IMAGE OF THE ISSUE

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Oesophagitis Dissecans Superficialis as the Presenting Sign of Isolated Oesophageal Mucous Membrane Pemphigoid

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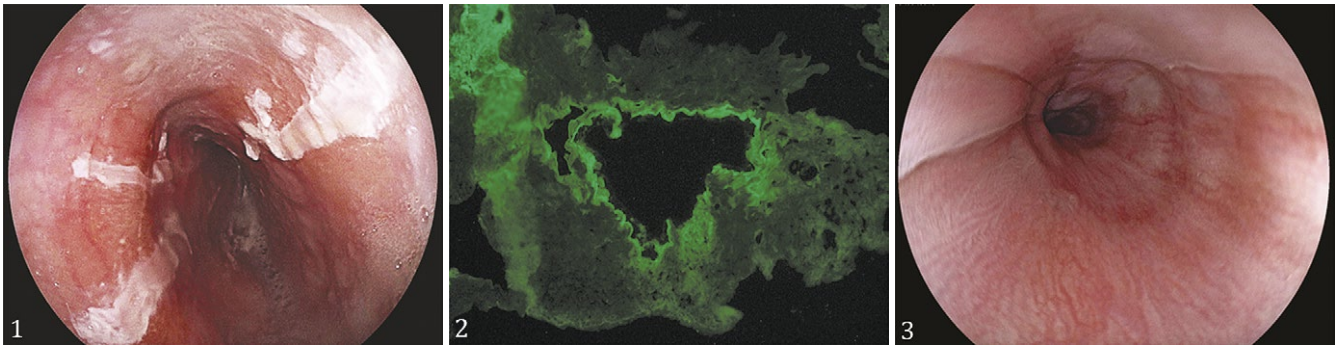

An 84-year-old man presented with recent-onset odynophagia and a two-year history of dyspepsia and heartburn. Oesophagogastroduodenoscopy (OGD) disclosed longitudinal sheets of sloughed squamous tissue with normal underlying mucosa involving the mid oesophagus and consistent with oesophagitis dissecans superficialis (ODS) (Fig. 1). Histopathology showed detached epithelial fragments lacking a basal layer with a nonspecific inflammatory infiltrate. Direct immunofluorescence (DIF) of perilesional mucosa revealed linear IgG deposits along the basement membrane zone (BMZ) (Fig. 2). Indirect immunofluorescence and enzyme-linked immunoassay for anti-BP 180, anti-BP 230 and anti-desmoglein antibodies were negative. Oesophagus-limited mucous membrane pemphigoid (MMP) was diagnosed. Pulsed intravenous methylprednisolone (125 mg/day for five days) followed by steroid tapering was given in combination with pantoprazole 40 mg/day, without any improvement. Thereafter, oral cyclophosphamide 100 mg/day was added. Over the next two months, symptoms gradually subsided concomitant with a normalization of the endoscopic picture (Fig. 3). Clinical remission persisted at the 37-months follow-up visit.

Oesophagitis dissecans superficialis is an exfoliative oesophageal disorder which may be associated with physical and chemical traumas, drugs, collagen diseases and autoimmune blistering diseases [1], notably pemphigus vulgaris and, more rarely, MMP, a mucous membrane-dominated subepithelial bullous disease caused by antibodies to different autoantigens of the BMZ [2]. Cases of classic MMP with oral, ocular and/or genital lesions manifesting also as ODS have been rarely reported [3, 4], possibly due to the underestimation of oesophageal symptoms by dermatologists and the inexperience with such lesions for most endoscopists. Furthermore, considering that oesophageal MMP usually occurs in the context of classic MMP with oral involvement and is typical in late stages [5], the exclusive oesophageal involvement at disease onset makes our case even more exceptional.

This case highlights that DIF should be performed when an endoscopic picture of ODS raises the suspicion of MMP, even in the absence of other skin and/or mucosal lesions.

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Conflicts of interest: None to declare.

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