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Souhir Monastiri *

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Case Report

Case Report Bullous Pemphigoid with Esophageal Involvement

Souhir Monastiri

Department of Gastroenterology at Sahloul University Hospital in Tunisia.

*Corresponding Author: Souhir Monastiri, Department of Gastroenterology at Sahloul University Hospital in Tunisia.

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Abstract:

Bullous pemphigoid (BP) is the most common chronic acquired bullous dermatosis that affects mainly elderly subjects. This condition is characterized by the presence of antibodies directed against the dermal-epidermal junction. It invades the skin and generally respects the other mucous membranes. The discovery of bullae in the esophageal mucosa is exceptional. We report an observation of PB with esophageal bullosis discovered in a 47-year-old patient following acute dysphagia.

Key words: bullous pemphigoid; cytomegalovirus; herpes virus

Introduction:

Bullous pemphigoid (BP) is a bullous dermatosis that may involve the oral mucosa in less than 20% of cases [1, 2] and usually respects other mucous membranes. The description of bullae in the esophageal mucosa is exceptional in BP [3-4] and remains unknown to gastroenterologists. The association with other digestive pathologies, in particular ulceration, is possible. We report an observation of PB with esophageal bullosa.

Observation

This is a 47-year-old patient with no notable pathological history, who presented in August 2022, a pruritic erythematobullous rash disseminated on the back, abdomen and seat associated with ulcerated oral lesions. In addition, acute and progressive painful dysphagia is noted, followed by an episode of hematemesis of small volume. The clinical examination shows tense, non-confluent skin bullae with clear fluid and resting on an erythematous epidermis (figure 1). In the oral cavity, the jugal surfaces and the soft palate are the site of multiple bullae and hemorrhagic ulcerations (Figure 2). The general condition was preserved and the rest of the somatic examination was unremarkable, notably there was no

ocular or genital involvement. The biological examinations were normal. Viral serologies were negative, especially for a recent infection with herpes virus (HSV) and cytomegalovirus (CMV). Skin biopsies were taken and the histological study showed a subepidermal detachment with perivascular eosinophilic infiltrates. Direct cutaneous immunofluorescence shows immunoglobulin G (IgG) and complement (C3) deposits at the dermal-epidermal junction [6]. The oeso-gastroduodenal endoscopy shows the presence of multiple bullae in the lower 1/3 of the oesophagus, resting on an erythematous and haemorrhagic mucosa (Figure 3) with local ulcerations [5]. The bullae are fragile, collapsing on insufflation and rupturing on passage of the fiberscope (Figure 4), leaving a clear or serum fluid. The bacteriological and mycological histological study of the biopsy specimens verifies the absence of mycosis, virus-induced lesions, tumor infiltration, epithelial atypia or esophageal tuberculosis. The patient was put on 0.5 mg/kg/day of oral corticosteroids and dermocorticoids for 3 weeks, followed by a gradual reduction. The evolution is marked by a rapid improvement of the cutaneous and esophageal lesions without sequelae stenosis.



Figure 1: Tense, non-confluent skin bullae with clear liquid on an erythematous epidermis.



Figure 2: Oral ulcerations.

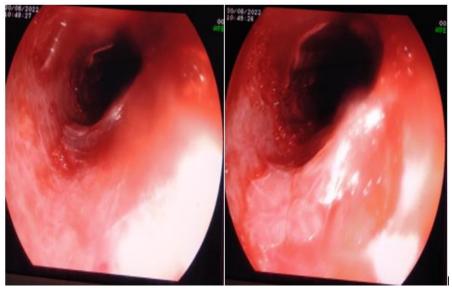


Figure 3: esophageal bullae.

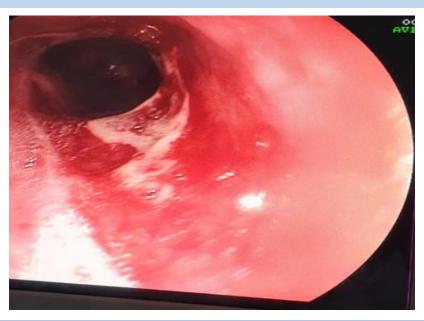


Figure 4: fragile esophageal bullae.

Conclusion:

Esophageal involvement in bullous pemphigoid appears to be underestimated, asymptomatic forms may go unnoticed. An upper endoscopy should be performed systematically in the presence of any PB, not only to detect esophageal bullosa, but also to look for an associated digestive pathology.

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