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A Painful, Blistering Rash

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CASE

A 67-year-old man presented with painful oral and skin lesions developing over the last 2 months. The lesions initially formed in his mouth and lips, and slowly spread to his torso, groin, and extremities (Figure 1). The lesions began as blisters that broke easily and were exquisitely painful to light touch. In the past few days, he also developed painful lesions on his eyes that were associated with redness and photophobia. The patient saw his primary care doctor at the onset of illness and failed to improve with courses of azithromycin, ciprofloxacin, and tetracycline. He was no longer able to tolerate anything by mouth because of pain and he had lost a significant amount of weight. His skin exam was notable for Nikolsky's sign.

DIAGNOSIS

Pemphigus vulgaris is an autoimmune blistering disorder mediated by auto-antibodies against epidermal cell antigens.¹ It has an incidence of approximately 1 in 100,000, and the usual age of onset is 40-60 years of age.² Patients present with painful, flaccid bullae that rupture and form erosions (Figure). Shearing stress on skin can lead to development of new erosions (Nikolsky's sign). Oral lesions are the initial symptom in 50-60% patients and may precede cutaneous lesions by months. Diagnosis is confirmed by histology and direct immunofluorescence of peri-lesional skin. Mortality is 70% if untreated, and primarily results from sepsis, fluid loss and malnutrition from oral lesions.³

Our patient was admitted for fluid resuscitation and intravenous steroids, and had a skin biopsy consistent with pemphigus vulgaris.



Figure. Extensive erosions and flaccid bullae associated with autoimmune blistering disorder.

funding sources and financial or management relationships that could be perceived as potential sources of bias. The authors disclosed none.

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