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Hallopeau's Acrodermatitis Continua of the Nail Apparatus: A clinical and Pathological Study of 20 Patients

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The clinical diagnosis of Hallopeau's acrodermatitis (HA) restricted to nail and digital pulp may be difficult, and even dermatologists often fail to recognize this condition. The aim of this study was to review the clinical and pathological features of 20 patients, observed over a period of 5 years (1988–1993), who were affected by HA limited to the nails. Our study shows that HA of the nail unit more commonly affects middle-aged females. In all our patients HA of the nail was restricted to one digit and not associated with other manifestations of pustular psoriasis. HA of the nail unit is characterized by a chronic course. None of our patients had a complete clearing of the dermatitis during the follow-up period. In 4 patients the acute phases of HA were treated with the non-steroidal anti-inflammatory agent nimesulide 200 mg/day, with great improvement of inflammatory signs and subjective pain within a few days. In these patients, prolongation of treatment with nimesulide during remission phases prevented relapses of the dermatitis.

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The clinical diagnosis of Hallopeau's acrodermatitis (HA) restricted to nail and digital pulp is often difficult. The aim of this study was to review the clinical and the pathological features of 20 patients, observed over a period of five years (1988–1993), who were affected by HA limited to the nails. Treatment and long-term prognosis of this condition are also discussed.

MATERIALS AND METHODS

Clinical findings and course

Twenty Caucasian patients (7 males and 13 females, mean age 46.4 years) were affected by a relapsing pustular dermatitis limited to the distal portion of the digits. Only one digit was involved in all cases: a finger in 15 patients, a toe in 5.

The dermatitis, which lasted from 2 months to 12 years (mean duration 27 months), was characterized by a relapsing course with acute recurrences followed by brief periods (20–45 days) of partial remission. Most of our patients had been unsuccessfully treated with topical and systemic antimycotics or antibiotics for months or even years.

A mechanical trauma had preceded the onset of the dermatitis in 5 patients. None of the 20 patients presented other clinical signs of vulgar or pustular psoriasis, including geographic tongue.

The family history for psoriasis was negative in all but 2 patients.

All patients were in good health, except for a 54-year-old woman who was affected by multiple autoimmune syndrome (MAS) type I.

Clinically, in acute phases the soft tissues of the distal portion of the affected digit were erythematous and enlarged, with multiple pustules that produced yellow scale crusts. Involvement of the nail bed with pustules and scale crusts that caused various degrees of onycholysis was evident in all patients (Fig. 1). In 2 patients the proximal nail fold showed inflammatory changes resembling acute paronychia. Twelve patients had nail abnormalities due to nail matrix involvement, including nail plate surface abnormalities (4 patients), onychomadesis (2 patients) and partial nail plate destruction (6 patients).

During acute stages patients complained of intense pain, spontaneously or following the use of the digit.

In the phases of remission, the digital pulp and periungual tissues showed mild erythema and scaling and the exposed nail bed was covered by hyperkeratotic scales. Mild atrophy of the pulp and the periungual tissues was observed in 2 patients who had been affected by the disease for several years.

None of our patients complained of rheumatological or osteoarticular symptoms.

Radiological examination of the affected finger showed focal resorption of the distal phalangeal bone in 2 cases.

Follow-up and treatment

Mean follow-up of the 20 patients was 10 months (range 2 months to 5 years). Nine of our patients have been followed for more than 2 years (mean 33 months, range 24 to 58 months).

In all patients the dermatitis showed a chronic-relapsing course during observation time. A complete clearing of the skin lesions was never observed.

Topical treatment with betamethasone 17-valerate 21-acetate in combination with 2% salicylic acid and 0.1% tretinoin improved the inflammatory signs and the local pain but never induced remissions or prevented relapses of the dermatitis. Two patients were treated with topical cyclosporin A (2% in oily solution twice a day) without clinical results.

In 4 patients the acute phases of HA were treated with the non-steroidal anti-inflammatory agent nimesulide 200 mg/day, with great improvement of inflammatory signs and subjective pain within a few days. In these patients, prolongation of treatment with nimesulide during remission phases prevented relapses of the dermatitis. However, the acute skin lesions recurred in all cases if drug intake was interrupted. In 2 patients a complete nail regrowth was observed after 3 months of nimesulide therapy. In one of them, nail plate regrowth was associated with the development of a band of longitudinal melanonychia. All these patients have been treated with nimesulide for 4 to 6 months without side-effects.

