CLINICAL VIGNETTE

Pustular Psoriasis Masquerading as Chronic Onychomycosis

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Acrodermatitis Continua of Hallopeau (ACH) is a rare variant of pustular psoriasis. ACH typically begins as an eruption of sterile pustules on the distal aspect of one digit and eventually progresses to inflammation of the entire digit along with severe onychodystrophy. The condition is frequently misdiagnosed as an infection and may be confused with a primary immunodeficiency[1]. We present a patient who was treated for apparent chronic, refractory onychomycosis for 10 years before nail biopsy revealed psoriatic inflammation consistent with ACH.

A 45-year-old man with a 10-year history of suspected onychomycosis of all fingers and toes was referred to Allergy-Immunology for evaluation of possible immunodeficiency, as his symptoms had been refractory to treatment. When his symptoms began, only a few nails were involved. The patient had been treated with topical and systemic antifungal agents, including ketoconazole, terbinafine, fluconazole and ciclopirox. Despite treatment, the condition progressed to involve all his nails. The inflammation became so debilitating that he could no longer work as a barber. Interestingly, he did report some relief with his most recent regimen of systemic fluconazole and topical ciclopirox, although he still experienced constant pain. On exam, his fingertips and toes were swollen, erythematous and tender to palpation (Figure 1). There were profound, well-demarcated induration and erythema around all nailbeds, and the nails were dystrophic and hyperkeratotic. The remainder of his exam was unremarkable, with no evidence of oral thrush, sinusitis, or pulmonary disease. He had no joint swelling or rash. On review of systems, the patient denied frequent infections, joint inflammation, or other symptoms suggestive of autoimmune disease or immunodeficiency. Neither his history nor his exam suggested an underlying endocrinopathy. Initial labs, including CBC and basic metabolic panel, were within normal limits, and nail culture grew no bacteria.

Although there was some initial suspicion for chronic mucocutaneous candidiasis (CMC), his presentation and history were not entirely consistent. He denied any history of oral thrush, which is the most common manifestation[2]. Second, despite the long duration of his symptoms, he had never developed any of the endocrinopathies (hypoparathyroidism or adrenal insufficiency) typically associated with primary CMC in the context of autoimmune polyendocrinopathy candidiasis-ectodermal-dystrophy (APECED)[3, 4]. Further, the patient had no medical conditions that would predispose him to secondary CMC, such as diabetes, HIV, or chronic use of immunosuppressive medication[3].

The patient was referred to a dermatologist, who suspected that the patient may have ACH. Nail biopsies were performed, revealing psoriasiform and spongiotic dermatitis with intracorneal and subcorneal neutrophilic pustule formation. (Figure 2) Infectious stains were negative for fungal, bacterial and herpes simplex organisms supporting the diagnosis of ACH.

This case of local psoriatic inflammation went undiagnosed for a decade. Given the physical exam findings of diffuse nail and digit inflammation with associated onychodystrophy, chronic candidal onychomycosis was a consideration. However, unlike onychomycosis, the degree of the nailbed induration exceeded the extent of the hyperkeratosis. Moreover, chronic candidal onychomycosis is unlikely in a patient with no other evidence or history of immune deficiency. In contrast to the other entities in the differential diagnosis for this patient, particularly CMC and APECED, ACH is remarkable for the absence of systemic complications.

ACH is a localized variant of pustular psoriasis that develops without typical psoriatic plaques. Sterile pustules form on the distal digit and within the nailbed, leading to severe onychodystrophy. The inflammation often spreads to the entire hand or foot, and longstanding disease may result in local osteitis and osteolysis[5]. In elderly patients, ACH may

progress to generalized pustular psoriasis[1]. A constant histologic feature of ACH is the subcorneal/intraepidermal spongiform pustule of Kogoj[5, 6]. Regular acanthosis, parakeratotic hyperkeratosis, and intracorneal collections of neutrophils (Munro's microabscesses) may be seen as well[5, 6].

The etiology of this condition is unknown, although it has been associated with preceding trauma or infection[1, 7]. One report describes a case of ACH that developed after treatment with terbinafine for onychomycosis[8]. This article hypothesizes that terbinafine caused ACH, as terbinafine may be associated with dermatologic adverse effects and the development of psoriasis[9]. Similarly, our patient had a limited form of nail dystrophy initially diagnosed as onychomycosis that progressed after treatment with ketoconazole. This proposed association raises the possibility that fungal infection or treatment of fungal infection precipitated ACH in our patient. However, there is no way to determine if his early nail dystrophy was a true infection or simply an early manifestation of ACH. Similarly, the proposed association between terbinafine and ACH may represent a spurious rather than causal relationship, as the patient in the previous case report may also have been misdiagnosed with onychomycosis rather than early ACH.

ACH is notoriously difficult to treat. Traditional anti-psoriatic therapies frequently fail to adequately control ACH, and there are no established treatment guidelines. However, reports describe improvement in symptoms and even remission with a range of different therapies including acitretin, calcipotriol, calcineurin inhibitors, phototherapy, and TNF-alpha inhibitors[7]. The initial treatment plan for our patient includes acitretin and high-potency topical steroids.

This case illustrates the similarities in clinical presentation between chronic onychomycosis and pustular psoriasis. As in our patient, ACH may cause significant morbidity and diminished quality of life. His case underscores the importance of early referral for biopsy in cases of onychomycosis refractory to traditional treatment.

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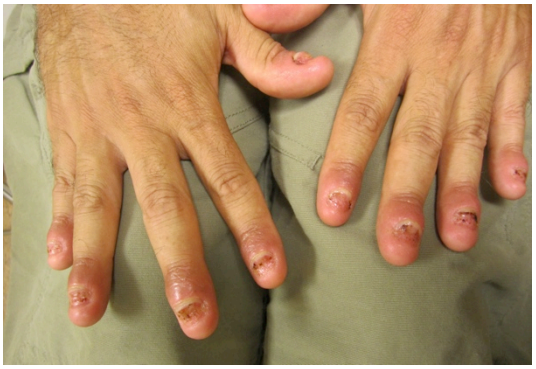
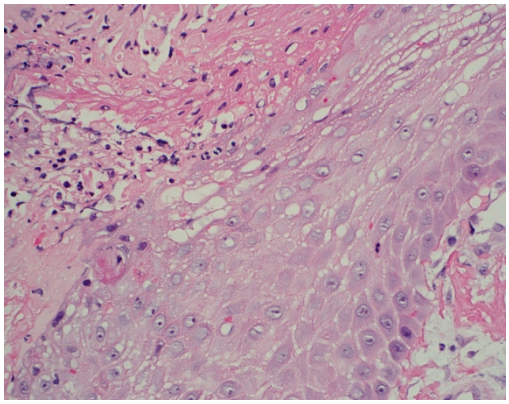
Submitted on April 8, 2014





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