Two Cases of Twenty-Nail Dystrophy

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Twenty-nail dystrophy is an idiopathic nail dystrophy in which all twenty nails are uniformly and simultaneously affected with excess longitudinal ridging and loss of lustre. The pathogenesis is controversial, and the treatment is unsuccessful. It is thought to have a self-limiting and reversible nature when it develops in childhood, but in adults, it is unusual and exists persistently. We report here two cases of adult patients with dystrophy of all twenty nails, whose ages were 58 and 55. The disease had been present for one year and may be associated with alopecia areata in the 58 year old and an idiopathic condition in the 55 year old. Negative results were obtained on mycological studies. Biopsies taken from the nail bed revealed marked hyperkeratosis consistent with nail dystrophy. (Ann Dermatol 11(2) 86~89, 1999).

Key Words : Twenty-nail dystrophy, Alopecia, Idiopathic

Twenty-nail dystrophy was the term described first by Hazelrigg, Duncan & Jarrett[1]. It's an idiopathic nail dystrophy in which all twenty nails are uniformly and simultaneously affected with excess longitudinal ridging and loss of lustre[2]. Although, it may be caused by psoriasis, lichen planus, alopecia areata, and ichthyosis, some cases are thought to be idiopathic[1,3-6]. It affects both children and adults[7]. The pathogenesis is controversial, and treatment is unsuccessful.

In the Korean literature, three cases have been reported. Two cases were idiopathic[8,9] and one case was associated with alopecia universalis with chronic thyroiditis[10].

CASE REPORTS

Case 1.

A 58-year-old woman was seen in our department because of dystrophic nails. The changes began insidiously one year previously and were first noticed in the finger-nails, followed by similar changes in the toe-nails. The nail plates were yellowish, dull, fragile, and longitudinally ridged(Fig. 1). She was treated with itraconazole for three months at the drug store, but no improvement was shown. A dermatologic examination revealed alopecia areata, and it had persisted for thirty years. However, other diseases such as lichen planus, psoriasis, and atopic dermatitis were not observed. Routine laboratory values including antinuclear antibodies were normal or negative and a mycological examination proved negative. A nail biopsy taken from the nail bed showed marked hyperkeratosis, mild acanthosis, focal hypergranulosis and mild infiltration of mononuclear cells in the upper dermis, which were consistent with nail dystrophy(Fig. 2).

Case 2.

A 55-year-old woman with nail dystrophy visited our department. The changes began about one year previously and involved all the finger- and toe-nails. The changes consisted of brownish discoloration, longitudinal ridging, and loss of normal lustre(Fig. 3). A physical examination including skin, hair, teeth and mucous membranes did not show significant findings. A family history was not

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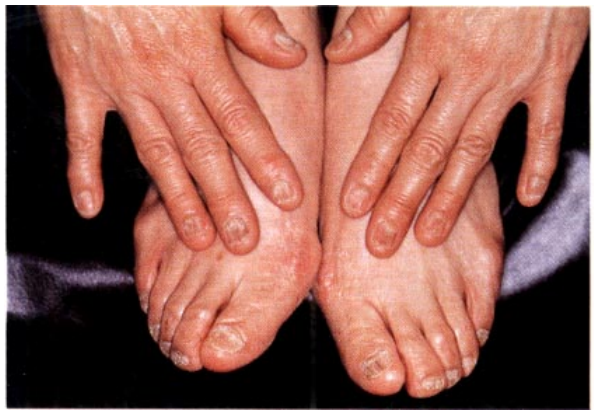

Fig. 1. The nails plates were yellowish, dull, fragile, and longitudinally ridged(case 1).

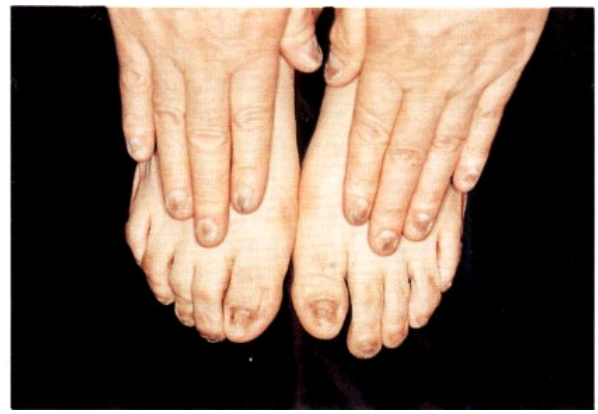

Fig. 3. Nail changes consisted of brownish discoloration, longitudinal ridging, and loss of normal lustre(case 2).

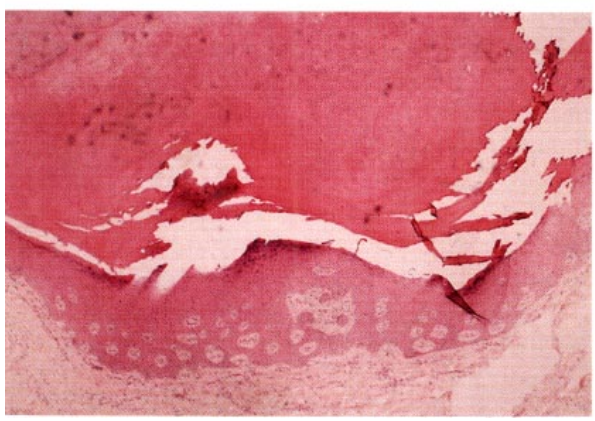

Fig. 2. Nail biopsy taken from the nail bed revealed marked hyperkeratosis, mild acanthosis, focal hypergranulosis and mild infiltration of mononuclear cells in the upper dermis(H&E stain, ×40)(case 1).

