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Case Report

Esophageal Involvement of Pemphigus Vulgaris

Vedat Goral[1*], Ali Balevi[2], Sevilay Aligulu[3] and Rukiye Nilgun Erdogan[4]

[1]*Department of Gastroenterology, Istanbul Medipol University School of Medicine, Istanbul, Turkey*
[2]*Department of Dermatology, Istanbul Medipol University School of Medicine, Istanbul, Turkey*
[3]*Medical Student, Istanbul Medipol University School of Medicine, Istanbul, Turkey*
[4]*Department of Pathology, Istanbul Medipol University School of Medicine, Istanbul, Turkey*

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ABSTRACT

Pemphigus is an autoimmune and life-threatening disease. Esophageal involvement of pemphigus vulgaris is rare. The most common presenting symptoms are odynophagia and dysphagia. Here, I present one cases of pemphigus vulgaris presenting with dysphagia because of esophageal involvement of the disease. In case, a 27-year-old female patient with a prior diagnosis of pemphigus vulgaris presented with dysphagia. Esophagogastroduodenoscopy showed diffuse mucosal exfoliation and oozing bleeding of the esophagus. The patient recovered after the administration of corticosteroids, pantoprazole and azathioprurine.

Introduction

Pemphigus is an autoimmune and life-threatening disease characterized by acantholysis (loss of keratinocyte-keratinocyte adhesion) caused by acantholysis (loss of keratinocyte-keratinocyte adhesion) caused by the mucous membrane and skin, directed against keratinocyte cell surface [1]. The main subgroups of Pemphigus are Pemphigus Vulgaris (PV), Pemphigus Foliaseus and Paraneoplastic Pemphigus. The most common subtype of pemphigus is PV [2-5]. PV is seen equally in both sexes. Although it differs geographically, its incidence varies between 0.1 and 0.5 per 100.000 per year [6-10]. In almost all cases during the course of the disease, the multi-layer flat epithelial mucosa, especially the oral mucosa, is involved. Pharyngolaryngeal, conjunctival, genital, anorectal mucosa are less often involved.

Odynophagia and dysphagia are the most common symptoms of esophageal involvement. However, patients with esophageal involvement can sometimes be asymptomatic. It is thought that many esophageal involvement cannot be detected because endoscopy is not performed mostly in patients without esophageal complaints. Some immunohistopathological studies show that esophageal involvement is higher than previous reports [2-4].

Here, I present one cases of pemphigus vulgaris presenting with dysphagia because of esophageal involvement of the disease. In case , a 27-year-old female patient with a prior diagnosis of pemphigus vulgaris presented with dysphagia. Esophagogastroduodenoscopy showed diffuse mucosal exfoliation and oozing bleeding of the esophagus. The patient recovered after the administration of corticosteroids, pantoprazole and azathioprurine.

Case Report

A 32-year-old woman complained about difficulty swallowing and pain in her chest about 3 months ago. The patient applied to our gastroenterology outpatient clinic with his current complaints and an upper gastrointestinal system endoscopy was performed. In the esophageal mucosa, an appearance that started around 20 cm, was circular, widespread, erosional, hemorrhagic and slightly narrowing in the lower region. Two biopsies were taken from here. The esophageal lumen was naturally observed. The Z line was crossed at 40.cm from the front incisors. At the pathology report, although no specific findings are seen in the DIF examination, it is rare with focal C3 accumulation and immunohistochemical examination. C4d positivity suggests an immunological mechanism. Intraepidermal suprabasal dissociation supports the diagnosis of pemphigus vulgaris.

