

Hemorrhagic bullous esophagitis complicating a rare skin disorder

Ali Raza^a, Maria Hernandez^b, Kashif Ahmed^b, Jonathan Kushner^b

University of Cincinnati Medical Center, Cincinnati, Ohio, USA

A 77-year-old female presented to the hospital with difficulty swallowing for one week. She reported severe retrosternal pain immediately after swallowing food. The patient denied any weight loss or heartburn. Eight weeks prior to presentation, she developed painful skin blisters on her extremities and trunk. Medications included aspirin and amlodipine. Physical examination revealed generalized pruritic eruption of the skin. The patient underwent esophagogastroduodenoscopy (EGD), which revealed severe diffuse esophagitis with nodular surface (Fig. 1). Separation of the superficial epithelial surface from underlying tissue was noted at various locations (Fig. 2). The stomach and duodenum were normal. Biopsies of the esophagus showed fragments of necrosis with marked acute inflammation, without evidence of fungal or viral infection, dysplasia or malignancy. Biopsies of the skin lesions revealed sub-epidermal vesicles with neutrophil predominance. Direct immunofluorescence microscopy revealed intense linear deposition of immunoglobulin G (IgG) along the dermo-epidermal junction. Indirect immunofluorescence study with saline-split skin and patient's serum showed IgG directed against the dermal side only. The diagnosis of epidermolysis bullosa acquisita (EBA) was made and the patient was treated with intravenous steroids and discharged on high dose cyclosporine and proton pump inhibitors. She reported complete resolution of dysphagia at a short-term follow-up.

EBA is an extremely rare autoimmune disorder with the prevalence of 1 case per 5 million people [1]. Esophageal mucosal involvement in EBA is rare and it can be appreciated on EGD and histology as separation of the superficial epithelial layer and may result in ulcers, fibrosis, and stenosis [2].

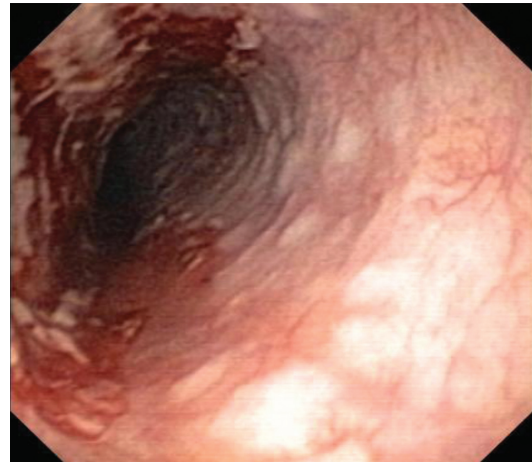


Figure 1 Diffuse esophagitis with edema, ulceration and nodular surface

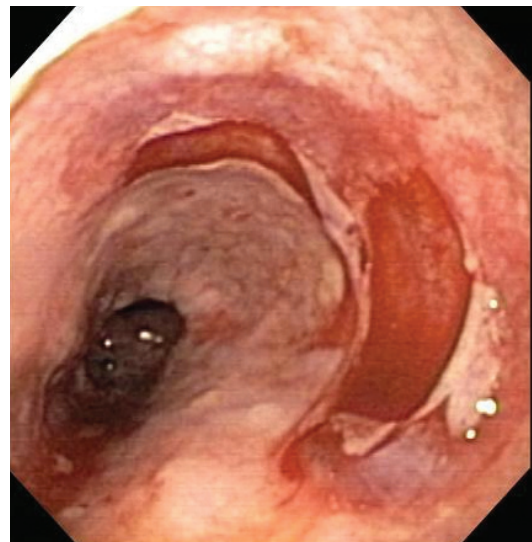


Figure 2 Superficial epithelial layer of esophagus separated from the underlying tissue

^aDepartment of Internal Medicine, University of Cincinnati, Cincinnati, Ohio (Ali Raza);

^bDepartment of Internal Medicine, Division of Digestive Diseases, University of Cincinnati, Cincinnati, Ohio (Maria Hernandez, Kashif Ahmed, Jonathan Kushner)

Conflict of Interest: None

Correspondence to: Ali Raza 231 Albert B. Sabin Way. MSB 6202, University of Cincinnati Medical Center
PO Box 670595, Cincinnati, Ohio 45267-0595, USA
e-mail: razaai@ucmail.uc.edu
Tel: +1 513-558-2595, Fax: +1 513-558-3878

Received 16 November 2011; accepted 8 December 2011

References

1. Ishii N, Hamada T, Dainichi T, et al. Epidermolysis bullosa acquisita: what is new? *J Dermatol* 2010;**37**:220-230.
2. Delgado L, Aoki V, Santi C, et al. Clinical and immunopathological evaluation of epidermolysis bullosa acquisita. *Clin Exp Dermatol* 2011;**36**:12-18.