Pustular ulcer with a blistering border

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A woman develops a rapidly expanding necrotic ulcer over her ankle. What is the cause and how can it be treated?

Case history

Over a three-week period, a 43-year-old woman developed a rapidly expanding necrotic ulcer over her right medial ankle. The ulcer had a pustular and haemorrhagic base and a peripheral dusky blistering edge (Figure 1). Cultures for bacteria and fungi were negative. A skin biopsy from the edge of the lesion showed sheets of neutrophils undergoing leucocytoclasis and filling the dermis (Figure 2). The patient had a known history of Crohn's disease.

Differential diagnosis

Bullous pyoderma may be seen in a range of conditions, including the following.

- **Bacterial infection**, particularly due to staphylococci or streptococci, may present as a bullous impetigo, deeper necrotic lesions (ecthyma) or granulating pustular lesions with sinuses (botryomycosis). Cultures will usually isolate these organisms readily.
- **Halogenoderma**, secondary to iodide- or bromide-containing oral medications, may produce bullous and pyodermatous lesions that are usually multiple. Skin biopsy often shows epithelial hyperplasia as well as eosinophils mixed with neutrophils, but prominent leucocytoclasis is not a feature. Careful review of the patient's drug history, including over-the-counter preparations, is important in reaching a diagnosis.
- **Pustular vasculitis** may present as blistering pustular lesions associated with skin infarction. This may be seen in the setting of rheumatoid disease, Wegener's granulomatosis or Behçet's syndrome. Clinical review for associated features such as palpable purpura or digital infarcts is necessary for diagnosis. Skin biopsy will reveal vascular necrosis with an angiocentric neutrophilia undergoing leucocytoclasis.
- **Pyoderma gangrenosum** (or bullous Sweet's syndrome) is the correct diagnosis. It presents as single or multiple, tender, progressive ulcers, which may resemble an infective process because they are pustular. Initial lesions may appear as a necrotising folliculitis. The edges of the ulcers are often dusky blue–black and undermined. The lesions may be induced by local injury including surgery.

Pyoderma gangrenosum is seen particularly in association with inflammatory bowel disease, rheumatoid or seronegative arthritis, plasma cell dyscrasias and myeloproliferative processes. Bullous lesions may be seen more frequently with leukaemia. Cultures of the skin biopsy specimen may be required to exclude infection, including deep fungal infection. Biopsy is helpful in the early neutrophilic stages and to exclude alternative processes such as vasculitis.

Treatment

For localised lesions, intralesional or topical corticosteroids may be effective. For more widespread lesions, systemic corticosteroids, dapsone, minocycline (Akamin, Minomycin), clofazimine (Lamprene) and cyclosporin (Cysporin, Neoral, Sandimmun) have all been effective in individual cases.

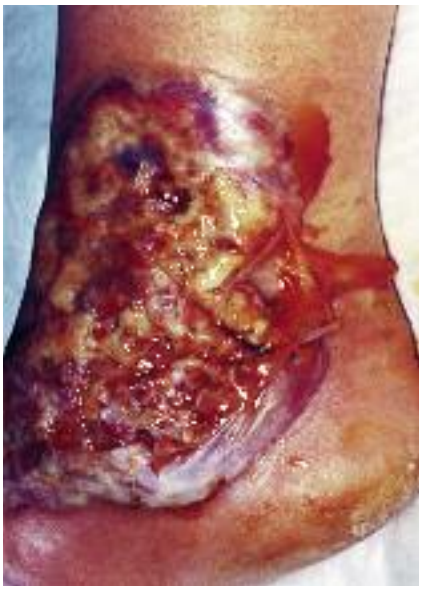

Figure 1. Necrotic and pustular vegetative ulcer with overhanging dusky blistering edge.

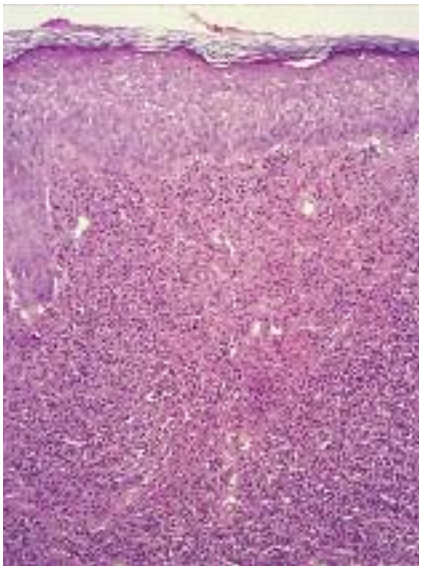

Figure 2. Skin biopsy demonstrating sheets of neutrophils undergoing leucocytoclasis and filling the dermis.

Keypoint

Pyoderma gangrenosum is an unusual distinctive reaction associated with sterile necrotic ulcers, often with a pustular element, that fail to respond to antibiotic or antifungal therapy. It should prompt clinical review for associated systemic diseases. MT

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