

Exanthematic Type of Pustular Psoriasis Consisting of Two Types of Pustular Lesion

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A 35-year-old female developed generalized pustules within a short period of time. Clinically, two distinct types of pustules were observed, viz. erythematous patches studded with crops of small pustules, and isolated large pustules with a red halo. Histologically, the former were subcorneal spongiform pustules, whereas the latter were unilocular pustules involving the hair follicular infundibulum or a subcorneal unilocular pustule on the palmo-plantar skin. On the basis of the sudden appearance of the pustules without any pre-existing lesions of psoriasis and the histological findings of the spongiform pustules, we made a diagnosis of the exanthematic type of pustular psoriasis. The lesions responded in a dose-dependent fashion to oral cyclosporin. Key words: Hair follicle; Cyclosporin A.

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A variety of cutaneous diseases are characterized by the formation of sterile subcorneal pustules. They include pustular psoriasis, impetigo herpetiformis, acrodermatitis continua, subcorneal pustular dermatosis, acute generalized pustular bacterid (AGPB) and pustulosis palmaris et plantaris (PPP) (1). Their clinical and histopathological features vary to some degree and so do the underlying pathomechanisms, although common chemotactic factors are involved in the formation of the sterile subcorneal pustules (2). We describe here a case of exanthematic type of pustular psoriasis presenting two distinct types of lesions, viz. crops of small pustules and isolated large pustules. The latter were similar to those reported in pustular bacterid (3,4).

CASE REPORT

A 35-year-old Japanese woman, with a history of asymptomatic ventricular septal defect, was admitted to our ward because of generalized pustules on the trunk and extremities. Anamnesis disclosed occasional isolated small pustule

formation on her knees for 20 years. Four months prior to this hospital admission, she developed symptoms of a common cold, which persisted, however, without complete remission for a few months. Thereafter, crops of slightly itchy, small pustules on an erythematous base appeared on her lower legs. She also suffered from fever, arthralgia and loss of appetite. Despite treatment by a local physician, which consisted of 1500 mg cefaclor and 40 mg gentamicin sulfate daily for 4 days, the skin lesion became widespread. Later, it turned out that the results of bacteriological studies of the pustular contents performed by him were all negative.

On examination, variously sized erythematous patches with numerous pustules, 0.5-3 mm in diameter, and scales were noted on the trunk and extremities (Fig. 1). Some pustules were confluent. Other isolated larger pustules, 3-7 mm in diameter, were scattered with an erythematous halo on the palms, soles and extremities (Fig. 2). Body temperature was 37.8°C.

Laboratory investigation of her blood showed an increase in white blood cell (WBC) count, 10,400/mm³ (normal, 3,400-9,400/mm³) and C reactive protein (CRP), 6.7 mg/dl (normal, <1.2 mg/dl) and a decrease in serum Ca level, 7.9 mg/dl (normal, 8.6-10.5 mg/dl). Antistreptolysin O (ASO) was normal at 74 Todd (normal, <160 Todd). HLA typing revealed A2, A31(W19), BW62(15), BW61(40), CW3, CW7, DR2, and DR4. The presence of chronic tonsillitis was detected by otorhinolaryngological examination. Because bleeding tendency due to probable platelet dysfunction was found in her preoperative hematological studies, scheduled tonsillectomy was cancelled.

A biopsy specimen of the grouped small pustules on an erythematous patch showed Kogoj's subcorneal spongiform pustules filled with mass of necrotic keratinocytes and an infiltrate of neutrophils (Fig. 3). A cellular infiltrate of lymphocytes, histiocytes and some neutrophils with a little nuclear dust were present around the upper dermal blood vessels, which showed a swelling of endothelial cells. However, neither fibrinoid degeneration of the blood vessels nor extravasation of red cells was observed. No immunoglobulin was deposited on the vessel walls. Both of two biopsy specimens obtained from the solitary larger pustules showed unilocular subcorneal abscesses at the infundibulum of the hair follicles (Fig. 4), and that from a pustule on the palm was also found to be a unilocular pustule. No microorganisms such as bacteria or fungal elements were demonstrated in these histologic specimens.

The patient was given 5 mg/kg/day of cyclosporin A (CsA). After 5 days, we added 2 g/day of piperacillin. Over a few weeks the number of pustules decreased, while the fever, WBC count, CRP and serum Ca all normalized. However, when the dosage of CsA was reduced to 2 mg/kg/day, the number of pustules quickly increased, but again

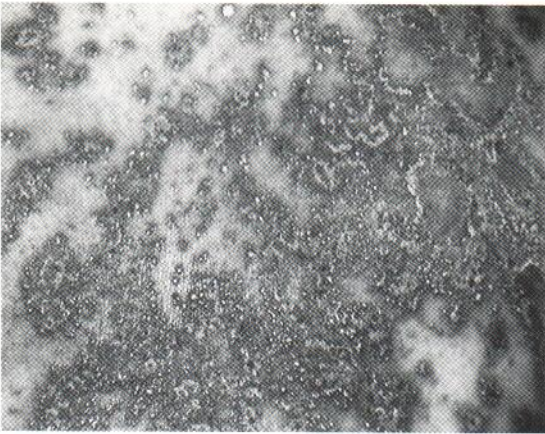


Fig. 1. Crops of pustules on an erythematous base on the back.

ceased to increase when the dose of CsA was increased to 5 mg/kg/day.

