An unusual cause of cutaneous ulceration in association with seronegative rheumatoid arthritis

M Saha[1] • K L Gibbon[1,2] • A S Jawad[3] • R Cerio[1] • V M Jolliffe[1]

[1]Department of Dermatology, Royal London Hospital, London E1 1BB, UK
[2]Whipps Cross Hospital, Leytonstone, London E11 1NR, UK
[3]Department of Rheumatology, Royal London Hospital, London E1 1BB, UK
Correspondence to: Monika Saha. Email: monikasaha@doctors.org.uk

DECLARATIONS

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Palisaded neutrophilic and granulomatous dermatitis is a rare, cutaneous disease, associated with systemic diseases including rheumatoid arthritis and systemic lupus erythematosus.

Case Report

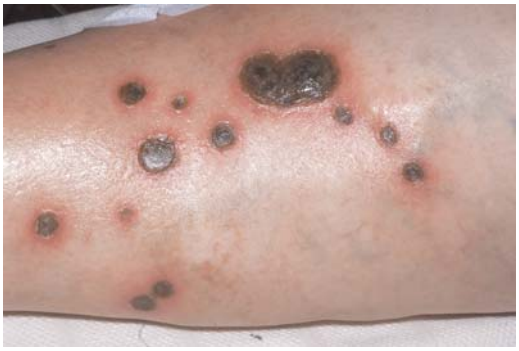
A 43-year-old Indian woman presented with a 6-month history of itchy papules on her forearms and abdomen. She had a 3-year history of seronegative erosive inflammatory polyarthritis affecting predominantly knee and ankle joints, Type 2 diabetes mellitus, hypercholesterolaemia and hypertension. Her medications (all long term) were glicazide, metformin, insulin, orlistat, simvastatin, bisoprolol, ramipril and amitriptyline. The patient had been diagnosed with seronegative rheumatoid arthritis and had been mostly controlled on codeine-based analgesia. In addition, she was treated with methotrexate for 2 months, 6 months prior to the rash, which was stopped due to abnormal liver function tests. Over the ensuing 3 months the lesions gradually became painful, ulcerated and more widespread, spreading to involve the buttocks, trunk and legs. Clinical examination revealed multiple erythematous papules and nodules, some of which had a central crust (Figure 1) whilst others were pustular. Her blood tests revealed a normal full blood count, negative autoimmune screen, rheumatoid factor and dsDNA but mildly raised inflammatory markers; (ESR 28, CRP 17). A skin biopsy from the arm showed ulceration, surface inflammatory crusting and acute on chronic dermal inflammation, with focal necrobiosis and granuloma formation accompanied by a focal neutrophilic infiltrate (Figure 2a,b). Special stains for organisms were all negative (PAS, Gram, ZN and Giemsa). A diagnosis of palisaded neutrophilic and granulomatous dermatitis was made in association with seronegative rheumatoid arthritis. The patient was treated initially, with prednisolone (30 mg), dapsone (50 mg) and dermovate NN cream topically with partial response. The dapsone was increased to 150 mg once daily with further response over the course of a year. The patient remains stable on dapsone 150 mg daily, permitting discontinuation of prednisolone. Lesions have healed with hyperpigmentation and there has been no further skin disease.

Discussion

Palisaded neutrophilic and granulomatous dermatitis is a rare dermatological entity with less than a 100 cases described.[1] It is a heterogenous disease and may represent a number of other conditions which have been previously described including Churg-Strauss granuloma, rheumatoid papule, interstitial granulomatous dermatitis, extravascular necrotizing granuloma and superficial ulcerating rheumatoid necrobiosis. In 1994 Chu *et al.*,[2] proposed the term 'palisaded neutrophilic and granulomatous dermatitis' (PNGD) to encompass these conditions and found an association with immune-mediated diseases.

There is both clinical and histological heterogenicity, depending on the stage of the disease. The diverse clinical presentations include skin-coloured or erythematous papules, nodules, or plaques and may be smooth, umbilicated or rarely ulcerated. Our patient had a dramatic picture of widespread ulceration with tenderness at multiple sites, which is an uncommon presentation. Lesions are often symmetrical and tend to occur on the extremities.

Figure 1
Multiple inflammatory, crusted lesions on the right leg



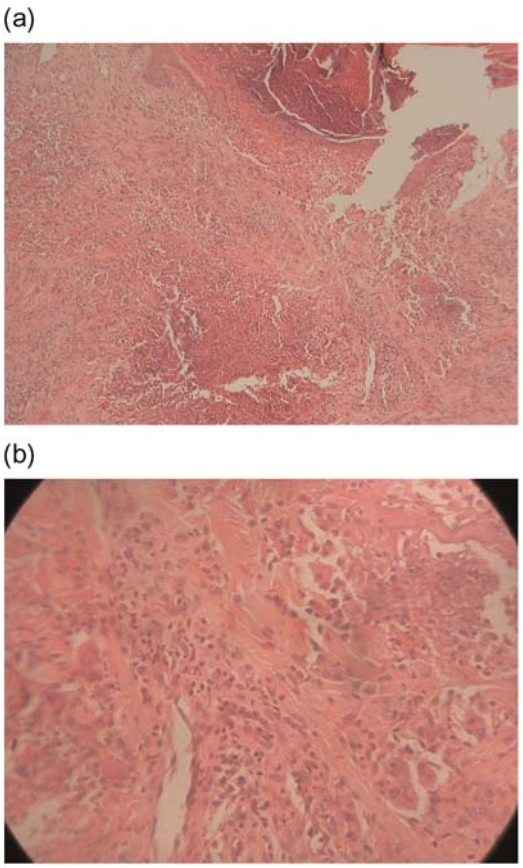
A characteristic and distinctive presentation of firm dermal linear cords on the trunk or neck is well described.[3]

Histologically early lesions show a leucocytoclastic vasculitis, dense neutrophilic infiltration and collagen degeneration. More longstanding lesions demonstrate palisaded granulomas surrounding leucocytoclastic debris, fibrin and altered collagen, whilst endstage lesions reveal palisaded granulomas and dermal fibrosis, with a less marked neutrophilic infiltrate.

Whilst the aetiology of this disease is unknown, there is a recognized association with systemic disease, most commonly immune-mediated diseases, in particular rheumatoid arthritis[4] and systemic lupus erythematosus.[3] PNGD has also been reported in association with lymphoproliferative disease, systemic vasculitides, inflammatory bowel disease and a number of other inflammatory disorders.[1] Our patient presented with a rare clinical presentation of ulcerated PNGD, with an underlying associated seronegative rheumatoid arthritis. This case is a reminder of the recognized association of PNGD and rheumatological disease.[4]

The disease can last from several months to a few years and relapses are not uncommon. Spontaneous resolution can occur in 20% of patients. However, treatment is usually required and topical and systemic steroids can be effective. Dapsone has also been used with success,[5] and other agents used include hydroxychloroquine, cyclosporin, colchicine, cyclophosphamide, non-steroidal anti-inflammatories and mycophenolate mofetil with varying benefit.

Figure 2
A skin biopsy from the arm (a) showing ulceration and an acute on chronic dermal inflammation with focal necrobiosis and granuloma formation (haematoxylin and eosin, original magnification x100) and (b) a neutrophilic infiltrate with necrobiosis (haematoxylin and eosin, original magnification x400)



In summary we present a case of ulcerated cutaneous PNGD in association with seronegative rheumatoid arthritis, highlighting the multiple cutaneous clinical presentations of this entity and reminding the clinician of this association.

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