

Desquamative esophagitis due to pemphigus vulgaris

A 48-year-old woman was admitted to our hospital with hematemesis that occurred after dinner. Two years previously, she had been diagnosed as having pemphigus vulgaris on the basis of bullous lesions in the oral mucosa and the presence of anti-desmoglein 3 (Dsg3) antibodies. Esophagogastroduodenoscopy (EGD) revealed desquamation of the mucosa, blood oozing from the entire esophagus (● Fig. 1), and a whitish strip, which was exfoliated esophageal mucosa, hanging loosely from the cardia (● Fig. 2). Prednisolone (30 mg/day) was administered. Two weeks later, EGD showed a dramatic improvement in the esophageal mucosa, which had an almost normal appearance (● Fig. 3).

Pemphigus vulgaris is a rare, autoantibody-mediated, blistering disease of the skin and mucosal membrane. The most frequently involved mucosa is the oral mucosa; oral mucosal lesions occur in nearly all patients during the course of the disease. It was long considered that the esophageal mucosa is rarely involved in pemphigus vulgaris [1]. However, in 1999, Gomi et al. reported that patients with pemphigus vulgaris had a high frequency (87.5%) of esophageal lesions (bullae or exfoliated erosions of bullae); these lesions led to exfoliative esophagitis [2]. Patients with mucosal pemphigus vulgaris have autoantibodies against the desmosomal cadherin Dsg3. These autoantibodies are responsible for loss of cohesion between epidermal cells, resulting in blister formation. Such a high frequency of esophageal involvement may be because Dsg3 is highly expressed in the stratified squamous epithelium of the esophagus as well as that of the oral mucosa [3]. Nikolsky's sign, a well-described clinical sign of pemphigus vulgaris, is positive when manual pressure on the skin of patients elicits the separation of the epidermis. A positive Nikolsky's sign has also been reported on the mucosal membrane of the esophagus and uterine cervix in patients with pemphigus vulgaris [4, 5]. In conclusion, when clinicians encounter idiopathic esophageal

lesions and extensive bleeding from the esophagus, especially with oral mucosal lesions, pemphigus vulgaris should be considered in the differential diagnosis.

Competing interests: None

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Bibliography

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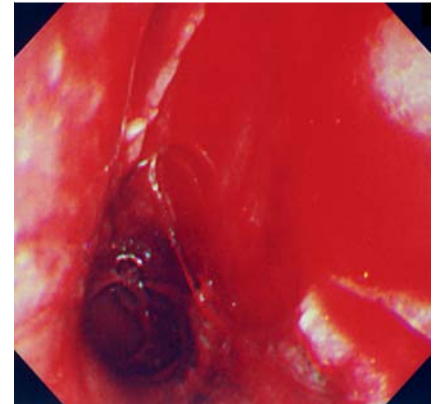


Fig. 1 Esophagogastroduodenoscopy (EGD) showing desquamation of the mucosa and blood oozing from the entire esophagus.

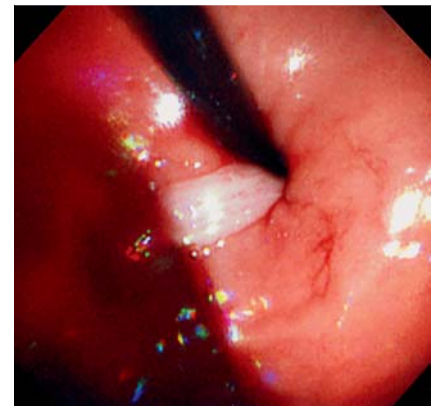


Fig. 2 EGD showing a whitish strip, which is exfoliated esophageal mucosa, hanging loosely from the cardia.

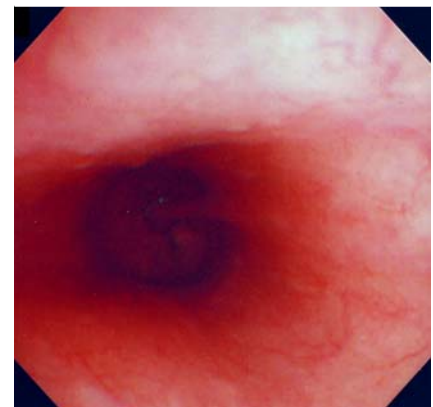


Fig. 3 EGD performed after 2 weeks, showing a dramatic improvement in the esophageal mucosa, which has an almost normal appearance.