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# Circumferential Acute Localized Exanthematous Pustulosis

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## Introduction

Acute localized exanthematous pustulosis (ALEP) is a rare localized variant of acute generalized exanthematous pustulosis (AGEP), first described by Prange et al. in 2005.<sup>1</sup> It is clinically and histologically consistent with AGEP with the exception of its limited distribution on the body. It is characterized by an acute eruption of numerous sterile, coalescing, non-follicular pustules on an erythematous background, most commonly affecting the face, neck and chest.

Contrastingly, AGEP starts in body folds including the axillary, inguinal, and inframammary regions and rapidly spreads to the trunk and limbs within a few hours. Fever, leukocytosis, elevated C-reactive protein, and neutrophilia may accompany the eruption.<sup>2</sup> Systemic symptoms are less common in ALEP. AGEP is drug-induced in 90% of cases, but bacterial and viral infections are also possible culprits.<sup>2, 3</sup> Antibiotics are the most commonly implicated drugs, but others that have been described include anti-hypertensives, antifungals, neuroleptics, analgesics, and NSAIDs.<sup>2</sup> All reports of ALEP have been drug-induced. We present a case of ALEP favored to be secondary to an infectious trigger which has not been previously reported.

## Case Presentation

A 16 year-old African American female with a history of atopic triad presented to the emergency department with a pruritic and painful eruption for 2 weeks which started around entirely of hairline and subsequently spread to further involve forehead, neck, and ears. She also endorsed a non-productive cough over the same period. Physical examination revealed sheets of non-follicular pustules on a background of erythema, circumferentially encircling the hairline (Figures 1,2). The helix and post-auricular sulcus were also involved with superficial erosions. She denied a history of new hair products, medications, or known contacts with similar symptomatology. She had been on Claritin, Advair, and Albuterol for a long period of time. A 4 mm punch biopsy was performed which revealed epidermal hyperplasia with neutrophilic spongiosis and a subcorneal pustule in addition to underlying superficial perivascular mixed-cell infiltrate consisting of lymphocytes, neutrophils, and scattered eosinophils. Bacterial culture was negative.

## Clinical Presentation

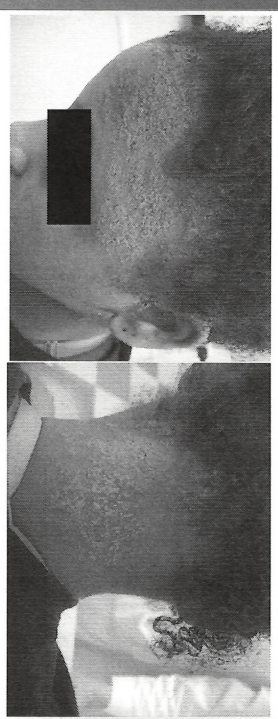


Figure 1

Figure 2

## Histology

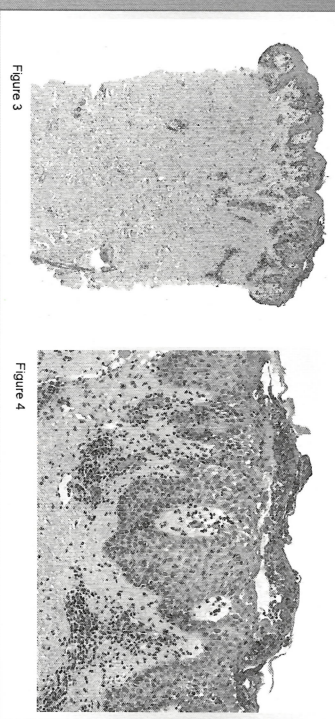


Figure 3

Figure 4

## Literature Review & Discussion

The pathogenesis of AGEP/ALEP, though not completely understood, is believed to be T-cell mediated through the production of high levels of cytokines like IL-3 and CXCL8, which recruit and activate neutrophils. Patch tests and in vitro tests have elucidated the important role of drug-specific CD4+ and CD8+ T cells in the development of the condition as these T cells produce significantly more CXCL8 in affected patients.<sup>4</sup> Histology is notable for spongiform subcorneal or intraepithelial pustules, perivascular infiltrates consisting of neutrophils and some eosinophils, focal necrotic keratinocytes, leukocytoclastic vasculitis, and papillary edema.<sup>2, 5</sup>