DISCUSSION

The clinical features of this patient are characterized by coexistence of disseminated erythematous patches studded with numerous fine pustules as noted in generalized pustular psoriasis and discrete large pustules with an erythematous halo scattered over the limbs, palms and soles as noted in generalized pustular bacterid (4). Biopsy specimens from the former revealed typical spongiform pustules, whereas the latter showed unilocular subcorneal pustules with only focal spongiform pustule formation at the infundibulum of the hair follicle in the case of extra-palmo-plantar lesions.

Our differential diagnosis included the following

diseases, i.e. impetigo herpeticiformis, acrodermatitis continua, subcorneal pustular dermatosis, PPP, AGPB and pustular psoriasis. However, except for pustular psoriasis, most of them could easily be excluded, both clinically and histopathologically. Impetigo herpeticiformis occurs during pregnancy. In acrodermatitis continua the disease starts as pustulation on the acral regions of the hands and feet. The eruptions of subcorneal pustular dermatosis occur clinically in the groins, axillae, submammary areas and the flexor aspects of the limbs. PPP is a chronic, relapsing disorder, whose extra-palmo-plantar lesions show unilocular, rounded pustules in the upper squamous layer (5). In AGPB, pustules develop on the dorsa of hands and extremities soon after focal infections. Histologically, leukocyteclastic vasculitis is noted in addition to unilocular pustules (4). Obvious spongiform pustules are usually absent in the last two dermatoses (1).

Pustular psoriasis, which is characterized histologically by spongiform pustule formation, was classified into four types by Baker & Ryan (6): 1) generalized von Zumbusch type, 2) annular type, 3) exanthematic type, and 4) localized type. In the exanthematic type the disease starts following upper respiratory tract infections such as common cold and chronic tonsillitis in individuals without any pre-existing lesions of psoriasis. The present case appeared to belong to this type. The patient suffered from symptoms similar to those of common cold for 3 months before the onset of the generalized pustular lesions. We could not determine the exact nature of this preceding disorder. It might be either a systemic manifestation of a mild form of pustular psoriasis or

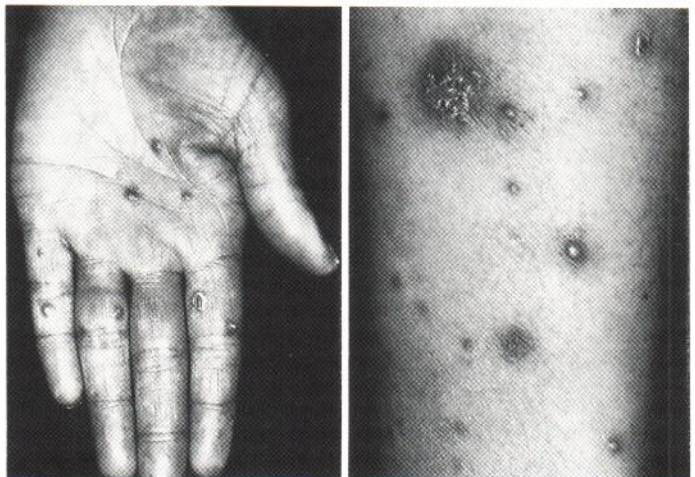


Fig. 2. Solitary pustules on the left palm (A) and the left forearm (B).



Fig. 3. Biopsy specimen of pustule in clusters on an erythematous base showing Kogoj's spongiform pustule. Hematoxylin-eosin, $\times 280$.

chronic tonsillitis. Pustular eruptions may result from intake of antibiotics as a form of drug eruption (7). But the drug eruption due to antibiotics that had been prescribed preceding the generalization of the pustular lesions was unlikely because the lesion did not regress after discontinuation of such treatment.

Burge & Ryan (3) reported 5 cases of acute palmoplantar pustulosis, which, they said, might be a limited form of the exanthematic type of pustular psoriasis. In contrast, our case is unique as the exanthematic type of pustular psoriasis, because of the coexistence of typical erythematous lesions of pustular psoriasis and isolated solitary pustular bacterid-like lesions. However, the role of a focal bacterial infection in our case was not clear, because no beneficial therapeutic effect was obtained with antibiotics. The pustular content itself was found to be sterile by bacterial studies.

Based on the report on the therapeutic effect of CsA in the treatment of generalized pustular psoria-

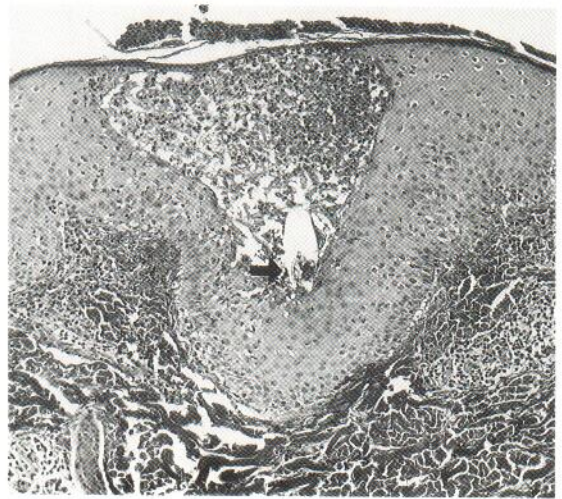


Fig. 4. Biopsy specimen of a solitary pustule showing an abscess in the infundibulum of the hair follicle. Arrow indicates a tiny hair shaft. Hematoxylin-eosin, $\times 88$.

sis (8), we administered CsA to this patient. CsA had a dose-dependent clinical efficacy.

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