Acrodermatitis Continua of Hallopeau in a 32-Year-Old Female: A Case Report*

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ABSTRACT

Acrodermatitis continua of Hallopeau (ACH) is a rare, chronic, and recalcitrant inflammatory disorder classified as a localized variant of pustular psoriasis. Patients usually present with relapsing episodes of subungual pustules, nail dystrophy, and scaling. We report a case of ACH in a 32-year-old female, which developed following a nail infection and exacerbated during pregnancy, with no medication for 2 years. She presented at the clinic with severe manifestations of anonychia and multiple bone resorption on the distal phalanges. The patient was started on topical medication of combination corticosteroid and vitamin D analogue and oral methotrexate initially at 10mg/week then increased to 15mg/week due to poor response. Despite compliance to medications and avoidance of possible irritants, the patient still had relapse of pustules on the nails.

Several treatment options for ACH are available such as topical steroids, vitamin D analogue, systemic biologics, and non-biologics such as methotrexate and cyclosporine. However, systemic biologics are considered the most efficacious for ACH but financial constraints often limit their use in resource-poor settings.

KEYWORDS: Acrodermatitis continua of hallopeau; nail disorder; subungual pustules; pustular psoriasis; Methotrexate; case report

INTRODUCTION

Acrodermatitis Continua of Hallopeau (ACH) is a rare, chronic, and often recalcitrant pustular eruption of the fingers and toes. Its etiology is still unknown but it is considered as a subtype of pustular psoriasis. Treatment for ACH involves all the possible treatment options for plaque type psoriasis which are topical steroids, topical vitamin D analogue, methotrexate, cyclosporine, systemic retinoid, phototherapy, and biologics[1-3]. In an ideal setting, recalcitrant acrodermatitis continua of Hallopeau should be shifted to biologic therapy as these provide higher chances of remission. However, in a resource poor setting, the options we can offer our patients are very much limited.

CASE REPORT

A 32-year-old woman presented in our clinic with a 2-year history of relapsing episodes of pustulation, onycholysis, and anonychia on the fingers of both hands. The patient reported an unrecalled nail infection prior to the onset of lesions.

Disclosures: The author has formally acknowledged and signed a disclosure affirming the absence of any financial or other relationships (including personal connections), intellectual biases, political or religious affiliations, and institutional ties that could potentially result in a conflict of interest.

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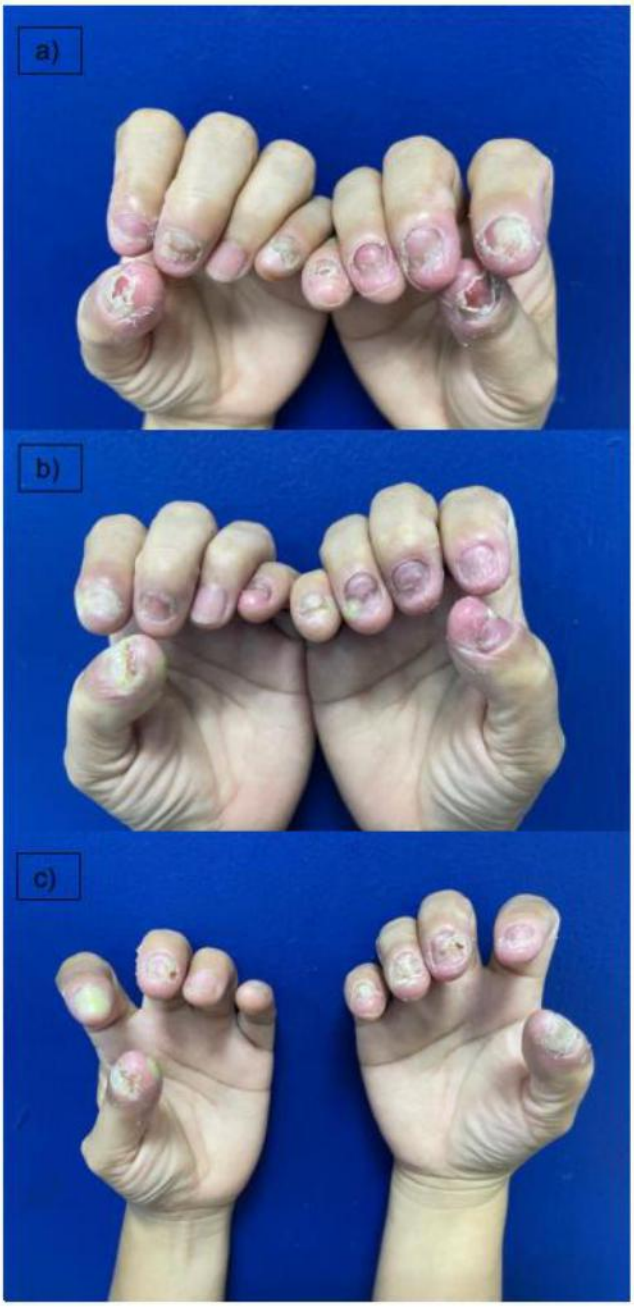
subungual and periungual areas. She reports rapid worsening of conditions during pregnancy.

