Esophagitis Dissecans Superficialis with Concomitant Bullous Pemphigoid: A Case Report

Shawn L Shah1*, Joan Paul2, Alicia T Dagrosa3, Erik Jenson4, Zilla Hussain1, Stuart Gordon1 and Michael Shane Chapman2

1Division of Gastroenterology and Hepatology, Dartmouth-Hitchcock Medical Center, Lebanon, New Hampshire, USA
2Division of Surgery, Section of Dermatology, Dartmouth-Hitchcock Medical Center, Lebanon, New Hampshire, USA
3Geisel School of Medicine at Dartmouth, Lebanon, New Hampshire, USA
4Division of Pathology, Dartmouth-Hitchcock Medical Center, Lebanon, New Hampshire, USA

*Corresponding author: Shawn L Shah, Division of Gastroenterology and Hepatology, Dartmouth-Hitchcock Medical Center, Lebanon, New Hampshire, USA, Tel: 603-650-7515, E-mail: ShawnLShah@gmail.com

Abstract

Esophagitis dissecans superficialis (EDS) is a rare endoscopic finding characterized by sloughing of the esophageal mucosal lining, which has been infrequently associated with autoimmune bullous dermatoses. We report a case of a 77-year-old man who presented with diffuse mucocutaneous bullous lesions and odynophagia. Direct immunofluorescence of a skin biopsy confirmed a diagnosis of bullous pemphigoid and esophagogastroduodenoscopy revealed linear fissures and sloughing of the esophageal mucosa consistent with a diagnosis of EDS. With only two previously reported cases of EDS associated with bullous pemphigoid, prompt endoscopic evaluation should be considered in patients with bullous lesions and gastrointestinal symptoms.

Abbreviations

BP: Bullous Pemphigoid; C3: Complement 3; EDS: Esophagitis Dissecans Superficialis; IgG: Immunoglobulin G; MMP: Mucous Membrane Pemphigoid; PPI: Proton Pump Inhibitor

Introduction

Esophagitis dissecans superficialis (EDS) is an exceedingly rare endoscopic finding that is characterized by sloughing of the esophageal mucosal lining [1]. First described by Rosenberg in the late 1800s, EDS can be visualized endoscopically as stripped off or denuded mucosa, vertical furrows or circumferential cracks along the esophagus [2,3]. The clinical presentation tends to be variable, ranging from an absence of symptoms to chest discomfort, dysphagia, odynophagia, and reflux [4,5]. While the clinical significance of EDS is poorly understood, the etiologies associated with the finding are rather diverse and include medications, such as bisphosphonates and nonsteroidal anti-inflammatory drugs, chemical irritants, celiac disease, collagen vascular disease and autoimmune bullous dermatoses [1,2,6,7]. Interestingly, esophageal involvement in vesiculobullous dermatoses is rather uncommon with most reported cases being associated with pemphigus vulgaris and mucus membrane pemphigoid (MMP) [5]. Here we present an unusual case of EDS associated with bullous pemphigoid (BP).

Case

A 77-year-old man with essential hypertension and morbid obesity presented with a 2-month history of diffuse mucocutaneous bullous lesions with recent onset of odynophagia and melena stools that had developed over the week prior to admission. Pertinent home medications included losartan-hydrochlorothiazide, propranolol, and levothyroxine. He reported pruritic blisters that had spread from his wrists to his upper and lower extremities with concomitant involvement of the oral cavity. The patient had presented to an outside emergency department twice during this 2-month period and was empirically treated with a 5-day course of oral prednisone (60 mg daily) each time with nominal improvement.

Upon presentation to our institution, the patient was afebrile with a normal heart rate but with evidence of orthostatic hypotension. Physical exam findings were notable for tense bullae along the wrists and ankles, edematous urticarial papules and plaques with overlying hemorrhagic crust along the abdomen and upper and lower extremities encompassing approximately 30% of the body surface area, superficial erosions along the upper and lower extremities covering approximately 5% of the body surface area, and punctate erosions along the inside of the lower lip and tongue (Figure 1). The patient’s face, back, and genitalia had no pertinent dermatologic findings. Nikolsky and Asboe-Hansen signs were both negative. Laboratory data were notable for a neutrophilic leukocytosis (12,800 cells per microliter), a peripheral eosinophilia (1,600 cells per microliter) and a normocytic anemia with a hemoglobin of 8.8 grams per deciliter.

A biopsy of the perilesional skin under direct immunofluorescence demonstrated linear immunoglobulin G (IgG) and complement 3 (C3) deposition at the dermoepidermal junction (Figure 2:...
with topical triamcinolone 0.1% ointment. On day 4, he was started on an oral prednisone taper (60 mg for 5 days followed by 40 mg for 5 days, then 20 mg for 5 days, and then 10 mg for 5 days). The patient was discharged on hospital day 8 with complete resolution of his odynophagia and melenic stools, as well as marked improvement of his bullous mucocutaneous lesions. He was sent home with instructions to complete the aforementioned oral prednisone taper and continue the PPI. The patient was started on mycophenolate mofetil (500 mg twice daily) as an outpatient and achieved full resolution of his mucocutaneous lesions. A repeat endoscopy was declined by the patient with no further episodes of odynophagia or melenic stools.

