A 64 year old woman presented with two weeks of redness, pain, swelling, and exudation of the periungual skin around all fingernails and toenails. Examination found she had complete destruction over the nailfolds, and the periungual skin was erythematous with blisters (fig 1).

She was treated for bacterial and fungal paronychia without any improvement, and the results of fungal microscopy and culture of periungual skin were negative.

Two weeks later, she developed cutaneous blisters/bullae on the trunk, which were thinly walled and relatively flaccid. The rest of the skin became fragile, and light stroking caused the epidermis to peel off. There was no mucous membrane involvement.

After a further week the blistering over the trunk became severe (fig 2).

No mucosal lesions were seen on examination.

Histology from a skin biopsy of the trunk lesions showed acantholysis in the suprabasal layer without spongiosis or exocytosis. The patient declined nail biopsy. Direct immunofluorescence of blister lesions revealed in vivo IgG deposition on the keratinocyte surface in all epidermal layers. Indirect immunofluorescence was negative. Enzyme linked immunosorbert assay results are shown in table 1.

Investigation with serum tumour markers and positron emission tomography revealed no further tumour presence.

**What is the diagnosis?**

**Answer**

Pemphigus vulgaris, confirmed by suprabasal acantholysis without spongiosis or exocytosis on histology, and raised levels of desmoglein-1 and desmoglein-3 antibodies on enzyme linked immunosorbert assay.

Pemphigus vulgaris is an autoimmune blistering disease typically manifesting as cutaneous and mucosal lesions. Mucosal changes are more common than nail changes in pemphigus vulgaris, but not all patients develop them.
Paronychia is the most common nail manifestation of pemphigus vulgaris (present in 13.4–80% cases1,3-5) and can be a sign before pemphigus vulgaris exacerbation or relapse.3-5

Although nail structure is similar to skin and mucosa structure, the number and function of antigen-presenting cells is lower in the nail matrix than in the epidermis,1 which might be why nail changes are not always present in pemphigus vulgaris. Paronychia in pemphigus vulgaris may mimic paronychia caused by bacterial and fungal infections. Microscopy and culture can facilitate diagnosis in such cases. Also, consider acral pustular psoriasis if periungual pustules are present rather than bullae. Nail changes may also be a sign of paraneoplastic pemphigus, which is why tumour presence was excluded. Other signs specific to paraneoplastic pemphigus are painful mucosal erosions, severe intractable stomatitis, and conjunctival involvement, meaning that paraneoplastic pemphigus is unlikely in this case.

Treatment options include rituximab and methylprednisolone.

**Learning points**

Consider pemphigus vulgaris when presented with nail changes.

Consider pemphigus vulgaris when paronychia does not respond to antibiotics or antifungals.

Mucosal lesions are not always present in pemphigus vulgaris.

**Patient outcome**

After three weeks of methylprednisolone (60 mg/day), skin and nail changes improved dramatically, and no new eruptions occurred. Remission was achieved after eight weeks on a gradually reducing dose.

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### Table 1: Results of enzyme linked immunosorbent assay

<table>
<thead>
<tr>
<th>Test</th>
<th>Result (U/mL)</th>
<th>Normal range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum autoantibody level against desmoglein-1</td>
<td>193.3</td>
<td>&lt;20</td>
</tr>
<tr>
<td>Serum autoantibody level against desmoglein-3</td>
<td>124.1</td>
<td>&lt;20</td>
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