Acute Generalized Pustular Bacterid: An Uncommon Dermatosis That Commonly Presents With Acral Pustules

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To the Editor:

We read with great interest the very well-presented article entitled "Acute Localized Exanthematous Pustulosis Due to Bemiparin" by Gómez Torrijos et al [1]. The authors reported the case of a patient with pustular eruptions on the palms, which were finally diagnosed as a localized subtype of acute generalized exanthematous pustulosis (AGEP). We recently saw a similar case with acral pustular eruptions, in which the diagnosis of acute generalized pustular bacterid (AGPB) was established based on the triad of acral distribution of the pustules, sudden onset of the disease, and concomitant remote localized bacterial infections.

A 42-year-old man had a 2-day history of pustular eruption on both hands that had developed suddenly. Several days earlier, he had had a sore throat with fever of 38.5°C. He denied having taken drugs and had no history of drug allergy, including to cefuroxime. Similarly, he had no history of psoriasis. Physical examination showed many isolated, discrete pustules on both hands (Figure). The results of routine testing of blood (6.2×10^9/L), urine, and stool and a basic biochemistry panel (ALT, 34 µ/L; AST, 26 µ/L) were normal. Laboratory results revealed C-reactive protein of 53 mg/dL and an erythrocyte sedimentation rate of 35 mm/h. Bacterial culture from a throat swab demonstrated multiple group A ß-hemolytic streptococci. Repeated cultures of pus for bacteria and fungus were negative. Skin biopsy of a pustule showed subcorneal spongiform accumulation of neutrophils, slight dermal edema, and perivascular infiltration (Supplementary Figure). No evidence of leukocytoclastic vasculitis was found. The patient was diagnosed with AGPB. Intravenous cefuroxime was administered at a single dose of 1.5 g twice daily for 7 days. The pustules cleared, with gradual resolution of the sore throat. No pustules were observed during the 1-year follow-up.

Skin manifestations of infections in other organs and tissues are diverse and are often the first observed signs of a disease. AGPB was first described by Andrews et al [2] in 1935. It has also been called pustular bacterid or pustulosis acuta generalisata [3], which is characterized by the presence of acral pustulosis, mostly with a focal infection [4].

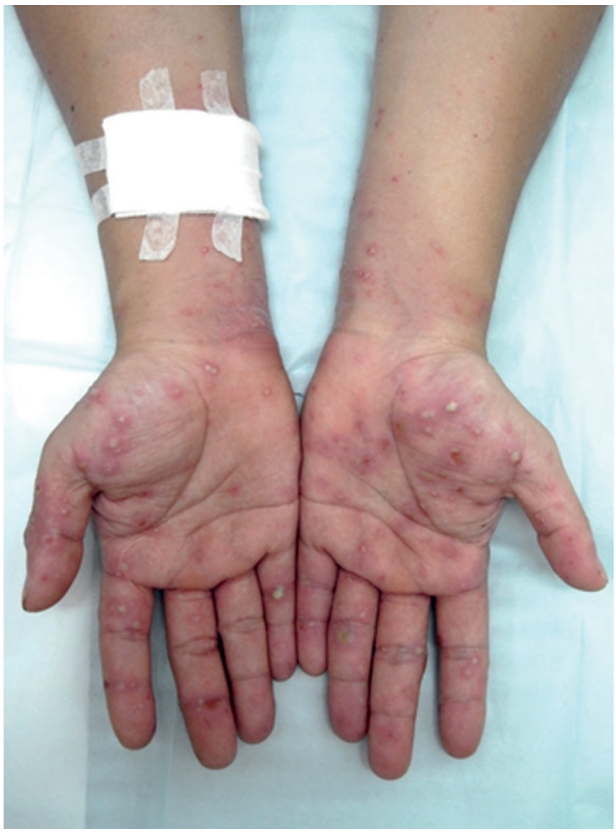

Figure. Multiple isolated discrete pustules surrounded by a narrow rim of erythema. The pustules vary in size from 2 to 6 mm in diameter and are distributed on both hands and, to a lesser extent, the forearms.