[*]*Correspondence to: Professor Dr. Vedat Goral, Department of Gastroenterology, Istanbul Medipol University School of Medicine, Istanbul, Turkey; E-mail: vedat.goral@medipol.com.tr; vegoral@hotmail.com*

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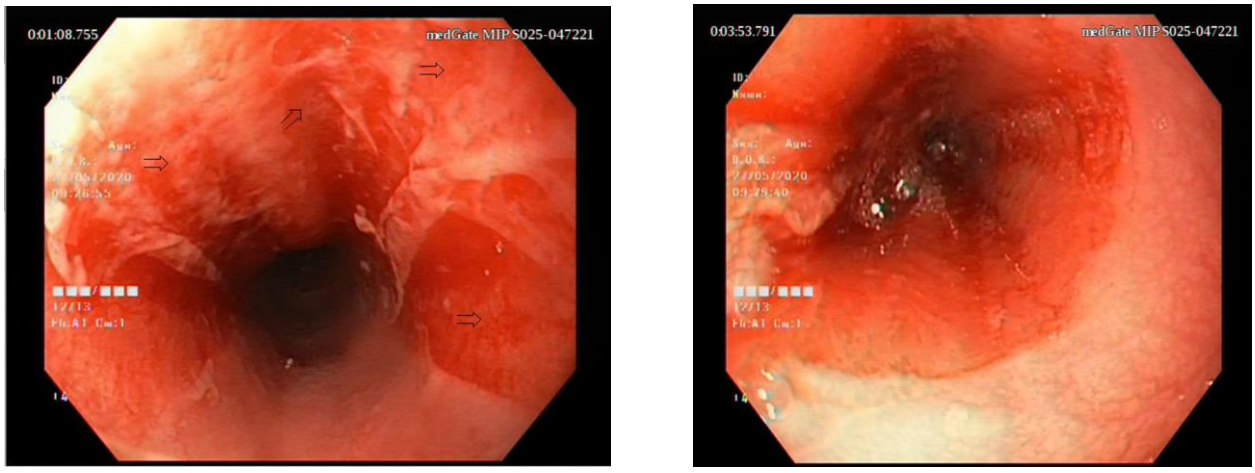
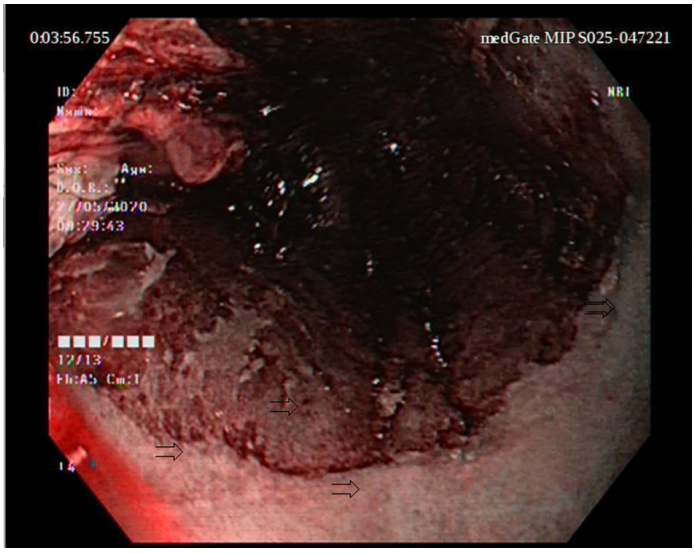
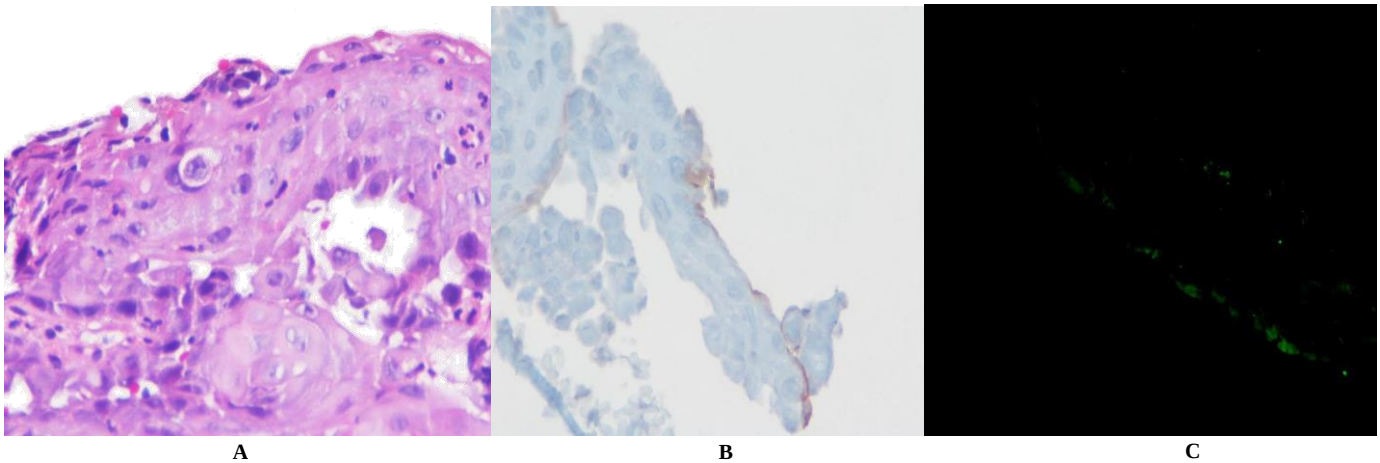


