Acral blisters with arthritis

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A woman has nonpruritic blisters and fragile skin on the dorsal surfaces of her hands. What is the cause of this blistering and how can it be treated?

Case history

Over a two-month period, a 52-year-old woman developed bilateral nonpruritic blisters (Figure 1) and fragile skin, localised to the dorsal surfaces of her hands. The individual blisters healed with scars. Over the preceding six months, the patient had been taking naproxen for arthritis. She had a background of chronic solar damage. A skin biopsy revealed a subepidermal blister with scant dermal inflammation (Figure 2).

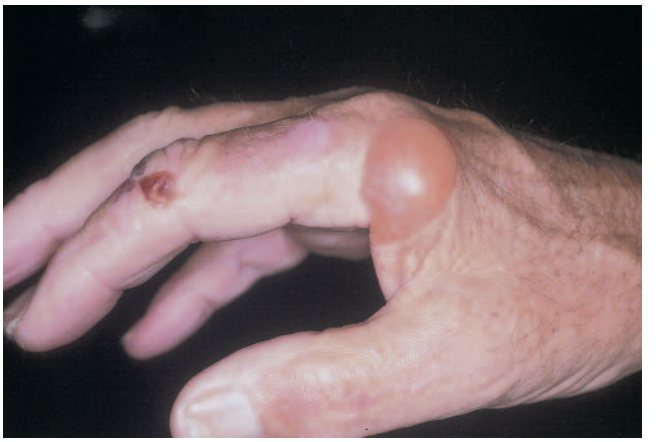

Figure 1. Acral blisters and erosions on the patient's hand.

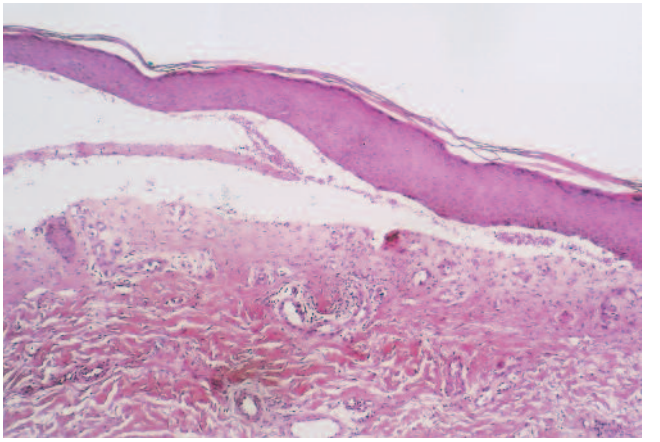

Figure 2. Skin biopsy showing a subepidermal blister with scant dermal inflammation.

Differential diagnosis

The differential diagnosis of acral blisters and skin fragility includes the following conditions.

- **Epidermolysis bullosa** is a heterogeneous group of inherited disorders that are characterised by mutations in adhesion molecules or anchoring fibrils that bind the skin. These disorders usually appear in early life. Mild forms may present as fragile skin with superficial blisters of the feet and hands, particularly the soles and palms. Skin biopsy may show dissolution of basal keratinocytes due to abnormalities in keratin tonofilaments, important in keratinocyte adhesion structures in the basal layer.
- **Porphyria cutanea tarda** presents in an identical clinical and histological pattern to that seen in this case. It is associated with elevated porphyrins produced by the liver, which can be tested by quantitating urinary porphyrins. Porphyria cutanea tarda is aggravated by excess iron and oestrogens.
- **Pseudoporphyria** is the correct diagnosis. It may be induced by a variety of drugs, including naproxen, nalidixic acid, tetracyclines, frusemide and retinoids.

Pseudoporphyria may also be seen in acral areas in individuals who have chronic renal failure, with or without dialysis, and in those with chronic sun damage or chronic UV exposure via tanning beds. A careful drug history and a history of exposure to sunlight or artificial light are important in reaching a diagnosis. Porphyrin studies are required to exclude porphyria cutanea tarda. Naproxen-induced blisters are seen particularly in children and may be associated with pitted angular or linear facial scarring.

Treatment

Removal of the offending agent usually results in slow resolution of the blisters over several months. Sun protection will be required. Other nonsteroidal anti-inflammatory drugs, such as diclofenac, indomethacin or COX-2 inhibitors, may be substituted for naproxen.

Keypoint

Acral blisters and fragile skin can be induced by a variety of drugs. This may be clinically indistinguishable from porphyria cutanea tarda. MT

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