

A Case of Generalized Acrodermatitis Continua of Hallopeau

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Acrodermatitis continua (AC) of Hallopeau is one of the chronic relapsing pustular eruptions of the extremities. Generalization of AC sometimes occurred spontaneously, or induced by withdrawal of drugs such as corticosteroid, or pregnancy. We report a case of generalized AC in a 50-year-old woman who has been treated intermittently with various medications other than systemic retinoids. The histopathologic findings of the pustules on the fingertips and trunk revealed subcorneal abscesses and/or spongiform pustules. The patient was treated with etretinate in the dose of 50mg/day, and UVB exposure. Two months after treatment, skin lesions were markedly improved. (*Ann Dermatol* 5:(2) 141-145, 1993)

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Acrodermatitis continua of Hallopeau is also referred to as dermatitis repens or acrodermatitis perstans¹. It was originally described by Hallopeau² in 1890 as a suppurative process usually of the hands, particularly the fingertips, that may occur after trauma or infection. Lesions tend to be asymmetric, painful, destructive, and refractory to treatment. Sequential crops of pustules on an erythematous base coalesce to form lakes of pus that eventually form crusts. Paronychia involvement often leads to dystrophy and essential loss of nail plate. Generalized involvement can sometimes occur with extensive skin, mucous membrane, and systemic reactions that may terminate fatally. Treatment of acrodermatitis continua has proved to be difficult, with many modalities of therapy being only temporarily effective if at all¹. We report a case of generalized acrodermatitis continua of Hallopeau which responded well to systemic etretinate.

REPORT OF A CASE

A 50-year-old female patient presented with pustules on the left 1st, 2nd and 3rd fingertips and the right 1st fingertip and nail dystrophy. At the age of 4, she had a blunt trauma to the right 2nd fingertip. About 20 years ago, she had several pustules on the left 2nd fingertip, and the pustules gradually developed on the distal portion and subungual area of other fingers, especially the left 1st, 3rd and 5th fingers and the right 1st finger. She complained of intermittent itching and pain on the lesions. When the patient first visited our department, she was diagnosed as localized acrodermatitis continua of Hallopeau. She received systemic and topical antibiotics, corticosteroid, methotrexate and local PUVA treatment, but there was no remarkable improvement clinically. She had no more medication and 4 months later, she was admitted to our hospital due to a generalized skin rash associated with fever, chill, and general weakness.

Physical examination revealed discrete or confluent pustules, and crusts with erythematous patches on the lower extremities and trunk, and pustules, crusted patches, periungual erythema, and nail dystrophy on the distal portion of the

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left 1st, 2nd, 3rd, and 5th fingers, and the right 1st, 3rd, and 5th fingers (Fig. 1a, 2a).

Laboratory data including a complete blood count, serologic test for syphilis, liver function test, renal function test, C-reactive protein, rheumatoid factor, antistreptolysin O test, serum cholesterol, triglyceride, electrocardiogram, chest roentgenogram, thyroid function test, hand roentgenogram, costoclavicular roentgenogram, KOH smear, and fungus culture were within normal

limits or negative except for an increased erythrocyte sedimentation rate (40mm/hr), and glycosuria (300mg/dl). Culture of the pustular contents of the fingertip lesions showed coagulase (-) staphylococcus. An X-ray film of lumbar vertebrae showed spondylolisthesis and herniation of nucleosus pulposus at the level of L3.

A biopsy specimen taken from pustules of the left 2nd fingertip showed large subcorneal abscess filled with many inflammatory cells mostly neu-

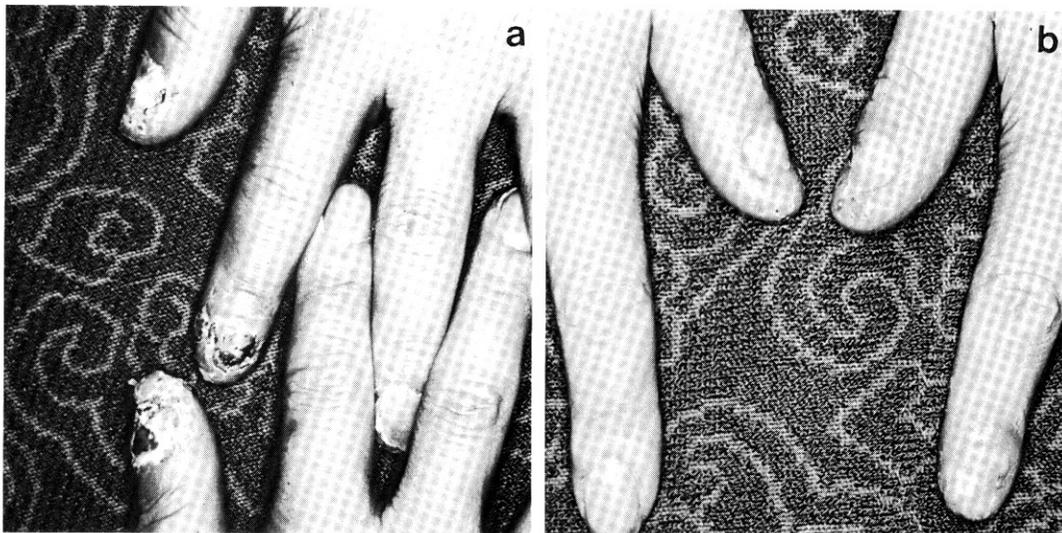


Fig. 1. Nail dystrophy, crusts and periungual erythema on distal portion of the fingers before treatment (a). Marked improvement of lesions 2 months after treatment (b).

