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Eosinophilic Annular Erythema Associated with Churg-Strauss Syndrome

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Dear Editor:

Eosinophilic annular erythema (EAE) is a rare entity of unknown etiology characterized by annular erythematous plaques with tissue eosinophilia[1]. Churg-Strauss syndrome (CSS) is a systemic granulomatous vasculitis characterized by peripheral eosinophilia and the infiltration of eosinophils into systemic organs. Herein, we report an interesting case of EAE associated with CSS.

A 73-year-old man presented with asymptomatic plaques on the trunk first detected 2 days prior. Physical examination revealed well-demarcated, annular and erythematous plaques with slightly elevated borders (Fig. 1). The patient had a medical history of chronic kidney disease and asthma. Histopathological examination revealed exocytosis, basal vacuolar degeneration, and hyperpigmentation in the epidermis and diffuse perivascular and interstitial eosinophil infiltrations in the dermis (Fig. 2). Based on these clinical and histopathological findings, he was diagnosed with EAE. Laboratory findings revealed an eosinophil count of 13,878/mm^3 (56.6% of total white blood cells) and anti-myeloperoxidase antibody (2.00 U/ml; normal range, 0～1.0 U/ml) and proteinase 3 antibody (4.58 U/ml; normal range, 0～1.0 U/ml) positivity. The patient also complained of pain and numbness of the lower extremities. An electrophysiologic nerve study revealed axonal-type sensorimotor polyneuropathy. These findings fulfilled the American College of Rheumatology criteria for the diagnosis of CSS. Intravenous methylprednisolone 10 mg/kg for 3 days was administered with gradual tapering to 0.5 mg/kg over 2 months. The skin lesions resolved and blood eosinophil count returned to normal after 2 months of follow-up.

Our case is unique in that the cutaneous manifestation appeared as well-demarcated, annular and erythematous plaques with slightly elevated borders. Differential diagnoses in our case included erythema annulare centrifugum (EAC), Wells syndrome (WS) and cutaneous manifestations of CSS.

EAC clinically presents as erythematous plaques that expand via peripheral extension with central clearing. Histopathologically, EAC is characterized by sharply demarcated "coat-sleeve" lymphocytes around superficial and deep blood vessels. In this case, histopathological findings of vacuolar degeneration and diffuse perivascular and interstitial eosinophil infiltration ruled out EAC.

WS is characterized by indurated urticarial plaques and pathological findings of tissue eosinophilia and flame

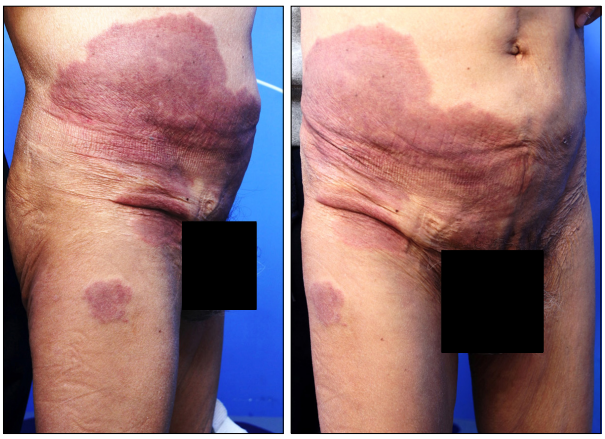

Fig. 1. Well-demarcated, annular and erythematous plaques with slightly elevated borders.

