CASE REPORT

DOI: 10.4274/cmj.galenos.2024.96268
J Cau Med Sci 2024;2(3):34-35

Esophageal Involvement and Bleeding in Bullous Pemphigoid Disease

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ABSTRACT

Bullous pemphigoid (BP) is the most common autoimmune bullous disorder and is characterized by autoantibodies against hemidesmosomal proteins in the skin and mucous membranes. BP typically presents as large, fluid-filled blisters on normal skin or red, enflamed areas of skin, mainly in the armpits, lower abdomen, inner thighs, and groin. Blisters on the lining of the oral and pharyngeal mucosa occur in up to 35% of cases, but blisters in the esophagus are rare, especially when there are no oral blisters. We report a case of newly diagnosed BP in a 48-year-old woman. She was admitted to our hospital with the sudden onset of hematemesis. This case highlights the importance of gastroenterologists who are cognizant of the potential association between skin illnesses and digestive disorders. It is important to exercise caution during endoscopic procedures in patients with pemphigoid disorders, especially in the absence of apparent symptoms.

Keywords: Bullous pemphigoid, hematemesis, endoscopy

INTRODUCTION

Bullous pemphigoid (BP) is the most common autoimmune bullous disorder and is characterized by autoantibodies against hemidesmosomal proteins in the skin and mucous membranes [1]. The incidence of BP, which is more common in older adults, is 2.5-42.8 cases per million per year, and its occurrence is gradually increasing due to longer life expectancy [2]. BP typically presents as large, fluid-filled blisters on normal skin or red, enflamed areas of skin, mainly in the armpits, lower abdomen, inner thighs, and groin. Blisters on the lining of the oral and pharyngeal mucosa occur in up to 35% of cases; however, blisters in the esophagus are rare, especially when there are no oral blisters. Patients with esophageal involvement may or may not have symptoms such as chest pain, difficulty swallowing, and pain during swallowing. Upper endoscopy may reveal blisters and necrosis in the esophagus [3].

CASE PRESENTATION

We report a case of newly diagnosed BP in a 48-year-old woman with a medical history of hypertension. Five months prior, she had been taking a penicillin group antibiotic for a dental enfection, after which bullous lesions appeared on her body. A skin biopsy confirmed the diagnosis of BP, and prednisolone at 1 mg/kg was started. During follow-up, the lesions regressed with oral prednisolone therapy. However, two months ago, she experienced an ischemic cerebrovascular event due to thrombosis in the truncus brachiocephalicus, resulting in left hemiplegia. Consequently, while continuing prednisolone treatment, additional acetylsalicylic acid therapy was initiated.

The patient was admitted to our hospital with the sudden onset of hematemesis. Urgent upper endoscopy revealed multiple blister lesions in the hypopharynx, a significant bleeding esophageal hematoma, and active esophageal bleeding (Figure 1). The procedure was completed by applying an Ankaferd blood stopper to the mucosal lesions where leakage-type bleeding was observed. In a follow-up endoscopy performed under sedation after two days of restricted oral intake and proton pump inhibitor infusion, it was noted that the bleeding in the mucosal lesions had ceased, and the edema had subsided (Figure 2). The patient, whose bleeding was controlled, was started on azathioprine treatment, as recommended by the dermatology department.

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Received: 25.08.2024 **Accepted:** 11.11.2024

