A rare case of angina bullosa hemorrhagica of the esophagus

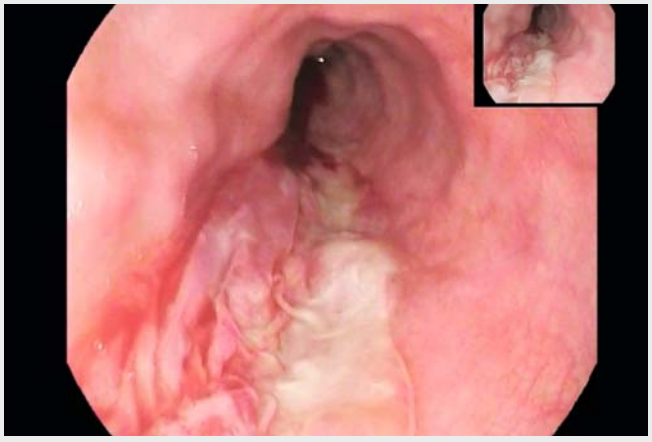
We report the case of a 52-year-old woman referred for esophagogastroduodenoscopy (EGD) because of a previous episode of transient acute chest pain with a negative cardiology work-up. EGD revealed a 15-mm hemorrhagic purple – red tense bulla in the thoracic esophagus at 26 cm from the incisors. The blister broke following passage of the endoscope, rapidly exposing the submucosal layer and causing transient bleeding (▶ **Video 1**; ▶ **Fig. 1**).

The patient's medical history was negative, except for a recent onset of multiple purple – red tense bullae of 1 cm in diameter involving the oral mucosa and tongue (▶ **Fig. 2**). Initially painless, the oral bullae became progressively more painful and ruptured with self-limiting bleeding. The patient underwent routine haematological investigations to rule out any bleeding disorder, followed by repeat EGD with a targeted biopsy. Direct immunofluorescence with specific anti-human IgG, IgM, IgA, and C3, and indirect immunofluorescence of the serum were negative.

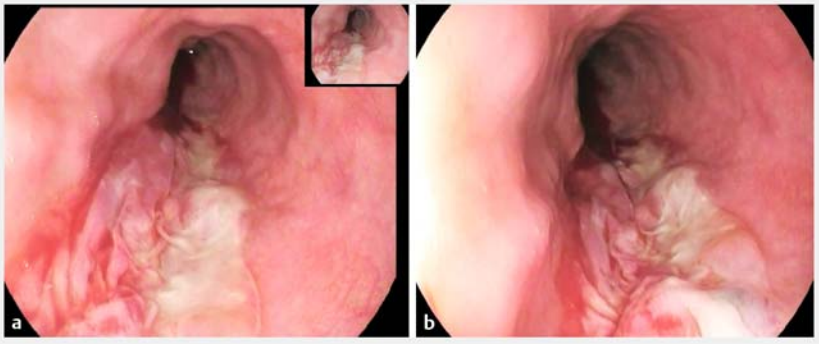
The clinical differential diagnosis included autoimmune blistering disorders, such as bullous lichen planus, mucous membrane pemphigoid, epidermolysis bullosa, and angina bullosa hemorrhagica (ABH). The patient's history, the spontaneous onset of symptoms, the presence of blood in the blisters of the oral mucosa, and the histopathologic findings were consistent with the diagnosis of ABH.

ABH is a rare benign condition, first described in 1933 by Dalina as traumatic oral hemophlyctenosis, characterized by sudden onset of blood-filled blisters in the oral cavity that burst without sequelae [1, 2]. The blisters are usually restricted to the soft palate; pharyngeal and esophageal blisters have rarely been described [3]. ABH is more common in women, with a peak incidence beyond the fifth decade. Its etiology remains unclear. ABH has been associated with a constitutional predisposition causing a weak anchorage of mucosal vessels. Masticatory trauma or dental procedures have been suggested as a precipitating factor [4, 5].

Esophageal ABH revealed by EGD has not been described before. Owing to the potential risk of rapidly expanding blood-filled bullae in the pharynx and the esophagus, it is especially important for gastroenterologists to be aware of this condition while performing upper gastrointestinal endoscopy.



