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Dermato-venereological Quiz

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A 43-year-old lady developed multiple blisters on trunk and proximal limbs for six months. The blisters were flaccid and easily ruptured leaving tender erosions on back, chest, abdomen, spreading to arms and thighs (Figures 1 & 2). The mucosal surfaces were not affected. There was no fever, systemic symptoms or malar erythema. The past medical history was unremarkable and she was not taking any drugs before onset of eruption. Skin biopsy was performed and histopathological sections were shown (Figures 3 & 4). ANA 1/80 speckled, anti-DNA and anti-ENA were negative

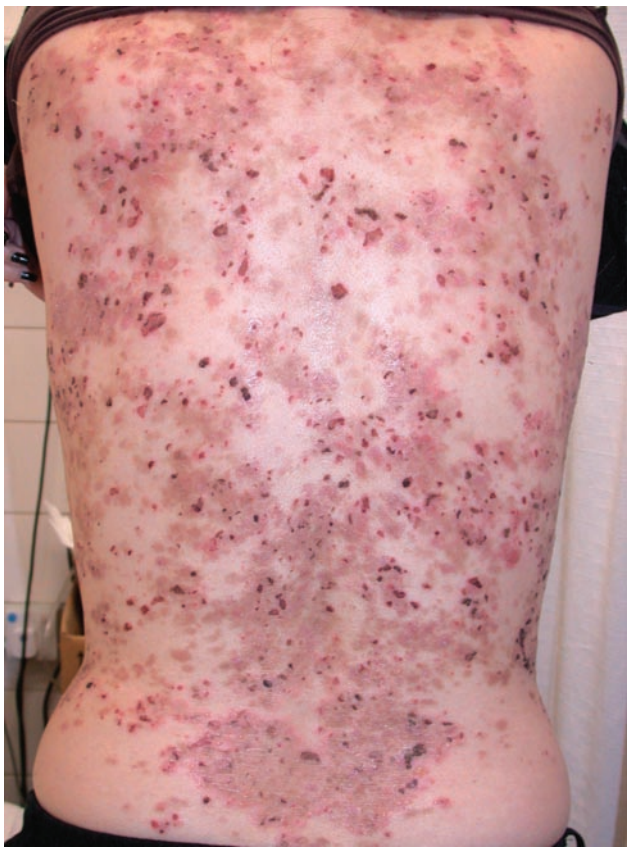

Figure 1. Multiple tender erosions located on trunk, neck and proximal limbs.

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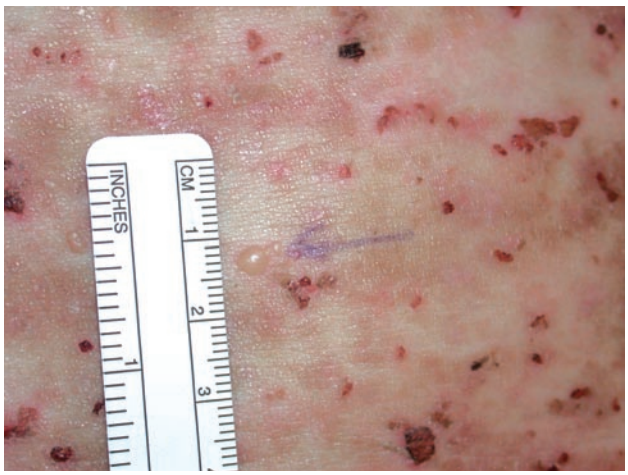

Figure 2. Flaccid blisters that readily ruptured leaving crusted tender erosions.