Upon consultation and work-up at our institution, the patient was already found to have anonychia on the 3rd and 4th digits of the right hand and 2nd digit of the left hand as well as bone resorption finding on the distal phalanges of the 1st, 2nd, and 3rd digits of both hands on x-ray. She also complains of pain on the distal interphalangeal joints of both thumbs.

Differential diagnoses for this case such as onychomycosis, bacterial paronychia, irritant contact dermatitis, parakeratosis pustulosa, and palmoplantar pustulosis, were ruled out clinically and through diagnostics such as nail biopsy, periodic acid-Schiff (PAS) staining, KOH, and culture. Histopathological findings of confluent parakeratosis, hypogranulosis, psoriasiform hyperplasia with marked epidermal acanthosis and elongated rete ridges with suprapapillary plate thinning as well as dilated vessels within the papillary dermis and markedly dense superficial and perivascular infiltrates consisting mainly of neutrophils and lymphocytes strengthened the diagnosis of ACH.

The patient was then started on combination topical medication of corticosteroid and vitamin D analogue ointment: betamethasone dipropionate 0.05% + calcipotriol 0.005% ointment 2x a day for 2 weeks. She was also advised to avoid wet work and other possible irritants. Baseline laboratories were also requested prior to initiation of systemic medication. On follow up and finding of normal laboratory results, the patient was ideally to be started on biologics but due to financial constraint, patient was started on methotrexate initially at 10mg per week which she continued for 1 month. Despite compliance to treatment, the patient still would have relapsing episodes of pustulation, onychodystrophy, and onycholysis on all the nails of the hands except for the 4th digit of the left hand. Oral methotrexate was later increased to 15 mg per week. The patient was advised to come back after 2 weeks but was then lost to follow up

Figure 1. Nail dystrophy, anonychia, and pustules **(a)** pre-treatment, **(b)** 2 weeks of medications and **(c)** 8 weeks of medications



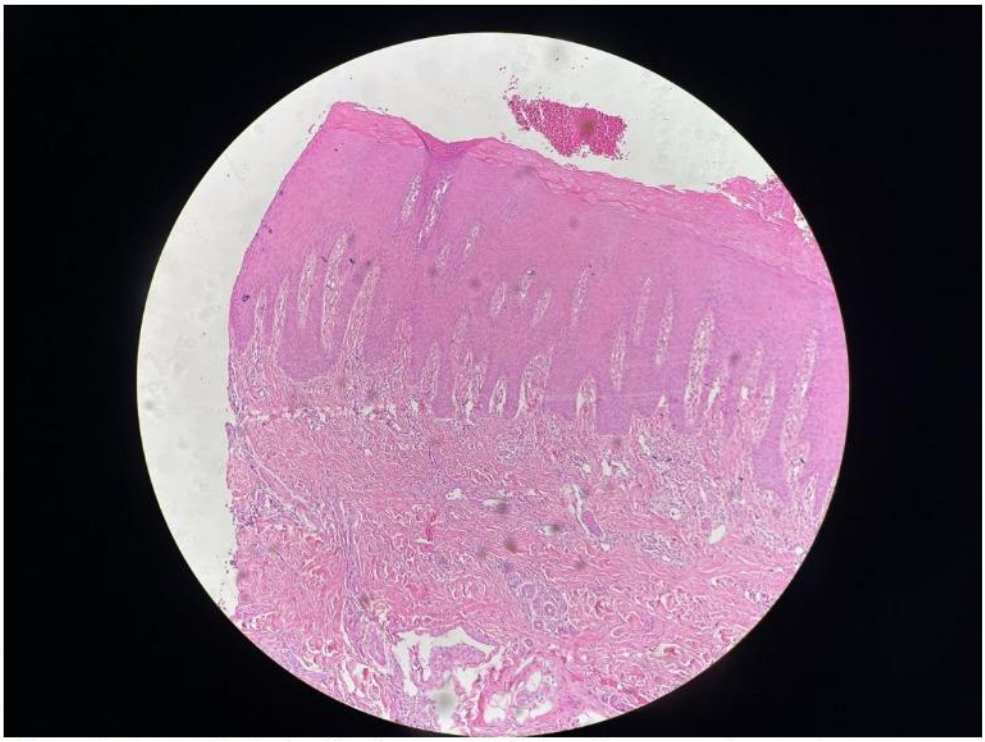


Figure 2: Histopathological findings revealed confluent parakeratosis, hypogranulosis, psoriasiform hyperplasia with marked epidermal acanthosis and elongated rete ridges with suprapapillary plate thinning as well as dilated vessels within the papillary dermis and markedly dense superficial and