Video 1 Angina bullosa hemorrhagica of the esophagus.



▶ **Fig. 1** Endoscopic images showing: **a** rupture of the esophageal blister following passage of the endoscope; **b** the exposed esophageal submucosal layer.

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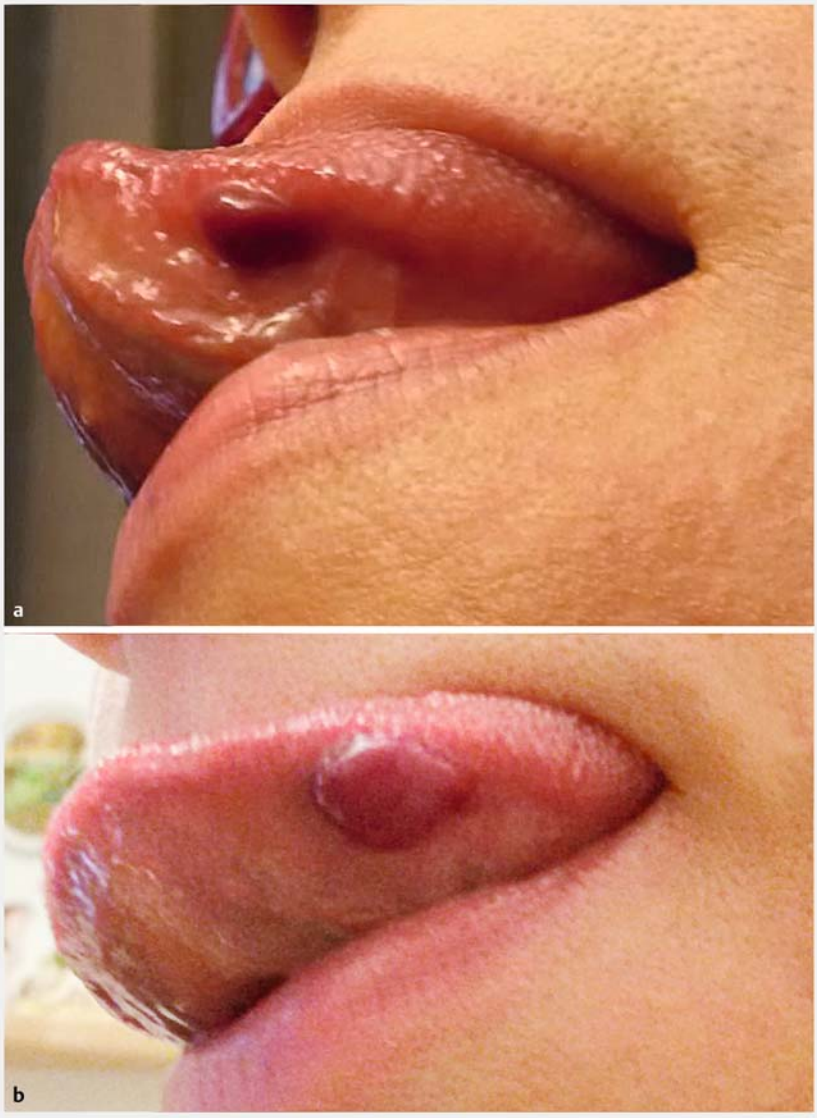
Competing interests

None



▶ **Fig. 2** Photographs showing: **a** a blood-filled blister on the tongue; **b** a hemorrhagic blister over the lateral tongue.

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References

[1] Badham NJ. Blood blisters and the oesophageal cast. J Laryngol Otol 1967; 81: 791–803
[2] Plantier F. Angina bullosa haemorragica. Ann Dermatol Venereol 2018; 145: 220–223
[3] Horie N, Kawano R, Inaba J et al. Angina bullosa hemorrhagica of the soft palate: a clinical study of 16 cases. J Oral Sci 2008; 50: 33–36
[4] Beguerie JR, Gonzalez S. Angina bullosa hemorrhagica: report of 11 cases. Dermatol Reports 2014; 6: 5282
[5] Kurban M, Kibbi AG, Ghosn S. Expanding the histologic spectrum of angina bullosa hemorrhagica: report of one case. Am J Dermatopathol 2007; 29: 477–479

Bibliography
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