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Case Report

Unusual Presentation of Acute Generalized Exanthematous Pustulosis with Unilateral Anterior Trunk Involvement

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Abstract

Acute generalized exanthematous pustulosis (AGEP) is a rare drug eruption characterized by the rapid occurrence of many sterile, non-follicular pustules, neutrophilic leukocytosis, high fever and spontaneous resolution usually within two weeks. The distribution of rash predilection in the trunk and intertriginous regions. In treatment, the causative drug must be initially discontinued. Topical corticosteroids, moisturizers, antipyretics, and systemic corticosteroids are generally used according to the patient's symptoms and AGEP severity. Cyclosporine and intravenous immunoglobulin therapy can be used in some patients with AGEP resistant to systemic corticosteroids in severe AGEP. We herein aim to report a unique case of AGEP that has different rash distribution and resistance to systemic corticosteroids.

Keywords: Acute generalized exanthematous pustulosis, rare presentation, trunk, unilateral

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Acute generalized exanthematous pustulosis (AGEP) is a severe cutaneous adverse reaction, often drug-induced, characterized by rapid onset of non-follicular, sterile pustules on an erythematous base with fever, neutrophilia, and peripheral blood leukocytosis.[1]

Symptomatic treatment is usually sufficient. In several cases, topical or systemic steroids can be used to control inflammation.[2]

Herein, we present a case of AGEP with an unusual rash distribution which was resistant to systemic corticosteroid.

Case Report

A 66-year-old male presented with fever, generalized redness, and itching two days after taking phenyramidol and acetaminophen/chlorzoxazone for muscle spasms. On the first examination, a well-demarcated rash affecting the left side of the trunk and both lower extremities, characterized by scaly, non-follicular pustules on an erythematous, edematous base, was observed (Fig. 1a). Interestingly, the distribution of the eruption was well demarcated on the

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left side of his trunk unilaterally and the right upper arm had not been involved (Fig. 1b). Blood tests showed leucocytosis (23.21*10^9/L) with an elevated neutrophil count (20.97*10^9/L). In addition, the C-reactive protein (CRP) rate was high (205.7). The other blood test parameters were within normal limits. He had a history of severe skin burns that affected his left body and left arm and a split-thickness skin graft was replaced on these burned areas which was taken from his gluteal and right axillar skin.

A 4 mm punch biopsy was obtained from the pustules in the erythematous area of his left upper leg. The histopathological examination revealed subcorneal pustule, spongiotic epidermis and eosinophil-rich infiltrate that supported the diagnosis of AGEP (Fig. 2).

Figure 1. (a) Non-follicular, confluent sterile pustules on the patients back. **(b)** Left-sided erythematous rash with desquamation. **(c)** The patient just before the discharging with mild hyperpigmentation.

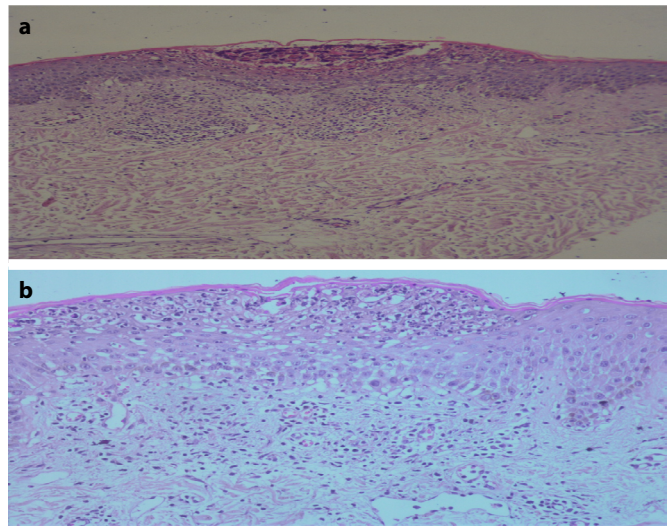

Figure 2. (a) Spongiotic epidermis and eosinophil-rich infiltrate. **(b)** Subcorneal pustule.

There was no bacterial growth in the blood culture sample and tissue culture sample. Therefore, we confirmed the diagnosis of AGEP according to AGEP validation score developed by the EuroSCAR study group.[3] Doppler USG was performed for venous thrombophlebitis in the right arm and leg due to unilateral involvement, though it concluded open and innate veins.

Initially, the patient received 60 mg/kg/day of intravenous methylprednisolone, topical steroids, and emollients. On the third day of his hospitalization, his condition worsened with fever and rash spread to the right side of his trunk, and edema occurred in his right arm and right foot. The treatment was switched to cyclosporine with a dose of 3 mg/kg/day on the 4th day of his hospitalization. Subsequently, the skin dropped fine scales, and the fever decreased. On the 14th day of hospitalization, the edema and redness decreased significantly (Fig. 1c). The control blood test revealed a 10.08*109/L leucocyte count and a 5.27 CRP rate. We reduced the dose of cyclosporine and discharged the patient. We considered acetaminophen as the causative drug. The drug patch test couldn't be performed due to cyclosporine use.

Discussion

Our patient had a well-demarcated distribution of rash on the left side of his trunk and both lower extremities. We could not find any AGEP case with a similar distribution pattern in the literature. Though we found one case of urticarial vasculitis with a unilateral rash pattern appeared on a previous mastectomy scar after anastrozole therapy. However, the reason for this distribution was unknown and authors suggested that previous breast surgery may have

caused anatomical deterioration of the vessels and regionally weaker circulation.[4] The pattern of our patient's rash distribution was compatible with the left lymphatic duct drainage areas. Therefore, we had Doppler USG imaging performed. But Doppler USG revealed that the vessels were open and innate. To explain this well-demarcated unilateral distribution pattern, we suggest that peripheral neural fibers and lymph vessels got damaged by skin burns. Hence, the neuromodulatory response was locally altered and the immunity overreacted on grafted skin tissue.[5]

The treatment for AGEP remains unclear and systemic corticosteroids are usually used as first-line therapy in severe AGEP cases. However, there are cases resistant to systemic corticosteroids. Cyclosporine selectively acts on T-cells by inhibiting calcineurin phosphorylase. Hence, it may play a role in inhibiting drug-specific T-cells in AGEP.[6] We searched the literature and found 4 patients who developed AGEP due to HCQ and were treated with cyclosporine because of their resistance to systemic corticosteroids.[7] Another remarkable aspect of our case is the successful use of cyclosporine in AGEP due to different drug from HCQ.

As a result, this case highlights that AGEP can be seen with atypical presentations and cyclosporine should also be considered for the management.

Disclosures

Informed consent: Written informed consent was obtained from the patient's parent.

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