Chronic Esophagitis Dissecans Secondary to Lichen Planus

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ABSTRACT
Chronic esophagitis dissecans secondary to lichen planus is a rare and often unrecognized pathology. The diagnosis is based on clinical (recurrent dysphagia and presence of cutaneous or genital lesions of lichen planus) endoscopic (desquamative esophagitis with proximal or multiple stenosis) and histological features. The treatment is still not well codified and the long-term prognosis remains unknown. We report a case of a woman with this affection with a 5 years follow up.

Introduction
Chronic esophagitis dissecans (COD) is a rare affection of unknown pathogenesis. It is the result of degenerative changes in the superficial layer of the esophageal mucosa. It is characterized by the association of chronic dysphagia, ruptured detachment of the esophageal mucosa in endoscopy, and histological features.

We report the observation of chronic esophagitis dissecans secondary to lichen planus revealed by acute dysphagia, with a follow-up of the 5-year-old disease.

Patient and Observation
A 68-year-old woman, having a history of hypertension and subtotal thyroidectomy for 8 years, consults in 2012 for high dysphagia, mixed with solids and fluids, paroxysmal, evolving for 1 month with a weight loss 10 kg in 2 months.

The anamnesis did not find any of caustic or foreign body ingestion nor notable dyspeptic syndrome.

The oral clinical examination did not show any erosive lesions and the abdominal as well as nodal areas check up were without particularities.

In esophagogastroduodenal fibroscopy, the esophagus is the site of flaky desquamation along its entire length, consisting of bubbles leaving an erythematous mucosa strewn with false membranes (Fig. 1), and a hiatal hernia at the cardiac level.

The biopsy and histopathological examination showed a non-suspicious esophageal mucosa.

Esophageal manometry revealed a nutcracker esophagus according to the Chicago classification.

The patient was treated with dual-dose proton pump inhibitors (PPIs), with a good clinical evolution for 2 years.

In 2014, the patient presented a recurrence of the same symptoms and the esogastroduodenal fibroscopy showed suspended esophageal ulcerations of atypical aspect (Fig. 2), a mucosa growing in tatters, an insurmountable stenosis easily bleeding on contact at 25 cm from dental arches and a pseudo diverticular orifice (Fig. 3, 4).

The biopsies performed were in favor of chronic esophagitis secondary to lichen planus disease, and the dermatological examination found secondarily vulvar atrophic lesions supporting the diagnosis of the disease.

In front of the endoscopic data, a thoracic CT scan was carried out urgently finding a pneumomediastinum suggestive of esophageal perforation on esophagitis dissecans (Fig. 5).

The patient was transferred for surveillance in specialized center of surgical digestive, left to fasting and under exclusive parenteral nutrition for 2 weeks, then put on systematic steroids associated with PPIs for several months. The evolution was favorable.

Away from the acute episode of perforation, oesogastroduodenal fibroscopy was performed with pediatric material finding esophageal stenosis passable by this device of 5 mm caliber, with a jump to 27 cm from the dental arches.

Very cautious and progressive esophageal dilatations (2 sessions spaced 3 weeks apart) with Savary candles were performed, from 5 to 11 mm, resulting in a clear improvement of dysphagia.

In 2016, the evolution was marked by the reappearance of dysphagia to solids on esophageal stenosis, requiring a new dilation session with candles of 11 to 14 mm.

Discussion
Chronic esophagitis dissecans (COD) is a rare pathology of relatively recent description. Its pathogenesis remains unknown, with treatment that remains poorly documented [1].

The diagnosis is based on clinical, endoscopic and histological data.

The presence of chronic dysphagia with a clear effect on the general condition should be reminiscent of think about the diagnosis and search for skin lesions or associated mucosa.

Esophagogastroduodenal fibroscopy found the ruptured detachment of the esophageal mucosa with a suspended stenosis, the biopsy found the dissection of the mucosa, without important inflammatory infiltrates [2].
Dysphagia is often explained by the sudden tipping of a mucosal flap, constituting a real tight "diaphragm", obstructing the oesophageal light [1].

Chronic dissecting oesophagitis may be secondary to certain systemic diseases: pemphigus vulgaris, bullous pemphigoid, lichen planus [1, 2]. This pathology was responsible for the oesophageal involvement in our patient.

Indeed, ODC secondary to lichen plan was first described by Al-Shihabi in 1982 [3]. It affects women slightly more frequently than men [4] and is rarely revealed by acute dysphagia [5].

The clinical manifestations of oesophageal involvement of lichen planus are varied from a dyspeptic syndrome to dysphagia. In addition, cutaneous involvement has often been described in association with esophagitis. The latter seems to be associated only with the chronic forms of lichen planus and not with the acute form [6]. In our patient vulvar involvement was concomitant with acute dysphagia.

At endoscopy, typically, the proximal esophagus is the most affected [7].

The lesion may be segmental or diffuse, characterized by suspended oesophageal stenosis, with ruptured delamination of the esophageal mucosa [8], as is the case with our patient. More rarely, bullosa can be involved in the esophagus [8].

Oesophageal biopsies may be suggestive of lichen planus but may not always confirm the diagnosis. Typical findings include basal keratinocyte damage and dense lymphocyte infiltration in the subepithelial layer. The presence of the anuclear remains, called the body of Civatte, is very characteristic of the lichen plan [9].

Therapeutically, no specific therapeutic approach to management of COD secondary to lichen plan has been established. Based on previous reports, different therapeutic schemes have been tried with contradictory results. Systemic corticosteroids have been proven effective and used as a first line of treatment: initially, oral prednisone 40-60 mg, daily for four to six weeks, with a response usually obtained at two weeks [10].

Oesophageal stenosis can be successfully dilated with temporary relief of dysphagia [10]. Dilatations should be done very carefully and gradually, given the risk of iatrogenic perforation on fragile esophagus.

Indeed, the oesophageal perforation can be a surgical but also medical treatment that can be indicated only in case of discovery of a crack without pleural or mediastinal incision [11].

The criteria for medical treatment were defined by Cameron & al in 1979 and Aljortay & al in 1997 [11]:
- Intramural perforation;
- Transmural perforation if it is well circumscribed, diagnosed early or on the contrary very late;
- The esophageal transit showing the passage of the contrast product preferentially in the esophageal lumen and not towards the fistulous
- The absence of an underlying obstacle related to a tumor or an unexpanded stenosis;
- Clinical signs of minimal sepsis;
- The appearance of clinical improvement within 24 hours after the start of treatment.

In our patient, the evolution over 5 years was marked by the recurrence of dysphagia on oesophageal stenosis requiring 3 sessions of endoscopic dilatation.

Conclusion
Chronic oesophagitis dissecans is a rare cause of dysphagia that is often overlooked. The chronic course of the disease can be laced with episodes of acute dysphagia. Other studies are essential in order to understand its physiopathology and to define appropriate management.

Competing interests
The authors declare no competing interest.

Author contributions
All authors contributed in the development of this publication and approved the final manuscript.

Figures

[Figure 1. Erythematous esophageal mucosa with desquamation.]
[Figure 2. Atypical aspect of suspended esophageal ulcerations.]
[Figure 3. Esophageal stenosis bleeding easily on contact.]
Figure 4. Bleeding esophageal stenosis with pseudodiverticular orifice.

Figure 5. Coronal section CT showing posterior mediastinal pneumo.

References