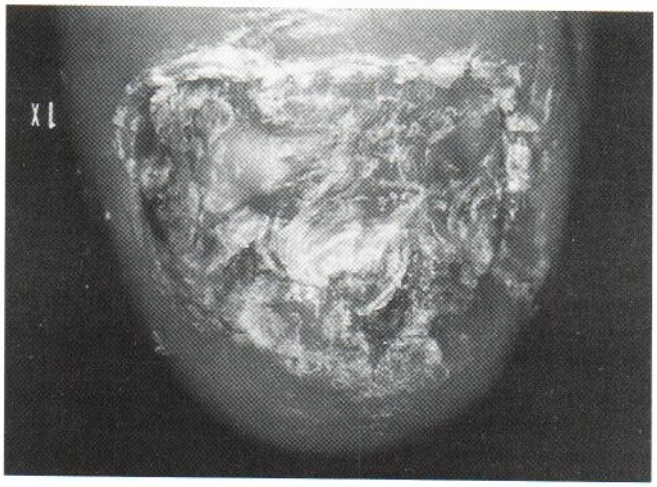

Fig. 1. Acute stage of HA. The nail bed shows a large pustule, hyperkeratosis and numerous scale crusts. The nail plate is absent.

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Fig. 2. The acanthotic nail bed contains mononuclear and neutrophilic exocytosis and coalescent spongiform subcorneal pustules (HE 80×).

Histology

Seven patients were submitted to a longitudinal nail biopsy and 13 to a nail bed biopsy. Specimens were formalin-fixed, paraffin-embedded and stained with H & E and PAS stain with and without diastase.

Pathological involvement of the nail bed was evident in all cases. The hyponychium was frequently affected in contiguity with the nail bed. Nail matrix involvement was observed in 6 of the 7 patients submitted to a longitudinal nail biopsy. In 2 of them the proximal nail fold was also affected.

Nail bed. The nail bed epidermis showed marked acanthosis with a variable number of spongiform pustules (Fig. 2). In 11 cases spongiform pustules coalesced to form intraepidermal or subcorneal pustules.

In 12 patients the nail bed showed a severe parakeratotic hyperkeratosis with numerous microabscesses of polymorphonuclear granulocytes in the parakeratotic horny layer. In 16 patients focal areas of hypergranulosis were also evident in the nail bed. The superficial dermis contained tortuous dilated blood vessels, sometimes filled with neutrophils, and a moderate to dense lymphocytic inflammatory infiltrate.

Nail matrix. In 3 of the 6 specimens with nail matrix involvement, the nail matrix epidermis showed moderate acanthosis, lymphocytic and neutrophilic exocytosis and spongiosis (Fig. 3). The proximal nail plate showed parakeratotic foci associated with Munro microabscesses. In 3 patients the nail matrix epidermis showed no inflammatory changes. We observed, however, the disappearance of the keratogenous zone, which was replaced by a 3 to 5 cell-thick granular layer (Fig. 4). The resulting nail plate was thin and markedly eosinophilic.

Proximal nail fold. In 2 patients the epidermis of the ventral portion of the proximal nail fold showed moderate acanthosis and parakeratosis with numerous spongiform pustules. This resulted in nail plate surface irregularities.

DISCUSSION

Our study shows that HA of the nail unit more commonly affects middle-aged females. In all our patients HA of the nail was restricted to one digit and not associated with other manifestations of pustular psoriasis. Even in patients with long-standing disease, HA was limited to the nail apparatus and the distal pulp, the skin of the digit never being involved. HA of the nail unit is characterized by a chronic course. Cyclical pustular recurrences at monthly intervals prevent a normal nail growth. None of our patients had a complete clearing of the dermatitis during the follow-up period. In our experience, bone resorption was not common and apparently not related to duration or severity of the disease.

Differential diagnosis with onychomycosis and contact dermatitis can be difficult, especially in chronic phases, although the typical history of periodical acute flares should strongly suggest HA. This possibly explains why 8 of our patients had been affected by the disease for more than 1 year before the correct diagnosis.