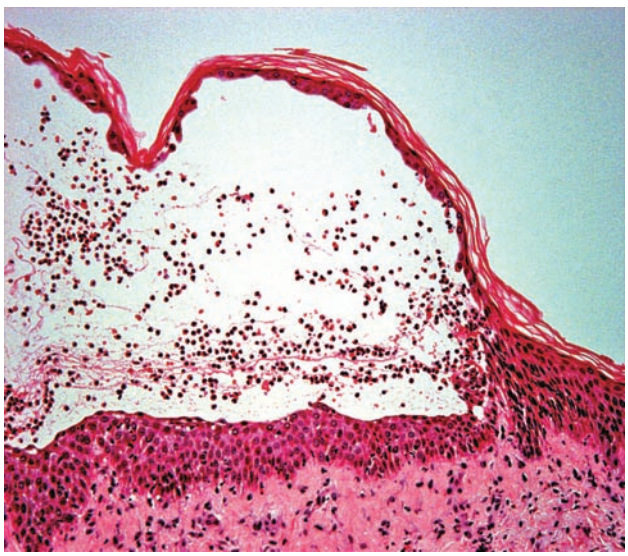

Figure 3. Superficial intra-epidermal bulla. (H&E stain, Original magnification x 4)

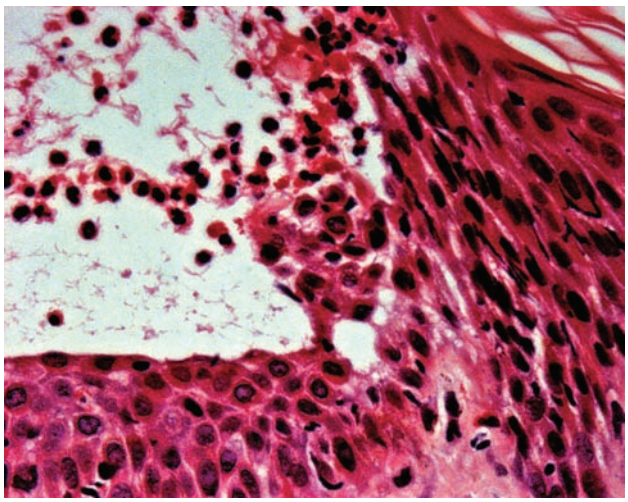

Figure 4. Acantholysis at the base of the bulla. (H&E stain, Original magnification x 40)

Questions

1) What are the differential diagnosis?
2) What are the histopathological features?
3) What is the most likely diagnosis and what investigations would help to confirm it?
4) Is oral mucosal involvement unusual and why?
5) What are the treatment options?

(Answers on page 103)

Answers to Dermato-venereological Quiz on pages 93-94

1) Differential diagnosis include auto-immune intra-epidermal blistering disorders like pemphigus foliaceus or erythematosus, pemphigus vulgaris, and drug induced pemphigus like eruption. Subepidermal immuno-bullous disease like IgA bullous dermatosis, dermatitis herpetiformis, bullous pemphigoid needs to be considered. Skin infections including bullous impetigo and tinea corporis have to be excluded.
2) The skin shows subcorneal bulla, containing a few dyskeratotic granular keratinocytes, eosinophils and neutrophils (Figure 3). There is a superficial perivascular inflammatory cell infiltrate of lymphocytes, a few neutrophils and eosinophils. Acantholysis is noted at the base of bulla (Figure 4). Fungal stains and Gram stain show no micro-organisms.
3) Pemphigus foliaceus. Direct IMF of skin showed positive staining of IgA, IgG and C3 in intercellular spaces but not DEJ. Indirect IMF demonstrated anti-skin antibody of intercellular pattern positive at 1/640 in this case. ELISA would be positive for anti-desmoglein-1 but negative for anti-desmoglein-3 antibodies, however this test is not readily available in our local setting.
4) The target antigen for pemphigus foliaceus, desmoglein-1 is located mainly in the superficial epidermis, resulting in blistering around the granular layer of skin. Oral lesions are unusual for pemphigus foliaceus. Although there is desmoglein-1 in oral epithelium, desmoglein-3 which also present in superficial oral epithelium keeps the cells from detaching.
5) Drug-induced pemphigus like eruption needs to be excluded. Drugs implicated include penicillamine, captopril, and rifampicin. Treatment for pemphigus foliaceus include wound care, antimicrobial treatment for secondary skin infection, topical and systemic steroids, second line agents include azathioprine, cyclophosphamide, cyclosporine or mycophenolate, and third line agents include plasmapheresis, IVIG or rituximab.