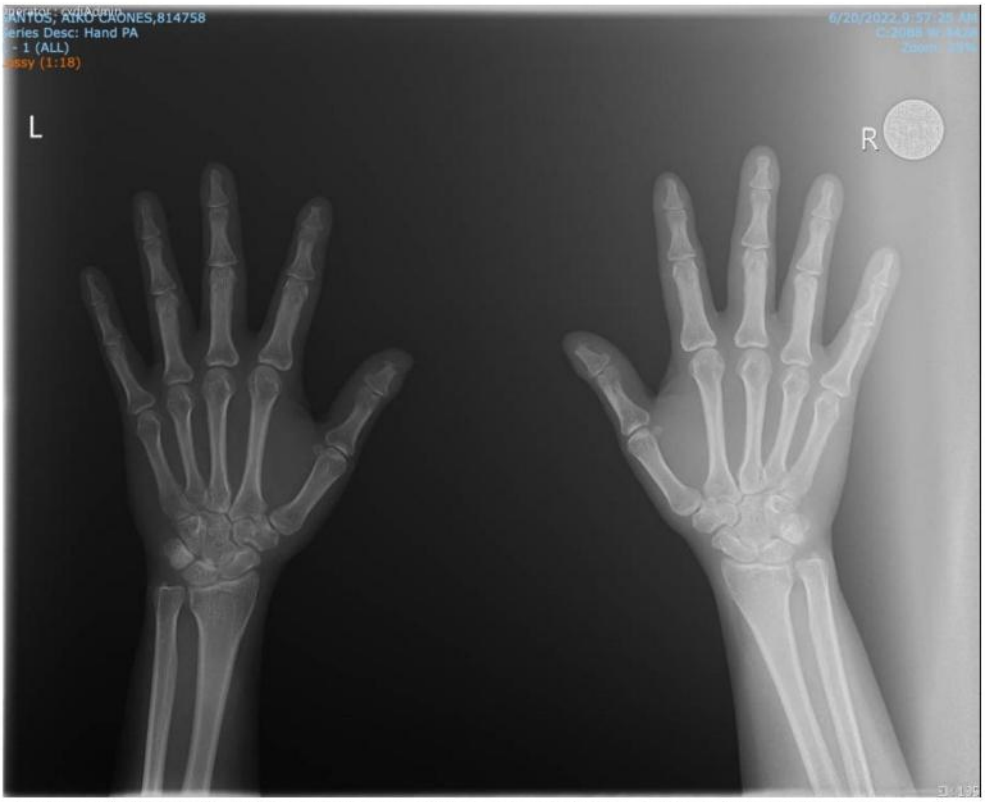


Figure 3: Radiographic evidence of bone resorption of the distal phalanges of the 1st, 2nd, and 3rd digits of both hands.

DISCUSSION

Acrodermatitis continua of Hallopeau (ACH) is a rare, chronic, and often obstinate inflammatory disorder. It is classified as a localized pustular psoriasis characterized by sterile pustules which initially affect the tips of the fingers[1-3]. A consensus statement from the European Rare and Severe Psoriasis Expert Network (ERASPEN) defines ACH as a primary, persistent (or more than 3 months) sterile, macroscopically visible pustules affecting the nail apparatus[4].

ACH is more common in middle-aged women, with very rare occurrence in childhood. Smoking history is common among patients but the reason is still unknown. ACH often begins after localized trauma or infection on a single digit[1]. Current evidence demonstrates that ACH is associated with a variety of genetic mutations in the genes IL36RN, CARD14, and AP1S3. The IL36RN gene encodes the IL-36 receptor antagonist (IL-36Ra), which normally acts to inhibit such pro-inflammatory signaling[3].

Pustule formation on the nail bed and nail matrix almost always occurs and may lead to severe onychodystrophy or even to anonychia. This eventually causes inflammation and sclerosis of the underlying soft tissue. Late stage of ACH can affect the bones resulting in atrophy of the distal phalanx. Spontaneous improvement is rarely observed and in some cases, elderly patients can progress to have generalized pustular psoriasis[1-3].

Treatment for ACH is mostly based on empirical data due to the paucity of case reports and research on this disease. Currently, the accepted treatment options include topical steroids, topical vitamin D analogue, methotrexate, cyclosporine, systemic retinoid, phototherapy, and biologics. Long-term strategy should be considered for the treatment of ACH due to its recalcitrant nature. Methotrexate is one of the most commonly used therapies due to its wide availability and price. However, very little is known about its effectiveness for ACH.

In countries where systemic biologics such as adalimumab, guselkumab, and secukinumab are widely used and available, most patients are given biologic medications early on, leading to less complications and higher chances of remission. However, the high cost of these medications makes them inaccessible to many patients, particularly in resource-poor settings. This financial barrier is a significant challenge in the treatment of ACH. A resource-poor setting should consider what the patient can sustain in the long run. Setting up financial aids for patients and modifying treatment regimen to suit what the patient can afford may also be tried, but safety and efficacy should be prioritized[1-3].

In conclusion, ACH is a rare and challenging condition to manage due to its recalcitrant nature and the paucity of research on effective treatments. Early and aggressive intervention is critical to prevent disease progression and improve patient outcomes. While biologics have shown the most promise in achieving long-term remission, their high cost limits access for many patients. In a resource-poor setting, treatment strategies must prioritize sustainability and cost-effectiveness while ensuring that patients receive the safest and most effective care possible.

INFORMED CONSENT

A written and signed consent for publication of this case report and its accompanying images was obtained from the patient.

DISCLOSURE

None of the authors were funded for this case report. There are no competing interests.

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