Trachyonychia: A Case Report and Review of Manifestations, Associations, and Treatments

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GOAL

To recognize the clinical presentations of trachyonychia

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Evaluate the etiology of trachyonychia with physical examination and diagnostic tests.
2. Recognize that trachyonychia has a variety of associations.
3. Recommend appropriate treatment for trachyonychia.

CME Test on page 323.

This article has been peer reviewed and approved by Michael Fisher, MD, Professor of Medicine, Albert Einstein College of Medicine. Review date: March 2003.

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Dr. Scheinfeld reports no conflict of interest. The author reports off-label use of flurandrenolone tape, triamcinolone, prednisolone, etretinate, psoralen, UVA light, and 5-fluorouracil. Dr. Fisher reports no conflict of interest.

Trachyonychia ("rough nails") is best considered a reaction or morphologic pattern with a variety of clinical presentations and etiologies. It may involve only 1 or as many as 20 nails (20-nail dystrophy). It can be a manifestation of lichen planus, psoriasis, alopecia areata, immunoglobulin A deficiency, atopic dermatitis, and ichthyosis vulgaris. Nail matrix biopsy results and physical examination findings help in establishing the cause of this condition, though often trachyonychia is an isolated finding. When trachyonychia occurs in childhood as a manifestation of lichen planus, it tends to resolve with time. We review a case of trachyonychia, its association, its diagnostic evaluation, and treatment options.

Accepted for publication February 28, 2003.

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Trachyonychia means "rough nails." This condition may involve only 1 or as many as 20 nails. It is best considered a reaction or morphologic pattern with a variety of clinical presentations

and etiologies. Clinical presentations are rough nails with a sandpapered appearance and numerous small superficial pits that make the nails shiny[1]; onychorrhexis, onychoschizia, distal chipping, and yellow onychauxis of the great toenail; and closely arranged longitudinal ridges, distal notching, and layered splitting.[2,3] Nail matrix biopsy results combined with clinical findings have linked trachyonychia with lichen planus generally,[4] lichen planus in children,[5] psoriasis,[6] alopecia areata,[7] IgA deficiency,[8] atopic dermatitis,[9] and ichthyosis vulgaris.[10] The term *20-nail dystrophy of childhood*[11] refers to a trachyonychia variant likely caused by lichen planus. Some who consider the term a misnomer—in part because not all nails are necessarily involved—think that perhaps it should be abandoned.[12]

Case Report

A 10-year-old girl presented with a 1-year history of worsening nail dystrophy. The patient had no history of psoriasis, atopic dermatitis, alopecia, or other skin disease, and family history was unremarkable. Except for dystrophy and hyperkeratosis identified on nails of both hands and both feet (Figure), physical examination findings were normal. Results of a fungal nail culture were negative, and the nail matrix biopsy specimen showed a bandlike lymphocytic infiltrate in the superficial dermis, with vacuolar alteration of the basal level. The diagnosis was trachyonychia secondary to lichen planus. Daily use of flurandrenolone tape and monthly intralesional injections of triamcinolone 2.5 mg/mL did not improve this patient's condition. After 4 months of injections in the distal nail folds, she was lost to follow-up.

Ten-year-old girl with a diagnosis of trachyonychia secondary to lichen planus had dystrophy and hyperkeratosis on nails of both hands and both feet.

