

Single Case

Acrodermatitis Continua of Hallopeau with Bone Resorption in an 8-Year-Old Patient: A Case Report

May Alorainy^a Mohammed Alshaya^b Ammar Al-Rikabi^c
Mohammed Ayes^d Fahad Alsaif^e

^aCollege of Medicine, King Saud University, Riyadh, Saudi Arabia; ^bSaudi Commission for Health Specialties, Riyadh, Saudi Arabia; ^cDepartment of Pathology, King Saud University Medical City, Riyadh, Saudi Arabia; ^dDepartment of Radiology and Medical Imaging, King Saud University Medical City, Riyadh, Saudi Arabia; ^eDepartment of Dermatology, King Saud University Medical City, Riyadh, Saudi Arabia

Keywords

Acrodermatitis continua of Hallopeau · Bone resorption · Osteolysis · Pustular psoriasis

Abstract

Acrodermatitis continua of Hallopeau (ACH) is an uncommon inflammatory disease manifesting as sterile pustular eruption of the fingers and toes. The disease is of a chronic relapsing nature and is often refractory to treatment. With longstanding disease, osteitis with consequent bone resorption of the underlying phalanges can occur, leading to disability. While the incidence of ACH is rare in children, complications like osteolysis have not been reported previously in this age group. In this paper, we report the case of an 8-year-old boy with severe ACH complicated by bone resorption.

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Introduction

Acrodermatitis continua of Hallopeau (ACH) is an uncommon inflammatory disease manifesting as a sterile pustular eruption of the fingers and toes. The disease is of a chronic relapsing nature and is often refractory to treatment. The disease is more common in mid-

dle-aged women; however, it is difficult to estimate its true prevalence due to the rarity of the condition [1, 2].

ACH is often instigated by local trauma or infection to a single digit, which may lead to proximal extension subsequently. The clinical presentation involves small pustules in one or more distal fingers and toes in the acute stage to hyperkeratotic and scaly plaques with continuing pus formation in the more advanced stages [3]. ACH evolving into generalized pustular psoriasis (GPP) has been reported in the literature [4].

In addition to the characteristic clinical features, the diagnosis is made by histopathological examination, which typically displays a picture similar to that of pustular psoriasis. Imaging studies are warranted to assess the extent of underlying bone involvement and thus the severity of the condition [3].

In this case report, we present an 8-year-old male patient with extensive ACH complicated by bone resorption of the distal digits.

Case Report

Our patient was an otherwise healthy 8-year-old Saudi boy who presented to the dermatology clinic at King Khalid University Hospital, Riyadh with pustular eruption of the fingers and toes associated with burning sensation, dryness, and scaling for 8 months. Nail changes ensued during the course of the illness, leading to nail detachment of all fingernails and numerous toenails; however, the left fourth and right second, third, and fourth toenails were intact. There was no history of joint pain, lower limb edema, or jaundice. The patient had no family history of psoriasis. Physical examination revealed scaly erythematous plaques with overlying pus and crust formation involving all distal digits and multiple areas on the palms and soles. Nails showed severe onychodystrophy with confluent pustules covering the nail bed; however, the left fourth and right second, third, and fourth toenails were spared (Fig. 1a, b). A skin punch biopsy was taken and revealed psoriasiform hyperplasia, parakeratosis containing numerous neutrophils (Munro-like microabscesses), and focal spongiosis with absent granular layer. The dermis showed increased vascularity within the dermal papillae. This was accompanied by the presence of chronic inflammatory cells infiltrating the upper dermis. These features were consistent with subacute psoriasiform dermatitis/pustular psoriasis (Fig. 2). On radiological examination, there was focal soft tissue swelling at the tips of fingers of both hands with irregular outline and fissure-like indentation. Underlying terminal tufts showed focal areas of bone resorption, particularly over the fourth and fifth left digits (Fig. 3). X-ray of the feet did not show any remarkable joint or osseous abnormalities. Based on the clinical and histological findings, a diagnosis of ACH was made. Other laboratory tests including complete blood count, liver function test, renal function tests, hepatitis B and C serology, anti-HIV test, QuantiFERON tuberculosis test, and chest X-ray were normal. The patient was started on etanercept 25 mg subcutaneously twice weekly.