Panel D). Additionally, a hematoxylin and eosin-stained sample from an intact bulla on the left volar wrist revealed a subepidermal blister with an eosinophilic infiltrate in the upper dermis (Figure 2: Panel A, Panel B and Panel C). The histopathological findings confirmed a diagnosis of BP. Given the patient’s continued melena, an esophagogastroduodenoscopy was performed, which revealed vertical linear fissures with sloughing of the superficial esophageal mucosa from the proximal to distal esophagus, suggestive of EDS (Figure 3). An esophageal biopsy was deferred given the friability of the mucosa and risk of bleeding. In addition to continuing treatment with a proton pump inhibitor (PPI), the patient was started on intravenous methylprednisolone (1 gram per day for 3 days) along with topical triamcinolone 0.1% ointment. On day 4, he was started on an oral prednisone taper (60 mg for 5 days followed by 40 mg for 5 days, then 20 mg for 5 days, and then 10 mg for 5 days). The patient was discharged on hospital day 8 with complete resolution of his odynophagia and melenic stools, as well as marked improvement of his bullous mucocutaneous lesions. He was sent home with instructions to complete the aforementioned oral prednisone taper and continue the PPI. The patient was started on mycophenolate mofetil (500 mg twice daily) as an outpatient and achieved full resolution of his mucocutaneous lesions. A repeat endoscopy was declined by the patient with no further episodes of odynophagia or melenic stools.
Bullous pemphigoid is an uncommon autoimmune immunobullous disorder that characteristically affects patients over the age of 60 [9]. Pathogenesis of BP involves tissue-bound and circulating autoantibodies directed against BP antigen 180 (BP180, BPAG2, or type XVII collagen) and BP antigen 230 (BP230 or BPAG1). Autoreactive T-cells then respond to BP180 and BP230 by stimulating BPAG2, or type XVII collagen) and BP antigen 230 (BP230 or BPAG1). This leads to the development of subepidermal blisters.

Discussion

The esophageal squamous epithelium consists of a dynamic cell population that is incessantly renewed; however, complete denuding of the esophageal mucosal is described as an entity known as esophagitis dissecans superficialis (EDS). EDS is primarily an endoscopic finding with no existing diagnostic criteria, although histopathologic features include marked parakeratosis and orthokeratosis, desquamation of the squamous layer, and epithelial inflammation and necrosis [1]. In their clinicopathological study of 12 cases of EDS, Carmack et al. noted the presence of suprabasal acantholysis, which is not typically seen in esophageal biopsies, as samples do not normally include the basement membrane secondary to the tangential position of the endoscope and forceps inside the esophagus [5,7,8]. Hokama et al. reported that enzyme-linked immunosorbent assay of these samples could be used to detect characteristic antibodies in a simpler and more quantifiable manner linked immunosorbent assay of these samples could be used to detect characteristic antibodies in a simpler and more quantifiable manner.

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Foroozan et al. reported the first case of EDS associated with mucous membrane pemphigoid (MMP) in 1967 [10]. Mucous membrane pemphigoid, or cicatricial pemphigoid, is a rare autoimmune bullous disease related to, but distinct from BP. Mucous membrane pemphigoid characteristically affects the oral and ocular mucosa with infrequent skin involvement, and is treated with dapsone rather than systemic corticosteroids as the mainstay of therapy. Esophageal involvement occurs in 4-8% of patients with MMP and typically involves mucosal erosions, webs, and strictures of the esophageal epithelium, as described in a study of 28 patients by Galloro et al. and colleagues argue that a successful biopsy can be safely achieved with a rocking biopsy forceps, as demonstrated in a study of 16 patients with immunobullous disease [8]. However, larger studies must be conducted as many consider endoscopic manipulation of the esophageal lesions were reported [4]. However, Galloro and colleagues argue that a successful biopsy can be safely achieved with a rocking biopsy forceps, as demonstrated in a study of 16 patients with immunobullous disease [8]. Indeed, in a case series of 28 patients with oral pemphigus vulgaris, no endoscopy-related complications manipulating the esophageal lesions were reported [4]. However, larger studies must be conducted as many consider endoscopic biopsies of such lesions to be risky given the extreme fragility of the esophageal mucosa with potential to aggravate existing lesions.

To date, there have been only two reported cases of EDS...
associated with BP (Table 1) [5,14]. Esophageal bullae or erosions have been reported with BP, but EDS or sloughing esophagus is an underreported and under-recognized entity [15-21].

Our patient experienced complete remission of his mucocutaneous lesions with systemic steroids and mycophenolate mofetil; however, he declined repeat endoscopy after his skin lesions resolved. Interestingly, the natural course of EDS appears benign despite its often impressive appearance under endoscopy. Most cases completely resolve after treatment of the underlying condition (e.g., autoimmune bullous dermatoses) in combination with a PPI [22]. However, since esophageal involvement can persist despite apparent clearance of skin lesions whilst on immunosuppressive therapy, EDS should be on the differential for patients presenting with melena and a history of an autoimmune blistering disorder [5]. Esophageal lesions can also be the presenting sign of bullous dermatoses, sometimes occurring months to years before the onset of the characteristic cutaneous lesions of BP or pemphigus vulgaris [23]. While only two prior cases of EDS associated with BP have been described in the English literature, prompt endoscopic evaluation should be considered among patients with bullous lesions and associated gastrointestinal symptoms.

References