QUIZ SECTION

MJS PUBLISHING

Chronic Pruritic Annular Rash in a 47-year-old Man: A Quiz

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A 47-year-old male patient presented with a widespread, intensely pruritic figurate rash for investigation. His medical history was significant for smoking, fibromyalgia, and dyslipidaemia. The rash first appeared more than a decade prior, and followed a relapsing–remitting course, with flare-ups occurring every couple of years and lasting for up to 2 years at a time. Skin biopsy obtained at the time of symptom onset revealed a sparse perivascular lymphocytic infiltrate. Antihistamines and topical steroids provided only temporary symptomatic relief. Additional biopsies obtained during subsequent flare-ups showed superficial and deep perivascular lymphocytic infiltrate, and an eosinophil-rich dermatitis.

The patient underwent comprehensive work-up at another hospital, including an autoantibody panel, fungal cultures, and total-body computed tomography, all of which were unremarkable. Initial skin swab for polymerase chain reaction (PCR) for *Mycobacterium leprae* was positive. The patient was treated with high-dose corticosteroids, dapsone, rifampicin and clofazimine, with transient improvement. Repeated PCRs for leprosy were negative, suggesting a possible initial testing error.

Physical examination revealed polycyclic annular erythematous plaques on the trunk and extremities, with indurated margins and central hyperpigmentation (Fig. 1A). Three 3-mm punch skin biopsies from lesions showed a conserved epidermis and a dermal mononuclear perivascular infiltrate composed of small lymphoid cells, with numerous interstitial eosinophils (Fig. 1B).

What is your diagnosis?

1: Granuloma annulare
2: Erythema annulare centrifugum
3: Chronic urticaria
4: Eosinophilic annular erythema

See next page for answer.

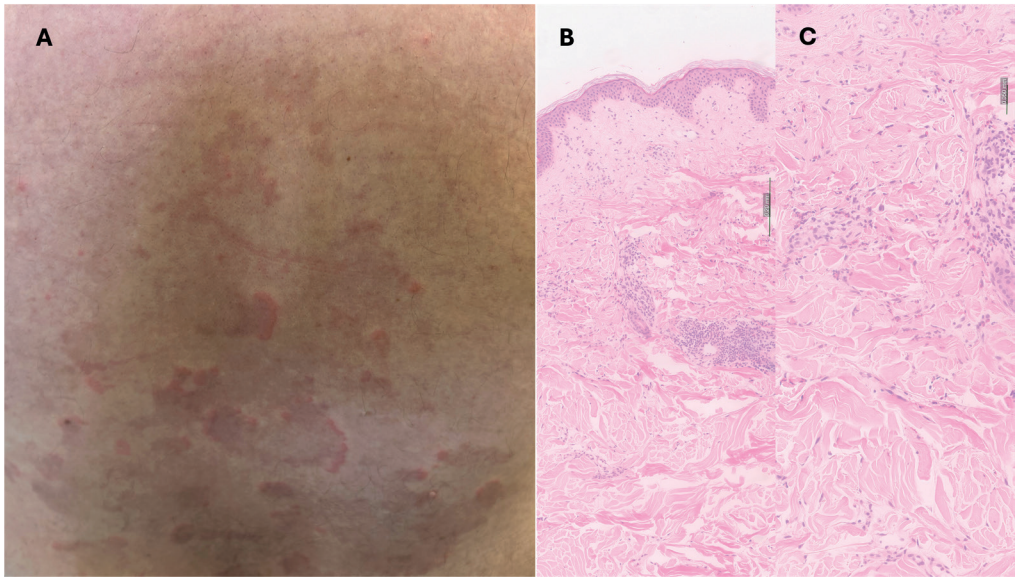

Fig. 1. (A) Polycyclic annular erythematous plaques on the patient's lower back. (B, C) Histopathological examination of a skin biopsy specimen (haematoxylin–eosin) showing a lympho-histiocytic infiltrate and numerous eosinophils, at a low (100X, B) and high (200x, C) magnification.

ANSWERS TO QUIZ

Chronic Pruritic Annular Rash in a 47-year-old Man: A Commentary

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***Diagnosis:* Eosinophilic annular erythema (EAE)**

Eosinophilic annular erythema (EAE) is a rare entity, characterized by recurrent annular erythematous plaques. Less than 100 cases have been reported to date, with a slight female predominance (1). The histological findings of EAE are characterized by a superficial and mid-dermal perivascular/interstitial infiltrate, rich in eosinophils, with an absence of "flame figures" or vasculitis (2). The condition is considered benign, though pruritus and disfiguring widespread lesions can impair quality of life in affected individuals. Several systemic conditions have been reported in conjunction with EAE, including malignancy (1). Paraneoplastic evaluation is therefore warranted upon diagnosis.

There is ongoing debate as to whether EAE represents a form of Wells syndrome (3). While long-standing EAE has been associated with peripheral eosinophilia, in most cases blood count changes were not noted (1). Clinically, EAE lesions are not associated with prodromal burning and resolve without scarring. Histologically, EAE lesions lack the characteristic "flame figures" found in Wells syndrome (4).

EAE lesions may present as wheal-like plaques, requiring differentiation from chronic urticaria. The histological image is similarly composed of dermal perivascular/interstitial lymphocytes and eosinophils. Unlike urticaria, EAE lesions persist for months and do not migrate. EAE is also not associated with angioedema or symptomatic dermographism. Other figurate erythemas include granulomatous disorders such as granuloma annulare and leprosy. Erythema annulare centrifugum (EAC) is a reaction pattern with superficial and deep variants. There is scale in the superficial lesions and raised margins in deep lesions. Marked eosinophilia favours a diagnosis of EAE over EAC. The major differential diagnoses of EAE are summarized in Table I.

No specific therapy is currently approved for EAE. Treatments used in reported cases include corticosteroids, immunosuppressive drugs such as cyclosporine and dapsone, and anti-malarials. Most cases of EAE are persistent and resistant to treatment. In recent years, biological therapy with dupilumab, benralizumab, and mepolizumab was reported as beneficial in several cases (1). One recent case report describes resolution of refractory EAE in a patient treated with tofacitinib, a selective JAK1 and JAK3 inhibitor (5). However, the chronic relapsing clinical course of EAE necessitates long-term follow up and management.

Table I. Main distinguishing features of eosinophilic annular erythema and other figurate erythematous lesions

Entity	Clinical features	Histology
Wells syndrome	Prodrome, morphea-like induration	Flame figures
Granuloma annulare	Usually few lesions, on extremities	Palisading granulomas or mucin deposition
Erythema annulare centrifugum	"Trailing" white scale in superficial form, not pruritic	Superficial and deep perivascular lympho-histiocytic infiltrate
Borderline leprosy	Hypoesthesia, alopecia	Granulomas
Urticaria	Transient pruritic wheals	Perivascular lymphocytes and eosinophils

The patient presented here was treated with UVA1 phototherapy and topical and systemic steroids, without sufficient improvement. Subsequently, oral methotrexate failed to provide symptomatic relief.

The authors have no conflicts of interest to declare.

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