Twenty-nail Dystrophy: Report of a Case and Review of the Literature

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A case of twenty-nail dystrophy in an 7-year-old girl is presented. The histopathological findings were incompatible with lichen planus. The differential diagnosis is briefly discussed, and the literature reviewed. *Key words: Nail disease; Lichen planus; Trachyonychia.* (Received November 10, 1983.)

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In 1977 Hazelrigg et al. (1) described under the term "twenty-nail dystrophy" (TND) a nail dystrophy of childhood in which all 20 nails are uniformly and simultaneously affected with excess longitudinal ridging and loss of lustre without other skin, hair and teeth abnormalities. This disorder was formerly described in 1950 by Alkiewicz (2) under the term "trachyonychia" and in 1974 Acthen & Wanet-Rouard (3) took up the term in a classification of nail dystrophies of either known or unknown causes. We recently had the opportunity to study a patient with onichodystrophy that involved all 20 nails.

CASE REPORT

An otherwise healthy 7-year-old girl was first seen in December 1982 for an asymptomatic nail dystrophy. Changes occurred at the age of 4 years and were noted first in the fingernails, followed in few months by similar changes in the toenails. There was no family history of psoriasis, alopecia areata, lichen planus, and other dermatologic and immunologic diseases.

Physical examination revealed (Figs. 1–2) all 20 nails to be thin, fragile, rough, opalescent, longitudinally ridged, and distally split. The proximal and lateral nail folds, as well as the skin, hair, teeth, and mucous membranes were all normal. The routine laboratory tests, including quantitative serum immunoglobulins, gave results within normal limits. Potassium hydroxide preparations and cultures of nail scrapings were constantly negative for pathogenic fungi and bacteria.

A biopsy of the matrix of a nail was performed according to Scher (4). The histologic examination showed psoriasiform epithelial hyperplasia with hypergranulosis. The dermis was free of inflammatory infiltrate (Fig. 3).

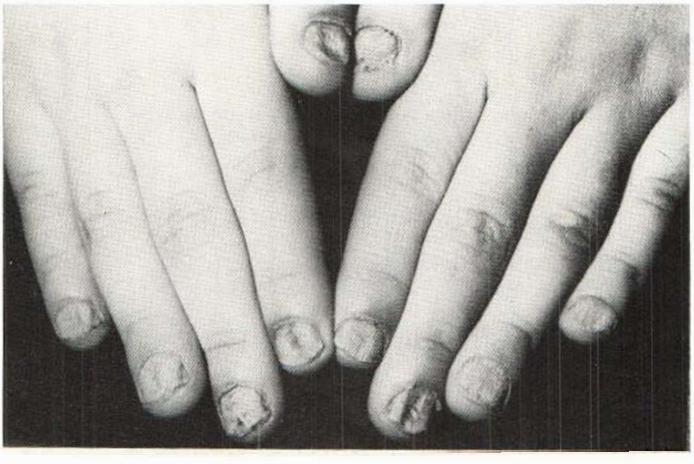

Fig. 1. Clinical appearance of the fingernails.

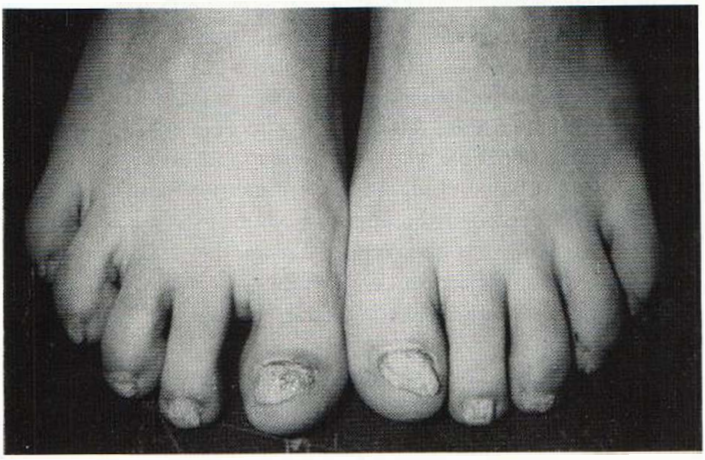

Fig. 2. Clinical appearance of the toenails.