The differential diagnosis includes:

- 1. Pustular psoriasis of von Zumbusch:** This entity is often very difficult to clinically and histologically differentiate from AGEP/ALEP. It is typically triggered by withdrawal of systemic steroids or potent topical steroids and infections, and hospitalization is frequently required.<sup>6</sup>
- 2. Sneddon-Wilkinson disease:** The pustules seen in Sneddon-Wilkinson are larger and described as half- and-half vesicles typically sparing the face. The condition most commonly presents in middle-aged women, and rarely occurs in children and adolescents.<sup>7</sup>
- 3. Pustular contact dermatitis:** The lack of recent contact with possible allergens or irritants also refutes a diagnosis of pustular contact dermatitis.

There are only a number of case reports describing ALEP, and all of them are drug-related.<sup>8</sup> Besides the chronic use of Claritin, Advair, and Albuterol, this patient did not start any new medications that could explain the eruption. With the history of upper respiratory symptoms accompanying the rash, the likely etiology in this case was a viral infection. Though viral infections have been recognized as a cause of AGEP,<sup>2</sup> to our knowledge, this is the first reported case of non-drug-related ALEP. Treatment includes discontinuing any causative agents and symptomatic relief, including topical steroids for pruritus and antihistamines if it is not a suspected causative drug. The disease is typically self-limited, and the rash can be expected to resolve in less than 15 days, which occurred with topical steroid use in this case.<sup>9</sup>

1. Prange B, Harris A, Kella A, Haddad-Levalley H, Rocco T, Thongji US. Acute localized exanthematous pustulosis (ALEP). *J Clin Dermatol*. 2005;23(12):722-723.

2. Fournier JC, Souchon-Lange F, Bismuth C, Gilibert JC, Bernard P, Loe C, Pichot F, Clauvy A, Chavannes C, Vallat L. Acute generalized exanthematous pustulosis. Analysis of 83 cases. *Arch Dermatol*. 2007;143(12):1432-1435.

3. Chung SS, Hwang YH, Yang CH, Ho S, Hwang JS. Clinical manifestations and characteristics of patients with acute generalized exanthematous pustulosis in Asia. *Acta Derm Venereol*. 2007;87(4):473-476.

4. Thongji US, Probst WL. Acute generalized exanthematous pustulosis: a skin to neurologic-mediated inflammatory process. *Neurology*. 2005;65(12):2023-2024.

5. Fournier JC, Souchon-Lange F, Bismuth C, Gilibert JC, Bernard P, Loe C, Pichot F, Clauvy A, Chavannes C, Vallat L. Acute generalized exanthematous pustulosis: analysis of 83 cases. *Arch Dermatol*. 2007;143(12):1432-1435.

6. Fournier JC, Souchon-Lange F, Bismuth C, Gilibert JC, Bernard P, Loe C, Pichot F, Clauvy A, Chavannes C, Vallat L. Acute generalized exanthematous pustulosis: analysis of 83 cases. *Arch Dermatol*. 2007;143(12):1432-1435.

7. Fournier JC, Souchon-Lange F, Bismuth C, Gilibert JC, Bernard P, Loe C, Pichot F, Clauvy A, Chavannes C, Vallat L. Acute generalized exanthematous pustulosis: analysis of 83 cases. *Arch Dermatol*. 2007;143(12):1432-1435.

8. Souchon-Lange F, Fournier JC, Bismuth C, Gilibert JC, Bernard P, Loe C, Pichot F, Clauvy A, Chavannes C, Vallat L. Acute generalized exanthematous pustulosis: analysis of 83 cases. *Arch Dermatol*. 2007;143(12):1432-1435.

9. Souchon-Lange F, Fournier JC, Bismuth C, Gilibert JC, Bernard P, Loe C, Pichot F, Clauvy A, Chavannes C, Vallat L. Acute generalized exanthematous pustulosis: analysis of 83 cases. *Arch Dermatol*. 2007;143(12):1432-1435.