AGPB manifests as sterile, isolated, small pustules with an erythematous halo. The rash is neither edematous nor scaly, and the diameter of the pustules has a range of several millimeters. AGPB mainly affects the palms and the soles, and, to a lesser extent, other parts of the limbs [4]. Occasionally, AGPB is generalized. There is usually intermittent fever, and a few cases present rare complications (eg, glomerulonephritis, arthralgia, and ankylosing spondylitis) as the disease progresses. AGPB sometimes co-occurs with Tietze syndrome and sternocostoclavicular hyperostosis. Most pustules resolve within 12 days after onset. Skin specimens in AGPB show subcorneal spongiform accumulation of neutrophils and perivascular infiltration. Leukocytoclastic vasculitis and neutrophilic panniculitis can sometimes be observed [4].

The features that point us to the correct diagnosis of AGPB include absence of psoriasis or other skin conditions, focal

infection (eg, tonsils, gums, sinuses, vagina), and clearance of the pustules by eradication of the infection [2,4]. The triad of sudden onset of the disease, acral distribution of the pustules, and concomitant sore throat with high fever in the case we report was consistent with AGPB. The differential diagnosis for AGPB is exhaustive and includes pustular psoriasis, AGEP, palmoplantar pustulosis (PPP), and dermatophytid reaction (Supplementary Material). Unlike patients with AGPB, most patients with pustular psoriasis have a relapsing course with Munro microabscesses and psoriasiform acanthosis, which are diagnostic. The patient with AGEP usually has a history of drug intake, and the pustules are tiny and mostly affect the inguinal folds or other intertriginous areas [5]. All these features are sufficient to confirm AGPB. PPP is a recalcitrant recurrent afebrile pustular dermatosis. In contrast to AGPB, the typical PPP pustule is confined to the palms and the soles. Although similar pustules present in dermatophytid reaction, they take the form of generalized eczematous eruptions caused by remote localized infection by tinea or staphylococcal colonization, which can be excluded in the case we report.

Many factors contribute to the formation of pustules in AGPB. Onset is usually shortly after a focal infection such as pharyngitis or tonsillitis by group A β-hemolytic streptococci or other bacteria [4,6]. It is speculated that superantigens and toxins from the bacteria upregulate the expression of tumor necrosis factor α and interferon γ, leading to activation of the complement C3 and C5a cascade. Complement C5a has been proven to be an attractant for neutrophil accumulation in the epidermis and results in pustular eruptions.

There is a proven causative relationship between AGPB and focal bacterial infections, and AGPB usually follows a focal bacterial infection. Clinicians should consider this diagnosis in individuals with sudden onset of acral pustular eruptions. Recognizing and eradicating focal infections are the most important steps in the management of AGPB and can reduce misuse and overuse of antibiotics. Pustules in AGPB usually resolve spontaneously in 7-14 days without relapse; therefore, most authors agree that aggressive treatment is unnecessary [4]. Antibiotics are still one of the mainstays of treatment and improve outcomes in those who have previously been infected or who could develop complications of glomerulonephritis and reactive arthritis [4]. The fact that AGPB with tonsillitis is aggravated after tonsillectomy indicates that eradication of the infection alone is not a radical cure for some patients. Corticosteroids showed no beneficial effect on the patient, although methotrexate and group A streptococcal vaccination have proven effective in some patients.

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Conflicts of Interest

The authors declare that they have no conflicts of interest.

References

1. Gomez Torrijos E, Cortina de la Calle MP, Mendez Diaz Y, Moreno Lozano L, Extremera Ortega A, Galindo Bonilla PA, et al. Acute Localized Exanthematous Pustulosis Due to Bemiparin. J Investig Allergol Clin Immunol. 2017;27:328-9.
2. Andrews GC, Machacek GF. Pustular bacterids of the hands and feet. Arch Derm Syphilol. 1935;32:837-47.
3. Laimer M, Koller J, Selhofer S, Massone C. Pustulosis acuta generalisata and streptococcal vaginitis. J Eur Acad Dermatol Venereol. 2009;23:583-4.
4. Kamiya K, Ohtsuki M. Acute generalized pustular bacterid. J Gen Fam Med. 2018;19:32-3.
5. Bavbek S, Sozener ZC, Aydin O, Ozdemir SK, Gul U, Heper AO. First case report of acute generalized exanthematous pustulosis due to intravenous iopromide. J Investig Allergol Clin Immunol. 2014;24:66-7.
6. Iraji F, Abtahi-Naeini B, Heydari H, Pourazizi M. Pustular Bacterid: Look at the Throat for Prevention over Treatment. Int J Prev Med. 2015;6:51.

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