Comment

Often, the onset of trachyonychia is insidious. The condition usually develops on all nails simultaneously. Trachyonychia also can occur on individual nails over many months. Peak age of onset is 3 to 12 years. Trachyonychia occurs, however, in multigenerational families,[13] in all age groups, in twins in the United States[14] and Europe,[15] in both sexes, and in all ethnic groups. This condition has been associated with ichthyosis vulgaris combined with alopecia universalis,[16] ungual lichen planus and alopecia areata,[17] koilonychia,[18] primary biliary cirrhosis,[19] and vitiligo.[20] In chronic graft versus host (GVH) disease, trachyonychia can be an isolated finding[21] or part of a constellation of cutaneous symptoms.[22] It may be associated with dystrophy, atrophy, and, often, ulceration of the lunula.[23] In the proper setting, the nail findings and clinical presentation of chronic GVH disease can resemble those of dyskeratosis congentia.[24] A mother and her 7-year-old daughter with chronic GVH disease had balanced translocation 46, XX, t(6q13;10p13).[25] A 15-year-old white boy with chronic GVH disease had recurrent episodes of immune thrombocytopenic purpura, autoimmune hemolytic anemia, and mild depression of immunoglobulin levels.[26]

Nail matrix biopsy results and physical examination findings help in establishing the cause of trachyonychia, though this condition often is an isolated finding.[27] In the case of lichen planus,[28] some patients also have flat polished purple papules on the body and white lacy or reticulated plaques in the mouth.[29] Nail biopsy specimens can show hyperkeratosis, hypergranulosis, and acanthosis in the ventral portion of the proximal nail fold and in the nail matrix; a bandlike lymphocytic

infiltrate in the superficial dermis; and vacuolar alterations in the basal layer. Nail abnormalities can develop in 1% to 10% of patients with lichen planus.[30] In the case of psoriasis, psoriasiform plaques sometimes develop on other body areas, and nail biopsy specimens can show psoriasis evidence such as psoriasiform hyperplasia and neutrophils. In the case of atopic dermatitis, spongiosis[31] (intercellular edema of the epidermis) also can occur in nail matrix biopsy specimens.[32] In the case of alopecia areata, lymphocytes can be present in the nail matrix, patches of nonscarring alopecia can develop on the scalp, and nail pits can develop in a gridlike pattern (giving a pounded brass appearance) on the nail plates. Evaluation of trachyonychia should include a check for fungus—a fungal culture or periodic acid–Schiff staining of a nail clipping. Some authors have suggested that longitudinal nail biopsy may be a useful diagnostic tool in certain cases of acquired nail dystrophy.[33]

Hazelrigg et al[11] stated that trachyonychia is self-limited and self-resolving in children. Specifically, trachyonychia tends to resolve with time when it occurs in childhood as a manifestation of lichen planus. Rarely, there is nail destruction in 20-nail dystrophy. If destruction occurs, the diagnosis is lichen planus—a form not restricted to the proximal nail fold but extended to the matrix. If the matrix is involved in lichen planus, a pterygium can develop—a manifestation rarely seen in 20-nail dystrophy.

Treatments for trachyonychia include intralesional injections of triamcinolone 2.5 to 3 mg/mL into the proximal nail folds.[2,34] Injections are painful and thus difficult in children. Medications for systemic treatment include prednisolone,[35] antimalarials,[36] and etretinate.[37] Seven-month therapy with topical psoralen and UVA light is reported effective.[38] In treating psoriatic nail disease, topical 5-fluorouracil[39] and cyclosporine[40] are useful. Clear nail hardeners can be applied to nails to improve their appearance.

In a study of 15 children, intramuscularly injected triamcinolone acetonide 0.5 to 1 mg/kg per month was prescribed for children with typical nail lichen planus.[41] Therapy duration was increased from 3 to 6 months, until the proximal half of the nail was normal. No treatment was prescribed for patients with 20-nail dystrophy or idiopathic atrophy of the nails. Treatment with systemic corticosteroids was effective in curing typical nail lichen planus. For 2 children, the disease recurred during follow-up. Recurrences were always responsive to therapy. Two children with 20-nail dystrophy improved without any therapy. Nail lesions caused by idiopathic atrophy of the nails remained unchanged during follow-up.

Trachyonychia and 20-nail dystrophy continue to present difficulties in classification, diagnosis, and treatment. With the advent of new immunomodulators, it is hoped that more effective treatments will be developed. Prompt diagnosis of these conditions aids in patient education and therapy.

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