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# Clinical Challenges Q&A

## 26. Mouth and Skin Ulcerations

A 25-year-old woman presented with a chronically sore mouth. Her soreness appeared one week after a skin rash appeared on her scalp, buttocks and abdomen. The patient suffered only from osteoarthritis and in crisis used piroxicam tablets. Clinical examination revealed oral malodour (halitosis) associated with a large superficial ulceration in her lower lip (Figure 1), a white coated, partially depapillated tongue, with painful red areas of ulcerations in her dorsum, buccal mucosae and floor of mouth. Together with the oral ulcers, a few large flaccid skin bullae were seen. Biopsy from the tongue near to the ulceration revealed epithelial acantholysis.

### Q1. Which is the most likely diagnosis?

- (a) Herpetic stomatitis;
- (b) Syphilis (second stage);
- (c) Pemphigus vulgaris;
- (d) Darier disease;
- (e) Erythema multiforme.

### A1. The answer to which is the most likely diagnosis?

*Pemphigus vulgaris*, an autoimmune intraepithelial blistering disease

affecting skin and mucous membranes, is the cause of oral and skin ulcerations in this patient. The presence of intact bullae in the mouth is extremely rare as these bullae are very fragile and break easily during mastication, causing painful ulcerations and halitosis, as seen in our patient. The hallmark of *pemphigus* is the formation of intra-epithelial bullae by antibodies against desmoglein (Dsg1 and 3) binding to keratinocyte desmosomes, finally causing acantholysis. Piroxicam, an anti-inflammatory drug, seldom induces acantholysis and may produce similar lesions. Intra-epithelial blistering may also be seen in *Darier disease*, *erythema multiforme* and herpetic infections but their clinical presentations are quite different. Secondary *syphilis* can also be presented with atypical but asymptomatic oral and skin ulcerations.

### Q2. Which investigations are most useful to confirm a pemphigus diagnosis?

- (a) Biopsy;
- (b) Immunofluorescence;
- (c) Serum electrophoresis;
- (d) Tissue culture;
- (e) Allergy tests.

### A2. The answer to which investigations are most useful to confirm a pemphigus diagnosis?

The most useful investigation for the diagnosis of pemphigus is a *biopsy*. This examination shows bullae within the epithelium. Additionally, direct *immunofluorescence* shows intercellular deposits of IgG and C3 in the epithelium while indirect *immunofluorescence* using monkey oesophagus substrate confirms the presence of specific antibodies in the patient's *serum*.



Figure 1. Large ulcer on lower lip.

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