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Chronically painful right thumb with pustules and onycholysis

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Clinical findings

A 67-year-old woman presented with a 5-year history of a chronically tender right distal thumb with pustules and nail dystrophy. She described recurrent episodes of pain and purulent discharge of her thumb and occasionally other digits. For years, these periods of discomfort had resolved with use of topical steroids, but most recently, the eruption on her thumb had been prolonged, and treatment with azithromycin and topical steroids had not improved her condition. She had no history of other skin disorders.

On examination, the right distal thumb was found to be tender to palpation. The nail bed and surrounding tissue were erythematous and oedematous, with extensive onycholysis. A well-circumscribed erythematous plaque with scale, fissures, lakes of pus and clear exudate extended beneath the lytic nail. The patient had no other skin lesions (Fig. 1).

Histopathological findings

Thumb X-ray, potassium hydroxide staining, and bacterial and fungal cultures were negative. The patient underwent nail-plate avulsion with two 3-mm punch biopsies of the nail bed. Pathology showed parakeratosis with neutrophils in the stratum corneum and spinous layer, an absent granular layer, epidermal hyperplasia, and suprapapillary thinning. The superficial dermis revealed dilated capillaries and sparse perivascular lymphohistiocytic infiltrate (figs 2 and 3). Staining

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with periodic-acid-Schiff was negative for fungal elements

What is your diagnosis?



Figure 1 The right distal nailbed is erythematous and oedematous with extensive onycholysis. A well-circumscribed erythematous plaque with scale, lakes of pus, and clear exudate extends beneath the lytic nail.

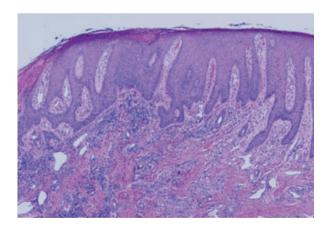


Figure 2 Parakeratosis with neutrophils in the stratum corneum and the spinous layer, absence of a granular layer, and presence of epidermal hyperplasia and suprapapillary thinning. The superficial dermis has dilated capillaries and sparse perivascular lymphohistiocytic infiltrate (haematoxylin and eosin, original magnification $\times\,40).$

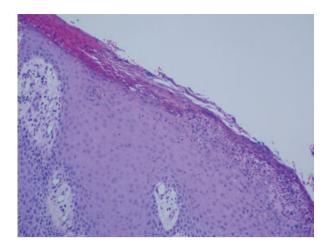


Figure 3 Parakeratosis with neutrophils in the stratum corneum and the spinous layer, absence of a granular layer, and presence of epidermal hyperplasia and suprapapillary thinning. The superficial dermis has dilated capillaries and sparse perivascular lymphohisticocytic infiltrate (haematoxylin and eosin, original magnification $\times 100$).

Diagnosis

Acrodermatitis continua of Hallopeau.

Discussion

Acrodermatitis continua of Hallopeau (ACH), first described in 1890, is a chronic, painful, sterile outbreak of pustules overlying an erythematous base on one or more distal digits. Significant nail-bed involvement, often with onychodystrophy, is typical. It can eventually result in anonychia or osteolysis of the distal phalynx. Rarely, it may progress to an acquired syndactyly, and in severe cases can involve the nasal tip in addition to the digits. Over time, it may progress proximally. 2.3

Although the eruption onset is often associated with trauma or infection, such an episode is not always present or remembered. Differential diagnosis based on clinical appearance includes chronic bacterial, fungal or viral paronychia, superinfected malignancy, secondarily infected contact dermatitis, or dyshidrotic eczema.

Histological findings are typically suggestive of a pustular psoriasis with characteristic subcorneal neutrophilic pustules. Indeed, this condition is considered by many to be a variant of pustular psoriasis, whereas by others it is considered a distinct entity. Progression to generalized pustular psoriasis, particularly in the elderly, has been reported.^{4,5}

ACH is often recalcitrant to treatment, though occasionally dramatic responses to therapy have been observed.³ Attempts to treat with systemic antimicrobials including tetracycline, azithromycin and dapsone, topical medications including calcipotriol and betamethasone, and immunomodulators such as prednisolone, methotrexate, acitretin, etanercept, infliximab, tacrolimus, colchicine, ciclosporin, psoralen ultraviolet A and combinations of these, have been met with mixed results.³ In our patient, treatment with tazarotene initially four times daily and then twice daily for 3 months was not successful. The patient also underwent phototherapy without improvement.

Learning points

- In patients presenting with chronic pustular outbreaks involving a distal digit or nail bed, culture may rule out infectious paronychia, and biopsy may be required to differentiate acrodermatitis continua of Hallopeau (ACH) from superinfected malignancy and contact dermatitis or dishydrotic eczema.
- In ACH, biopsy shows subcorneal neutrophilic pustules suggestive of pustular psoriasis.
- ACH is often quite recalcitrant to treatment. In rare cases, it may progress to anonychia, osteolysis, or acquired syndactaly.

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