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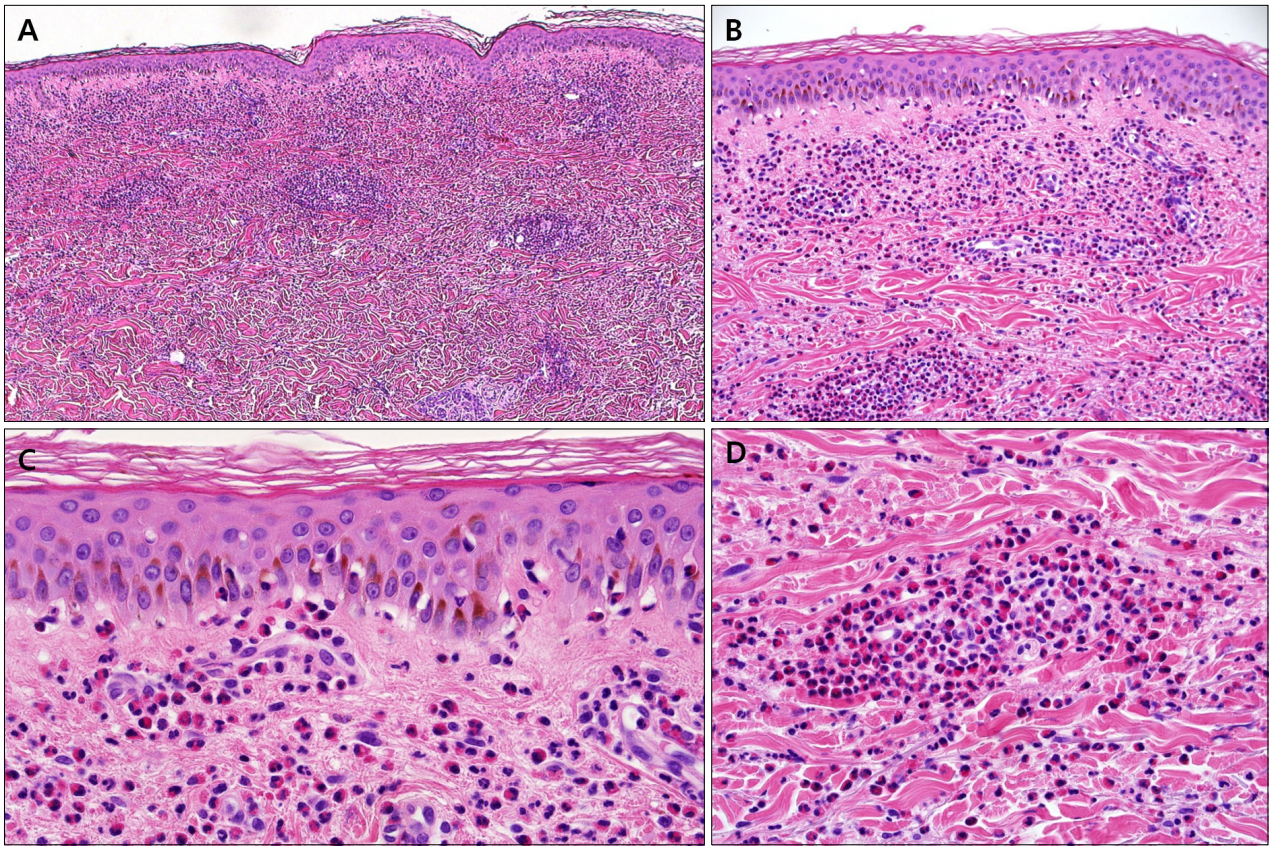

Fig. 2. Histopathological examination revealed exocytosis, basal vacuolar degeneration, and hyperpigmentation in the epidermis and diffuse perivascular and interstitial eosinophil infiltration in the dermis (H&E; A: ×40, B: ×100, C: ×200, D: ×200).

figures. The disease entity of EAE in the spectrum of WS is debateful. El-Khalawany et al.[2] suggested that EAE is a peculiar clinical variant in the WS spectrum. On the other hand, Howes et al.[3] suggested that a subset of EAE cases can be distinguished from WS by pathological findings of basal vacuolar change and dermal mucin, as seen in our case, without eosinophilic dust, flame figures or granulomatous inflammation. Cutaneous manifestations of CSS occurs in 60%～70% of all patients, and classically present as erythematous maculopapules, hemorrhagic lesions, or cutaneous and subcutaneous nodules[4]. The most common histological findings of CSS include extravascular granulomas and leukocytoclastic vasculitis, which were not observed in our case[4].

To the best of our knowledge, this is the first reported case of EAE associated with CSS. Some authors postulated that WS is closely related to CSS and that the two diseases might be part of the same pathogenetic process[5]. Our case will help establish EAE as a clinical entity.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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