HA of the nail is mainly a nail bed disorder, and the patholog-

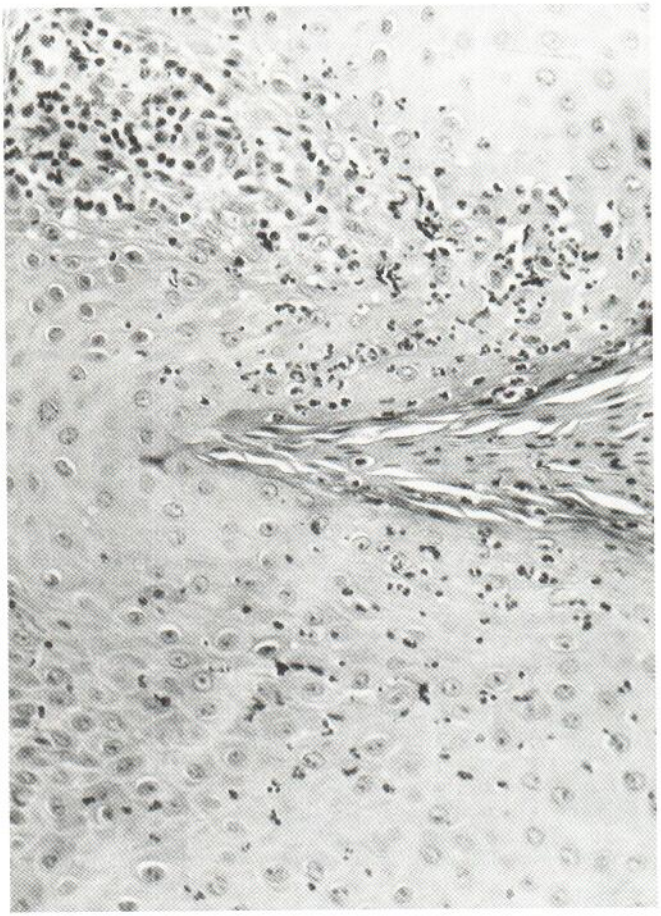

Fig. 3. The nail matrix shows neutrophilic exocytosis and mild spongiosis. Note parakeratosis of the newly formed nail plate (HE 125×).

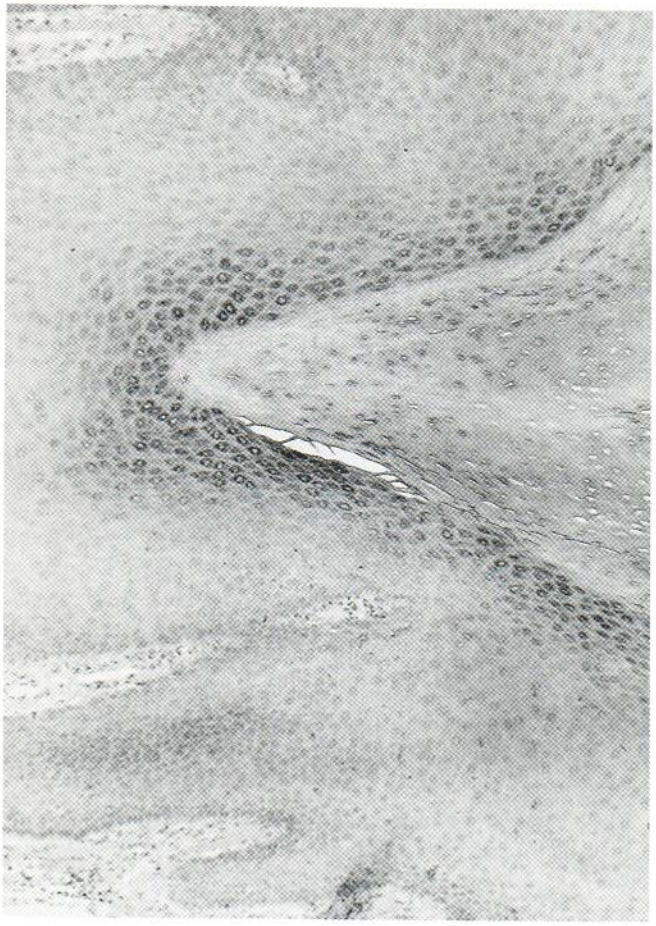

Fig. 4. Diffuse hypergranulosis of the nail matrix with disappearance of the keratogenous zone (HE 250×).

ical features of HA of the nail bed are comparable to those seen in pustular psoriasis of palms and soles (1). The nail matrix is only occasionally affected by the disease. When it affects the nail matrix, however, HA does not produce the typical large intraepidermal pustules but pathological changes similar to those seen in nail psoriasis (2). In some of our cases the nail matrix showed intraepidermal pustules, Munro microabscesses, focal parakeratosis and spongiotic changes. In other cases inflammatory changes were lacking and the nail matrix only showed indirect evidence of previous damage, i.e. a nail matrix keratinization through the formation of keratohyalin granules. This reactive pattern of the nail matrix is common to other inflammatory disorders, such of nail lichen planus (3) and spongiotic trachyonychia.

Treatment of HA is generally disappointing. Although systemic steroids (4), PUVA, retinoids (5) and cyclosporin A (6) may be effective, possible side-effects limit utilization of these drugs when the disease is restricted to one digit.

Topical treatment of HA does not produce satisfying results. Efficacy of topical steroids is poor, even when applied with occlusive dressing. Topical antimetabolites (mechlorethamine, fluorouracil) have been utilized with variable results in a limited number of cases, but a long-term evaluation of these drugs is lacking (7, 8).

Nimesulide (4-nitro-2-phenoxymethanesulfonanilide) is a methanesulfonanilide compound that blocks both classic and alternative complement activation paths and acts as a superoxide anion scavenger (9). It has a high potential to control the harmful effects of polymorphonuclear leukocyte oxidants (10, 11). Since few patients have received this treatment, it is too early to draw conclusions regarding its tolerability and efficacy; nimesulide, however, seems effective in treatment of HA, even though remissions are strictly dependent on drug intake.

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