CASE REPORT

A patient with odynophagia and unusual endoscopic findings

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SUMMARY

The case describes a 50-year-old woman presenting with a severe painful dysphagia to solids, impacting on her nutritional intake. She had a history of pemphigus vulgaris maintained in remission with azathioprine, with no evidence of active oral or cutaneous disease at the time of presentation. Endoscopy and histology from the distal oesophagus revealed oesophageal involvement of pemphigus vulgaris. This is a relatively rare clinical form of the disease, with only 58 cases reported worldwide. Patients with pemphigus vulgaris are also prone to infective or steroid-induced gastritis, which present in the same way. Early endoscopic evaluation is therefore essential to distinguish between oesophageal involvement of pemphigus vulgaris and other pathologies, which warrant significant differences in management.

BACKGROUND

Pemphigus vulgaris is an autoimmune condition of the skin and mucous membranes. Oesophageal involvement is rare, with only 58 cases reported worldwide.¹ Most patients present with dysphagia and/or odynophagia, although the abrupt onset of symptoms in this case is unusual.

The diagnosis of oesophageal pemphigus vulgaris may not be immediately apparent even in those with known cutaneous pemphigus, who are also prone to steroid-induced or infective gastritis.² Prompt endoscopy (and biopsy for histological analysis) is therefore essential,^{3–4} not least because there are obvious differences in the use of steroids in the management of oesophageal pemphigus vulgaris compared to steroid-induced gastritis. Distinguishing the underlying pathology is therefore key.

CASE PRESENTATION

A 50-year-old woman presented with acute onset of painful dysphagia to solids, developing following a meal the preceding day. The severity of her symptoms resulted in sitophobia having a negative impact on her weight. Her medical history was significant for cutaneous pemphigus vulgaris, diagnosed 2 years previously and maintained in remission with azathioprine.

Physical examination revealed dehydration. Of note, there was no clinical evidence of oral or cutaneous pemphigus.

Initial management included intravenous fluid rehydration and analgesia.

INVESTIGATIONS

The patient was referred for urgent upper gastrointestinal endoscopy, demonstrating a 6 cm long mucosal abnormality in the distal oesophagus (figure 1) from which biopsies were obtained (figure 2).

The endoscopic image shows non-specific patchy inflammation and sloughing of the distal oesophageal mucosa to reveal denuded submucosa. Notably, 4 months prior to this presentation, the patient had a routine gastroscopy, which was entirely normal, indicating that the endoscopic findings here were new. The non-specific endoscopic appearances meant that a broad differential diagnosis remained at this stage, including infectious oesophagitis and eosinophilic oesophagitis.

Subsequently, histology demonstrated suprabasal clefting and acantholysis (loss of intercellular connections),⁵ consistent with oesophageal pemphigus vulgaris. This is demonstrated by the arrow in figure 2, which indicates an acantholytic epithelial surface.

TREATMENT

Input from oral medicine and dermatology was sought, as well as dietician review to optimise nutrition. The patient was treated with intravenous methylprednisolone (1 mg/kg) for 1 week. This was converted to prednisolone 40 mg daily once oral intake was established, and azathioprine dosing was optimised. Dysphagia and odynophagia were slow to resolve, and recovery was complicated by steroid-induced proximal myopathy. Once

Figure 1 Endoscopic view of the distal oesophagus, showing non-specific patchy inflammation and sloughing of the distal oesophageal mucosa to reveal denuded submucosa.

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Figure 2 Oesophageal histology (H&E; ×20 magnification) demonstrating acantholysis (marked with arrow).

symptoms were adequately controlled, the prednisolone dose was weaned down at a rate of 5 mg every 5 days.

OUTCOME AND FOLLOW-UP

At follow-up 4 months after diagnosis, the patient's oesophageal pemphigus is in remission and she remains on azathioprine maintenance therapy. She has been established back on to a full normal diet.

DISCUSSION

Pemphigus vulgaris is caused by antibodies against the cellular adhesion protein desmoglein 3. The resulting loss of cohesion between keratinocytes leads to flaccid blisters and characteristic shearing of the epidermis following mechanical abrasion (Nikolsky's sign).⁶

Typical endoscopic findings of oesophageal pemphigus vulgaris are generally regarded to include flaccid bullae, as well as shearing of the oesophageal lining following abrasion.⁷ In this case, however, the endoscopic appearance was non-specific and the diagnosis was made on histological findings.

It is also interesting to note the absence of oral or cutaneous lesions, despite the oesophageal involvement in this case. This is particularly unusual. Most case-based reports in the literature describe patients with pemphigus vulgaris to have either cutaneous and/or oral lesions at the time of diagnosis of the oesophageal lesions.^{2 8}

While rare, oesophageal pemphigus vulgaris is an important cause of dysphagia and odynophagia in patients who have a history of the cutaneous form of the disease. Evidence to guide the management of these patients is limited. Case-based reports support the use of high-dose corticosteroids and steroid-sparing agents such as azathioprine.^{1 9 10} Multidisciplinary input, in particular, early dermatological and dietetic involvement, is key.

Learning points

- ▶ In patients with pemphigus vulgaris presenting with upper gastrointestinal symptoms, always consider oesophageal involvement of the disease as part of the differential diagnosis.
- ▶ Early endoscopic evaluation, in particular to obtain tissue for histological analysis, is essential to distinguish between oesophageal involvement of pemphigus vulgaris and other pathologies, which may warrant significant differences in management.
- ▶ Multidisciplinary involvement is essential in the management of oesophageal pemphigus vulgaris, in particular, early dermatological and dietetic involvement.

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