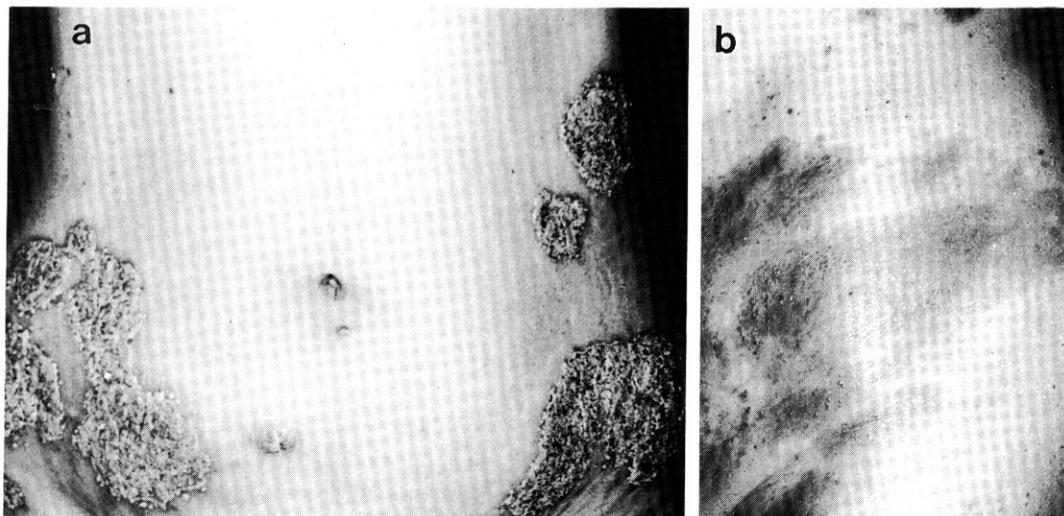


Fig. 2. Confluent crusted pustules on erythematous base on lower abdomen before treatment (a). Marked improvement of lesions 2 months after treatment (b).

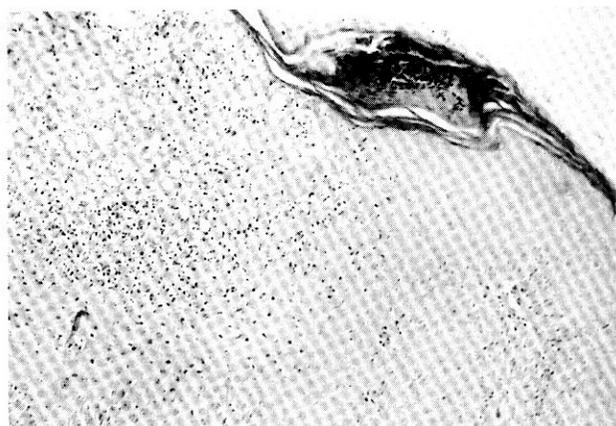


Fig. 3. Biopsy specimen of pustule on right abdomen shows spongiosis, acanthosis in upper stratum malpighii, and Munro's microabscess and spongiform pustules (H&E, $\times 100$).

trophils and RBC, and spongiform pustules on upper stratum malpighii. Pustules on the right abdomen showed Munro microabscess, spongiform pustules, acanthosis, and hypogranulosis (Fig. 3). Treatment was started with oral etretinate 50mg/day, and 10 days after administration of etretinate, the pustules were dried up and no new pustules developed on the trunk and extremities.

Also she received ultraviolet light B exposure 2 times per week, and itching was controlled with systemic antihistamines. One month later, the finger lesions improved. She complained of a mild dry mouth and cheilitis due to etretinate, and 2 months after admission and treatment, the lesions of the trunk and fingertip area were cleared (Fig. 1b, 2b). There were two times of recurrence to date when she was followed up for 15 months, but she was well controlled with only etretinate administered for 30 days and 50 days per each recurrence.

DISCUSSION

Pustular eruptions of the extremities have been categorized and labelled by many authors^{1, 3, 4, 5}. Among these labels are pustular psoriasis, pustular bacterid, acrodermatitis continua, pustular pompholyx, dermatitis repens, acrodermatitis perstans, and palmoplantar pustulosis. Acrodermatitis continua of Hallopeau is considered by many to be synonymous with acrodermatitis perstans, or dermatitis repens^{6, 7}. Today, some authors consider

acrodermatitis continua of Hallopeau, acrodermatitis perstans, and dermatitis repens as the same entity^{3, 4}. Others believe that dermatitis repens remains a distinct entity^{8, 9}. Although disagreements exist, many divide the persistent pustular eruptions of the extremities into three major categories 1) acrodermatitis continua of Hallopeau, 2) pustular psoriasis of the palm and sole and 3) pustular bacterid of the hands and feet^{1, 3, 5}.

