitretin was effective in clearing the nails of one patient with occupational trachyonychia due to psoriasis [53]. Excellent results were reported in a female patient with hypothyroidism and trachyonychia after 7 months of acitretin and clobetasol 8% nail lacquer for 10 months [54].
- *Alitretinoin.* More than 75% improvement was observed in 66.7% of a small cohort of patients after 6 months of alitretinoin 30 mg/day for idiopathic recalcitrant trachyonychia. Adverse event occurred in 42.9% of patients, and two (9.5%) withdrew from the study due to headache [55].
- *Biotin.* Oral administration of biotin 2.5 mg/day for 180 days reduced longitudinal ridging, thinning, and distal notching in two girls with idiopathic trachyonychia [56].
- *Cyclosporine.* The mean depth of nail roughness in four out of five patients with psoriatic trachyonychia improved after 3 months of cyclosporine treatment at a dose of 3 mg/kg/day [57]. *Oh* and *colleagues* recently showed a significant clinical response in 38 patients treated with cyclosporine 3–5 mg/kg/day with a pantothenic acid-complex-based dietary supplement in comparison to those treated with the dietary supplement alone. The latter authors reported that cyclosporine therapy group had more patients whose improvement was almost clear or whose improvement was marked or moderate than the control group. Moreover, the Dermatology Life Quality Index (DLQI) was significantly lowered in the cyclosporine group in comparison to the controls. Mild adverse effects in 10.5% of patients in the cyclosporine group were reported [58].
- *Corticosteroids.* Oral mini-pulse betamethasone (4 mg once daily for two consecutive days/week) cleared idiopathic trachyonychia after 6 months of therapy in a 12-year-old girl [59].

There is no gold standard therapy for trachyonychia, and the fact that many cases resolve spontaneously may deem treatment unwarranted. However, combination therapy may be helpful in recalcitrant trachyonychia, and it may also help minimize adverse events of systemic treatments by reducing drug dosage (Fig. 5.9). As trachyonychia is a self-limited disease, it is sensible to start with topical medications for 2–3 months and then add intralesional triamcinolone if no improvement is noticed, and finally, a systemic agent may be added in resistant cases.

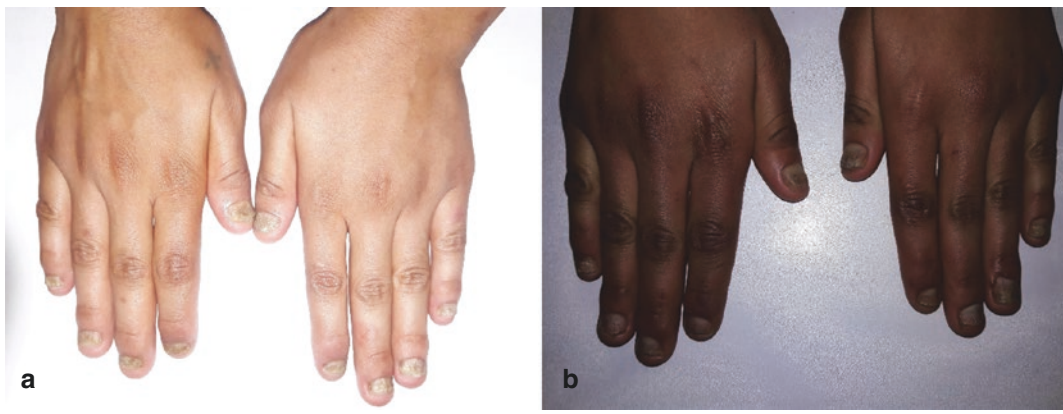

Fig. 5.9 Idiopathic trachyonychia before therapy (**a**) and 2 months after cyclosporine 50 mg/day and intralesional triamcinolone once monthly (**b**)

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