Bullous and Exfoliative Esophagitis

K Mönkemüller, Marienhospital, Bottrop, Germany, and University of Alabama, Birmingham, AL, USA
C Mel Wilcox, University of Alabama, Birmingham, AL, USA

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Abstract

Bullous pemphygoid is an uncommon cause of atypical esophagitis. Nevertheless, awareness of this condition is important to make a diagnosis and institute appropriate therapy. There are many autoimmune skin disorders that also affect the esophagus, as both skin and esophagus are covered with squamous epithelium. Some of these blister- and bullae-forming diseases are bullous pemphygoid, pemphygus vulgaris, cicatricial pemphygoid, epidermolysis bullosa acqusita, and lichen planus pemphygoides. Herein is presented a patient with bullous pemphygoid, focusing on the endoscopic and histological characteristics of the disease. This article is part of an expert video encyclopedia.

Keywords

Atypical esophagitis; Bullous esophagitis; Exfoliative esophagitis; Lichen planus; Pemphygoid; Pemphygus; Standard endoscopy; Video.

Video Related to this Article

Video available to view or download at doi: 10.1016/S2212-0971(13)70005-0

Technique

Esophagosgastroduodenoscopy (EGD).

Materials

Videoesophagogastorduodenoscope, standard biopsy forceps; Olympus, Tokyo, Japan.

Background and Endoscopic Procedure

Bullous pemphygoid is an uncommon cause of esophagitis. Nevertheless, awareness of this condition is important in order to make a diagnosis and institute appropriate therapy. There are many autoimmune skin disorders that also affect the esophagus, as both skin and esophagus are covered with squamous epithelium. Some of these blister- and bullae-forming diseases include bullous pemphygoid, pemphygus vulgaris, cicatricial pemphygoid, epidermyolysis bullosa acqusita, eosinophilic esophagitis, and lichen planus pemphygoides,

Nonautoimmune diseases presenting with vesicles, bullae, or exfoliative esophagitis include: herpes simplex virus, lichen planus, amyloidosis, eosinophilic esophagitis, esophagitis dissecans superficialis, graft-versus-host disease, and reactions to blood transfusion.

Patients with vesicular and bullous esophagitis present with a wide spectrum of symptoms including odynophagia, dysphagia, thorax pains, hematemesis, burning behind the sternum, and vomiting of esophageal casts of squamous epithelium. Owing to the fibrosing nature of these disorders, patients can develop strictures of the esophagus. The diagnosis of bullous esophagitis is established by a combination of clinical examination, laboratory data, endoscopy, and histology.

On EGD, patients with bullous pemphygoid often have a normal-appearing esophageal mucosa, but on touching this with the endoscope, bulla form rapidly. This may be called the endoscopic 'Nikolsky sign.' Therapy for bullous esophagitis is directed at the specific underlying cause. For autoimmune subepidermal blistering diseases, the therapy of choice is systemic steroids.

Key Learning Points/Tips and Tricks

- Bullous pemphygoid is a rare cause of esophagitis.
- Bullous and vesicular esophagitis can occur in a wide range of disorders, autoimmune and nonautoimmune.
- Careful attention to the history and physical examination are mandatory as systemic disorders affecting the skin can also affect the esophagus.

Complications and Risk Factors

Careful performance of EGD is mandatory as the slightest contact with the esophageal surface results in the formation of bullae.

In addition, patients with bullous or exfoliative esophagitis have a more vulnerable esophageal mucosa, which can predispose to complications such as laceration, bleeding, and perforation.

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Scripted Voiceover

Time (min:sec)	*Voiceover text*
00:00–00.36	A 65-year-old male presented with dysphagia, odynophagia and hematemesis.
	His past medical history was remarkable for chronic renal failure, atherosclerosis, and pemphigoid.
	On skin examination there were several red patches and small fluid-filled blebs on both hands and feet.
	His hemoglobin was decreased and the C-reactive protein was markedly elevated.
00:37–00:50	Esophagogastroduodenoscopy (EGD) disclosed an erythematous and edematous esophageal mucosa with two bloody fluid-filled blebs.
00:51–01:13	More blebs developed upon endoscopic contact with the esophageal mucosa.
	Biopsies of one of these lesions resulted in 'peeling-off' of the mucosa.
01:14–01:22	Histology revealed squamous epithelial detachment from the submucosal layers.
01:23–01:33	There was clear dilation of the intercellular spaces as shown in the periodic-acid Schiff (PAS)-stain.
01:34–01:50	This patient suffered from esophageal involvement from bullous pemphigoid type II, as demonstrated later by the presence of antibodies against the 180 kDa basement membrane antigen.
01:51–02:12	There are many autoimmune skin disorders that also affect the esophagus including bullous pemphigoid, pemphigus vulgaris, cicatricial pemphigoid, epidermolysis bullosa acquisita, and lichen planus pemphigoides.
02:13–02:45	Nonautoimmune diseases presenting with vesicles, bullae or exfoliative esophagitis include: herpes simplex virus, lichen planus, amyloidosis, eosinophilic esophagitis, graft-versus-host disease, reactions to blood transfusion, Behçet's disease, radiation esophagitis, and esophagitis dissecans superficialis.