A



B

Figure 1: A) Endoscopic view of esophagus, showing erosions and sheets of sloughed mucosa (arrows). **B)** Endoscopic (Narrow Band Imaging-NBI) view of esophagus, showing erosions and sheets of sloughed mucosa (arrows).



A **B** **C**

Figure 2: A) Intraepithelial dissociation and acantholytic cells (H-E, x100). **B)** Accumulation surrounding squamous cells with anti-C4d antibody in immunohistochemical examination. **C)** C3 accumulation with DIF.

A biopsy was taken by monitoring vesicle-nodular lesions in the mid-esophagus (Figure 1a & 1b). Starting from the biopsy site, it was observed that the esophageal mucosa was separated from the underlying tissue enveloping the esophageal lumen all around (Figure 2). Pathology report: Although no specific findings are seen in the DIF examination, it is seen rare with focal C3 accumulation and immunohistochemical examination. C4d positivity suggests an immunological mechanism. Intraepidermal suprabasal dissociation evaluated in favor of pemhigus vulgaris.

Discussion

The incidence of PV varies from 0.5 to 3.2 cases per 100,000, with an approximately equal male-to-female ratio.6 PV is a potentially life-threatening disease, with a mortality rate of approximately 5–15%. Complications secondary to the use of high-dose corticosteroids contribute to the mortality rate. Morbidity and mortality are related to the extent of disease, the maximum dose of systemic steroids required, and the presence of other diseases. Prognosis is worse in older patients with extensive disease. The pharyngo-laryngeal, genital, anorectal, and conjunctival mucosa are less commonly involved. Odynophagia and dysphagia are the usual complaints in patients with esophageal pemphigus.

Systemic corticosteroid therapy is effective at reducing or eliminating the clinical manifestations of PV, with as high as 400 mg of prednisone being administrated daily for patients with severe involvement. Oral or intravenous administration of cyclophosphamide, azathioprine, cyclosporine, and methotrexate may allow the reduction of corticosteroid dosage. Even with immunosuppressive therapy, up to 10% of patients die from their disease, because of electrolyte loss, wound infection, or treatment complications [10-14].

The patient was started on proton pump inhibitor and sucralfate therapy. Steroid and azathiopurine treatment was started with the recommendation of the dermatology clinic. The patient's complaints of swallowing difficulties disappeared, the patient is under pemphigus and his treatment is still ongoing. The patient recovered after the administration of corticosteroids, pantoprazole and azathioprurine.

As conclusion; pemphigus is an autoimmune and life-threatening disease. Esophageal involvement of pemphigus vulgaris is rare. The most common presenting symptoms are odynophagia and dysphagia. If the patient with pemphigus has dysphagia, endoscopy should be done.

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