DISCUSSION

TND is a rare entity in which all 20 nails are uniformly and simultaneously affected. Cardinal features include excessive longitudinal striations and loss of nail lustre in all 20 nails. The nail plate may be thin and fragile with distal notching, although thumb and great toenails may be thickened, yellow, and rough. The condition is thought to be idiopathic with insidious onset in early childhood. No treatment is reported to be effective, but the condition is felt to be self-limited with slow resolution. Even if most cases of TND are sporadic (1, 5, 6, 7, 8, 9, 10), some families have been reported in whom TND was transmitted through successive generations with an autosomal pattern of inheritance (11,

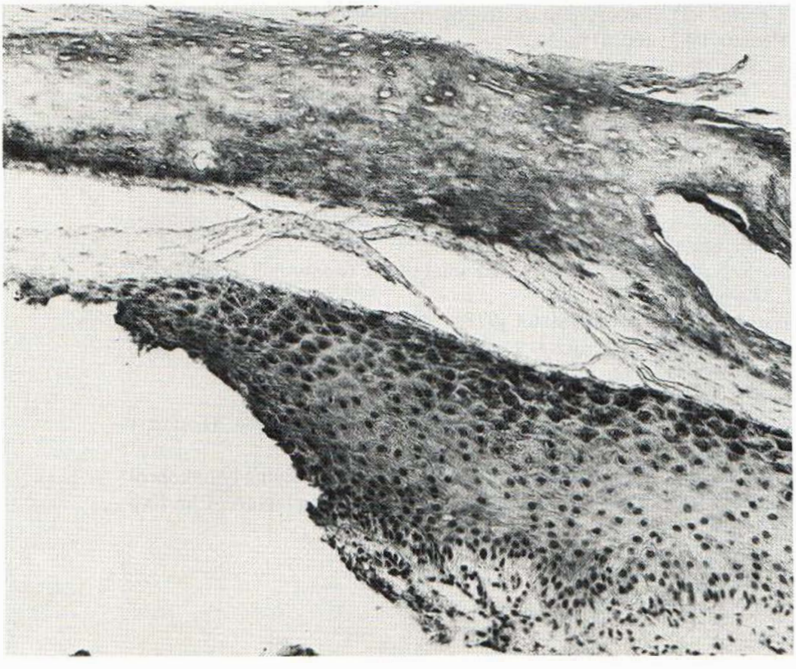

Fig. 3. Nail biopsy showing psoriasiform epithelial hyperplasia with hypergranulosis. Hematoxylin-eosin, ×100.

12). A few cases have been described in adults (13, 14). The pathogenesis of TND is controversial. Many cases associated with alopecia areata (6, 14, 15) have been reported. Scher et al. (9) believe that this condition represents a variant of lichen planus, as they found histological features compatible with a diagnosis of lichen planus. Wilkinson et al. (10) report a case in which the histopathological findings are felt distinctive (eczematous changes with mononuclear cell infiltrate), and not compatible with lichen planus. Electron microscopy features have been described by Braun-Falco et al. (16). A symmetrical nail dystrophy with clinical features suggesting TND preceded the appearance of systemic amyloidosis in one case (17). A selective IgA deficiency has been reported in another case (7). However, it has been hypothesized that immunological disorders could play a role in some cases of TND, particularly in those associated with alopecia areata, lichen planus and psoriasis.

Clinically, our patient did not show the characteristic picture either of psoriasis or of alopecia areata; moreover the histological findings were incompatible with lichen planus. The differential diagnosis of TND has been recently reviewed (16). The authors stress that nail biopsies are necessary to exclude some common disorders, especially lichen planus and psoriasis. The reported case was diagnosed as having TND because of the simultaneous involvement of all 20 nails in addition to slow progression of the disease, that was associated neither with peculiar histological findings nor with other skin signs.

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