Clinically, acrodermatitis continua may be seen in children, but is rare in young adults, unlike palmoplantar pustulosis, it is not unusual for it to begin in old age. It is commoner in females. The first lesions start on a finger or thumb more often than a toe. Onset is often related to minor trauma or infection at the tip of digits. The skin over the distal phalanx becomes red and scaly, and pustules develop. The nailfolds and nailbed may be involved, leading to nail dystrophy. Slow proximal extension is the rule, but this may be spread over years, eventually other digits may be involved. Bony changes can occur with osteolysis of the tuft of distal phalanges⁴.

Slawsky and Libow¹⁰ consider AC as a distinct disease with several features that distinguish it from other pustular eruptions. The presence of painful destructive lesions in an acral often paronychia distribution helps to separate this condition from PPP or pustular psoriasis. Studies of patients with AC and other pustular eruptions of the palms and soles have not demonstrated an increased incidence of HLA-B13 and-Bw17, as seen in psoriasis vulgaris^{11, 12}.

Acrodermatitis continua may evolve into generalized pustular psoriasis (GPP) especially in the elderly⁴. Several cases of generalized AC are reported in the literature, most cases^{8, 13, 14} are a spontaneous generalization of it, but some cases were induced by withdrawal of a steroid^{15, 16, 18}, administration of nystatin¹⁴, and by pregnancy¹⁷. However Lyons¹⁹ reviewed factors involved in the initiation of generalized pustular psoriasis episodes. These are tapering of systemic corticosteroid, intercurrent infections, pregnancy and drugs, including nonsteroidal antiinflammatory agents, antibiotics such as penicillin, sulfonamides, lithium, morphin, alcohol, salicylates with codeine etc, and sunlight, and finally emotional stress. Re-

cently Miyagawa et al²⁰ reported generalization of palmoplantar pustulosis after withdrawal of etretinate.

AC has been notoriously difficult to treat. Among reported successful therapeutic agents are sulfapyridine⁹, topical mechlorethamine, intramuscular steroids, psolaren and UVA, razoxane, and topical nystatin, neomycin and clobetasol¹⁹. More recently, treatment of AC type-pustular psoriasis with low dose cyclosporine²¹, and topical use of fluorouracil²² was reported.

The new retinoid etretinate has shown excellent results in treatment of patients with pustular psoriasis²³ and pustulosis palmaris et plantaris¹. Orfanos et al²⁴ indicate that starting patients on dosage of 75mg/day causes rapid resolution of lesions of pustular psoriasis. AC lesions improved with a daily dose of 50mg to 70mg of etretinate¹. This is similar to the results reported for pustular psoriasis^{23, 24}.

Numerous agents have been used in the treatment of generalized pustular psoriasis (GPP). It has become apparent that systemic steroids in the treatment of GPP, are at best "double-edged swords" due to precipitation of GPP, and prolonged exacerbations of GPP in many cases. Also these agents have proved to trigger a severe rebound of the diseases when withdrawal is attempted¹⁹. In addition to steroids, antimetabolites including methotrexate, hydroxyurea, azathioprine, aminopterin and 5-fluorouracil have proven beneficial in certain instances¹⁹. Among the non-antimetabolic drugs that have in some few instances been successful in the treatment of GPP are dapsone, clofazimine and colchicine. PUVA has also been used with some success¹⁹. Some authors^{25, 26} reported that combined treatment with etretinate with PUVA is more effective than etretinate or PUVA alone for palmoplantar pustulosis. Lastly, the retinoids are presently being evaluated for the treatment of pustular psoriasis including GPP with exciting results¹⁹. Two successful cases^{28, 29} of GPP were reported in Korea where skin lesions in a child and an infant were improved after treatment with etretinate. Clearing of GPP lesions was achieved with etretinate in doses averaging 1mg/kg/day clearing within 2 months, even in patients whose

disease had been refractory to methotrexate, steroids, and other agents for more than a year¹⁹. Maintenance therapy at lower doses was often required. White et al²⁷ showed that even with low doses of etretinate (30mg/day) for 12 weeks, their patients improved.

We treated our patient with etretinate (50mg/day) for about 3 weeks, and maintained a low dose (20 to 30mg/day) of etretinate for 5 weeks. She did not receive further medications, as her lesions were almost subsided and she was satisfied with the result of etretinate therapy. Although lesions were re-exacerbated twice on the fingertips and trunk during the follow-up period, they were well responsive to etretinate alone.

Most patients receiving etretinate experience some side effects. These side effects are dose-related and commonly consist of dry skin, cheilitis, dryness of mucous membranes, thirst, hair loss, elevated level of serum triglyceride and liver enzyme, and teratogenic potential²⁴. Our patient did experience mild thirst, dryness of mucous membranes, and cheilitis with mild elevation of serum triglyceride level.

In summary, we present a case of generalized acrodermatitis continua of Hallopeau, refractory to previous treatment of variable modalities including corticosteroid, methotrexate, and PUVA, that responded well to oral etretinate.

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