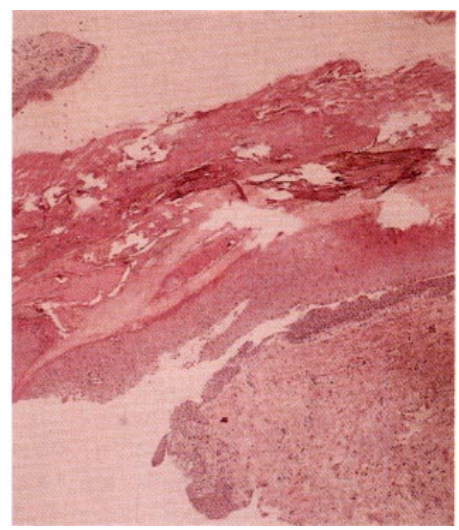

Fig. 4. Biopsy specimens showed marked hyperkeratosis, mild acanthosis, focal spongiosis and mild infiltration of mononuclear cells in the upper dermis(H&E stain, ×40)(case 2).

significant. On laboratory testing, there were no abnormal findings except mild hypercholesterolemia. A mycological examination turned out to be negative. Biopsy specimens from the nail bed showed marked hyperkeratosis, mild acanthosis, focal spongiosis and mild infiltration of mononuclear cells in the upper dermis consistent with nail dystrophy(Fig. 4).

DISCUSSION

Twenty-nail dystrophy is a clinical entity of onychodystrophy of all nails with or without other manifestations of skin disease. The term twenty-nail dystrophy of childhood was popularized by Hazelrigg et al[1]. in a report of children with acquried trachyonychia of all nails. They acknowledged the similarity of their findings to "excess ridging," a non-congenital nail abnormality described previously by Samman[11]. The six original cases were characterized not only by excess longitudinal ridging but also by onychorrhexis, onychoschizia, distal chipping, a dully opalescent appearance of the thin nail plates and, a yellowish

onychauxis of the great toenails. The onset was noted to involve either all the nails simultaneously or individual nails gradually through a period of several months. Cutaneous or mucosal signs of other diseases were not present, and twenty-nail dystrophy was considered to be a self-limiting abnormality that resolved slowly with age. Subsequent studies, however, suggested that twenty-nail dystrophy of childhood may not constitute a specific and distinct entity and also may not be confined to childhood.

It has been associated with several diseases such as alopecia areata, lichen planus, psoriasis and ichthyosis vulgaris and it can be called "idiopathic" in case of no other skin diseases[1,3-6]. In lichen planus patients, the incidence of coexistence has been lower than 10%[12,13]. In psoriasis and alopecia areata patients, it has been reported to range from 10% to 50% and 10% to 66%, respectively[14,15]. When it develops in childhood, we need to differentiate that from chronic mucocutaneous candidiasis which has widespread candida infection affecting mucous membranes, skin and nails and multiple endocrinopathies. In our patients, one had alopecia areata and the other was entirely free of skin, hair, dental, and bone diseases, and both of them had no significant family history.

Clinical manifestation consists of twenty opalescent dull nails. The nail plates are thin and fragile and have prominent, close-set, longitudinal ridging and distal notching with splitting into layers. The thumb-nails and great toenails are sometimes thickened, yellow, and rough. Potassium hydroxide preparations and Sabouraud agar culture for dermatophytes and yeasts are consistently negative[1].

Baran and Dupre[7] proposed the term "vertical striated sandpaper nails" and noted that adults, as well as children, could be affected. It is thought to have a self-limiting and reversible nature when it develops in childhood, but in adults it is rare and may exist persistently[15,16]. Our patients' ages were fifty eight and fifty five and the disease had been present in both cases for one year.

Histology of the twenty nail dystrophy is variable depending on the causes. Scher et al[17]. performed biopsies on several patients and uncovered histopathological findings of lichen planus. They concluded that twenty-nail dystrophy occurs in a subgroup of patients with lichen planus. However, Wilkinson et al[2]. presented histological findings that were incompatible with lichen planus and there was considerable distortion of the nail matrix with a fairly dense mononuclear inflammatory infiltrate below and within the matrix epithelium together with marked spongiosis. They thought that these changes suggested an eczematous disorder and that twenty-nail dystrophy of childhood might not constitute a single entity. Tosti et al[18]. suggested that nail dystrophy associated with alopecia areata was characterized by spongiosis, exocytosis, and mild to moderate lymphocytic infiltration in the proximal nailfold, nail matrix, nail bed, and hyponychium and proximal nail matrix was generally more severely affected than the distal matrix. In our patients, the histopathological findings showed marked hyperkeratosis, mild acanthosis, focal hypergranulosis, small focal spongiosis and mild infiltration of mononuclear cells in the upper dermis.

Treatment of twenty-nail dystrophy is unrewarding. Topical steroids, intradermal injections of triamcinolone acetonide suspension into the proximal nail fold, and extended administration of oral griseofulvin have been unsuccessful[1]. Some authors reported the effect of topical PUVA[19,20], although it is still controversial. In our cases, one patient was treated with topical PUVA and the other with topical steroids. However, there had not been any beneficial effects until the time of writing.

We report here two cases of twenty-nail dystrophy, which developed in a 58-year-old and a 55-year-old.

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