Interstitial granulomatous dermatitis associated with myelofibrosis

Derrick Phillips[1]
Erma Qazi[1]
Su Enn Low[2]
Nitin Khirwadkar[3]
Kok Ngan[4]

Author details can be found at the end of this article

Correspondence to:
Derrick Phillips;
derrickphillips@nhs.net

A 61-year-old woman presented with an 18-month history of weight loss, night sweats and a non-pruritic rash affecting her face, arms and torso (Figure 1). Her medical history included eczema and psoriasis localised to the hands, valve replacement surgery, hypertension and gastro-oesophageal reflux.

Blood tests revealed mild anaemia, significant lymphopenia and a negative autoimmune panel. A computed tomography scan identified hepatosplenomegaly and mesenteric lymphadenopathy. Haematological investigations confirmed a diagnosis of myelofibrosis, and skin biopsies revealed features consistent with interstitial granulomatous dermatitis (Figure 2).

Interstitial granulomatous dermatitis is a rare inflammatory eruption associated with autoimmune diseases (particularly rheumatoid arthritis), as well as haematological and solid organ malignancies (Cases-Merida et al, 2018). It presents with erythematous, indurated papules and plaques that are symmetrically distributed on the trunk and proximal limbs. The 'rope sign' is characterised by linear cord-like plaques affecting the lateral trunk and is pathognomonic (Kim et al, 2017; Takahashi et al, 2017). To the authors' knowledge, this is the first reported case of interstitial granulomatous dermatitis associated with myelofibrosis in the UK.

Author details

[1]Department of Dermatology, Liverpool University Hospitals NHS Foundation Trust, Liverpool, UK

[2]Department of Histopathology, Whiston Hospital, Rainhill, Prescot, UK

[3]Department of Histopathology, Liverpool University Hospitals NHS Foundation Trust, Liverpool, UK

[4]Department of Dermatology, Whiston Hospital, Rainhill, Prescot, UK

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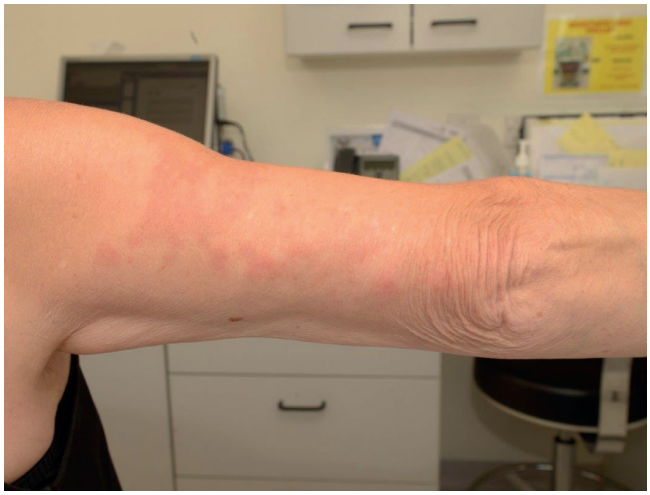

Figure 1. Annular and discoid-looking infiltrative, erythematous plaques suspicious for discoid lupus or Jessner's lymphocytic infiltrate.

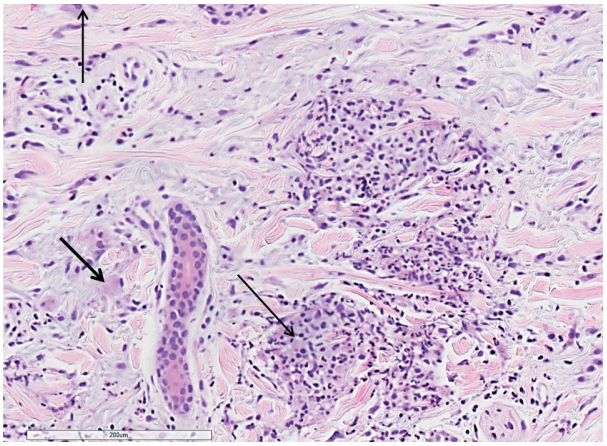

Figure 2. Light microscope image; haematoxylin and eosin stained skin specimen from arm, ×40 magnification. Interstitial histiocytes (black arrows) with admixed polymorphs and focal leukocytoclastic vasculitis, in keeping with interstitial granulomatous dermatitis.