Discussion

The clinical features of ACH vary according to the stage of disease development. It presents acutely with small pustules affecting distal fingers or toes which often coalesce to form lakes of pus. These pustules usually rupture, leaving a shiny erythematous base where new

pustules develop. As the disease progresses, the affected areas tend to become hyperkeratotic and scaly with continuing pustulation. Involvement of the adjacent nail bed and matrix frequently result in severe onychodystrophy or even anonychia. With longstanding disease, osteitis with consequent bone resorption of the underlying phalanges can occur, leading to disability [2].

ACH is known to be a rare occurrence in childhood. A study evaluating the patterns and characteristics of 125 children with psoriasis included only 1 patient diagnosed with ACH [5]. A previous case report of ACH in a child of similar age was described, but manifestations were limited to a single digit only [6]. Additionally, Kiszewski et al. [7] reported ACH in a 2-year-old boy who had a more diffuse involvement of the distal portions of the fingers and toes. Although past cases were reported in children, this, to our knowledge, is the first case of extensive ACH complicated by osteolysis to have occurred in this age group.

ACH is thought to be a variant of pustular psoriasis, as both entities share a recessively inherited mutation in *IL36RN*, which encodes interleukin-36 receptor antagonist (IL-36Ra). This mutation leads to deficiency of IL-23Ra, a condition termed DITRA which was identified in familial types of GPP, palmoplantar pustular psoriasis, ACH, and other pustular skin disorders. This is further supported by cases of ACH evolving into GPP [8]. Similarly, Abbas et al. [9] reported a male patient diagnosed with ACH with a homozygous missense mutation c.338C>T (p.Ser113Leu) in *IL36RN* together with his sister who had a history of GPP. Furthermore, in a recent report, a Chinese woman with longstanding ACH complicated by osteolysis was found to have a homozygous c.115+6T>C mutation in *IL36RN* [10].

The absence of treatment guidelines is attributed to a lack of controlled studies, which is justifiable by the infrequency of the illness. Many therapeutic modalities have been used for ACH with equivocal results. These include topical preparations such as corticosteroids, calcineurin inhibitors, and vitamin D analogs. Moreover, many reports have evaluated the use of phototherapy and systemic therapies, including immunosuppressive medications and biological therapy, with variable results [2]. Since ACH is considered a variant of psoriasis, it is assumed that inhibition of tumor necrosis factor α could achieve a successful response [11].

Statement of Ethics

Informed consent was obtained from the patient's legal guardian.

Disclosure Statement

The authors have no conflict of interest of any type related to this work. No funding was received from individuals or corporations in relation to this paper.

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Fig. 1. a Multiple erythematous crusted plaques of the distal digits of the feet involving the toenails. Courtesy Fahad Alsaif, MD. **b** More severe involvement of the digits of the hands bilaterally with evident detachment of the nail plates of the fingernails. Courtesy Fahad Alsaif, MD.

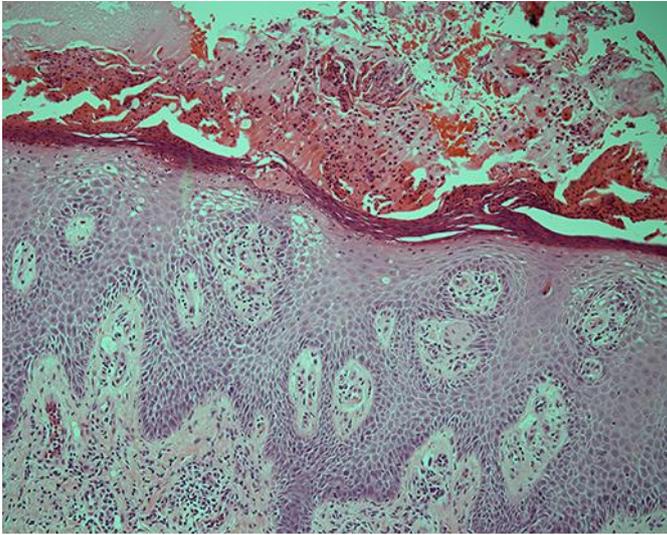


Fig. 2. Photomicrograph showing hyperplastic squamous epithelium with focal mild spongiosis and parakeratosis associated with scaly crust formation containing numerous neutrophils within the parakeratotic layer. Note the presence of chronic dermal inflammation. Hematoxylin and eosin stain, $\times 400$. Courtesy Ammar Al-Rikabi, MD.



Fig. 3. X-ray of the hands showing bone resorption of the distal phalanges mainly involving the fourth and fifth left digits. Courtesy Mohammed Ayesh, MD.