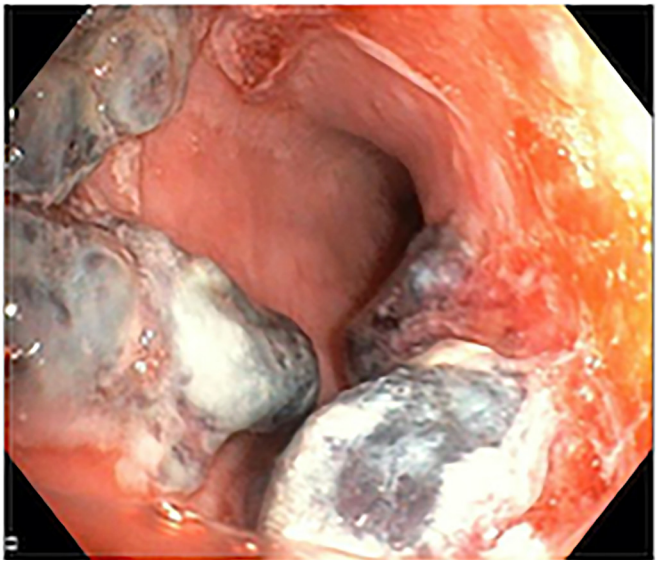

Figure 1. Esophageal hemotamas

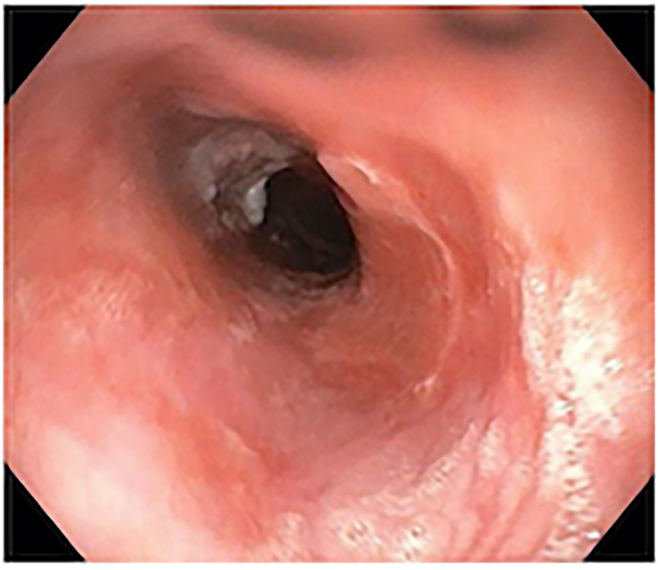

Figure 2. The esophagus after treatment

DISCUSSION

BP is the most common autoimmune disease causing blisters on the skin. It is becoming more common in older adults. BP typically presents as itchy, inflamed skin covered with blisters. However, the appearance of the blisters can vary greatly, and sometimes there are no blisters. Therefore, BP should be considered in any older adult with itchy, enflamed skin [4].

This is a rare case of a patient with a unique gastrointestinal symptom related to skin disease. Pemphigoid disorders, including BP and mucous membrane pemphigoid, are a very rare group of autoimmune skin diseases. These lesions are caused by antibodies that attack the basement membrane of the squamous epithelium, triggering enflammation.

It is noteworthy that the gastrointestinal symptoms exhibited by the patient were not consistent with the observations made during the endoscopic examination. The patient did not exhibit any gastrointestinal problems. Performing esophagogastroduodenoscopy can be challenging in such situations, as the esophagus has the potential to develop blisters and undergo sloughing even with minimal contact from the endoscope. It is advisable to employ a cautious and deliberate approach when manipulating the endoscope to minimize the risk of hemorrhage, lacerations, and perforation. The primary objective of treatment is to effectively manage the underlying immunological disorders. Systemic corticosteroids are commonly used as the initial therapeutic approach to induce remission. However, in cases of extensive gastrointestinal bleeding resulting in hemodynamic instability, therapeutic endoscopy may be used as an intervention to halt the bleeding.

This case underscores the importance of gastroenterologists who are aware of the possible association between skin diseases and gastrointestinal symptoms. Caution should be exercised during endoscopic procedures in patients with pemphigoid disorders, even in the absence of overt symptoms.

Ethics

Informed Consent: Written informed consent was obtained from the patient before procedure.

Footnotes

Authorship Contributions

Surgical and Medical Practices: D.A., Concept: D.A., Design: M.C.Y., Data Collection or Processing: K.K., Analysis or Interpretation: D.A., Literature Search: K.K., M.C.Y., Writing: K.K., D.A.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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