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Boerhaave's Syndrome - An Unusual Presentation of Eosinophilic Oesophagitis

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Abstract

Eosinophilic Oesophagitis (EoE) is a chronic disease of the oesophagus characterised by mucosal eosinophilia and oesophageal dysfunction. It is a common but under-recognised cause of oesophagitis, most commonly affecting young males with a history of atopy.1 The estimated prevalence ranges from 13 and 49 cases per 100,000, although this is likely to be underestimated due to reduced awareness of this condition.2 Oesophageal rupture as a complication of EoE has been widely reported in association with food impaction and as a complication of endoscopy. Spontaneous oesophageal perforation (Boerhaave's syndrome) is less common.3 we present an unusual case of Boerhaaves's syndrome as a presenting feature of EoE in a previously asymptomatic patient. To our knowledge this has not been previously reported.

Keywords: Eosinophilic oesophagitis; Boerhaave's Syndrome; EoE

Background

Eosinophilic Oesophagitis (EoE) is an increasingly recognised chronic, allergic, inflammatory condition of the oesophagus, with the incidence varying from 1 to 20 new cases per 100,000 people per year. While EoE was initially described in the 1970s, it has only been a recognised clinic pathological disorder since the 1990s. Adults typically complain of dysphagia, food bolus impaction or chest pain. There is heterogeneity in the presentation of EoE and dysphagia may present later which may contribute to delay in diagnosis. The oesophagus, in the normal state, is devoid of eosinophils, and inflammation driven by eosinophils is characteristic of EoE. Nonetheless, there is significant variability in eosinophil concentrations throughout the length of the oesophagus, with a more distal location of eosinophilia steering towards a diagnosis of gastro-oesophageal reflux disease (GORD). Thus, confirmation of EoE involves obtaining 2-4 biopsies from the proximal, mid and distal oesophagus - a protocol which has been shown to improve diagnostic accuracy [1]. Treatment options include dietary restrictions (usually a 6-food elimination diet) as well as pharmacological treatment with acid suppression and topical corticosteroids. A typical regimen initially involves acid suppression with an eight-week course of a proton pump inhibitor followed by repeat gastroscopy and biopsy after 8-12 weeks to assess endoscopic and histological response. If either symptoms or endoscopic evidence of EoE persist, topical corticosteroid therapy is commenced with dose titration based on clinical and/or histological response [1,2].

Case Report

An adolescent male patient, with a background of mild asthma treated with salbutamol as-required, was referred to our centre with persistent pyrexia, night sweats and weight loss. He denied any gastrointestinal symptoms including dyspepsia, dysphagia or chest pain. Routine bloods revealed a C-reactive protein of 52, normal white cell count and normal eosinophil count. A contrast-enhanced computed tomography (CT) scan of the thorax revealed a 4.4cm mediastinal mass with associated lymphadenopathy. Linear endoscopic ultrasound (EUS) demonstrated multiple inflammatory appearing posterior mediastinal lymph nodes but the mass could not be demonstrated. The EUS endoscope was

subsequently exchanged for a diagnostic gastroscope, which revealed longitudinal furrowing and trachealisation of the oesophagus in keeping with eosinophilic oesophagitis. This diagnosis was later confirmed histologically with biopsies showing 66 eosinophils per high-powered field (hpf). The patient was commenced on omeprazole 20mg twice daily and swallowed (rather than inhaled) fluticasone 440mcg twice daily. A post 8-week therapy thoracic magnetic resonance imaging (MRI) scan showed a reduction in the size of the inflammatory mass from 4.4cm to 1.5cm in diameter and resolution of lymphadenopathy. Eight months after starting treatment, the patient has remained well and continues on topical fluticasone and omeprazole (Figures 1-3).

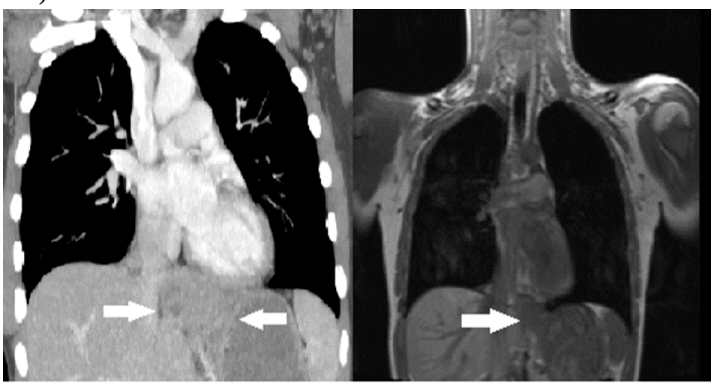


Figure 1a: *Contrast enhanced computed tomography (CT) scan demonstrating a 45mm peri-oesophageal anterior mediastinal mass with displacement of the left diaphragmatic crus (white arrows).*

Figure 1b: *Follow-up magnetic resonance (MR) thorax demonstrating a thickened gastro-oesophageal junction measuring 17mm by 6mm (white arrow) reduced in size in comparison to index CT scan and with resolution of lymphadenopathy.*

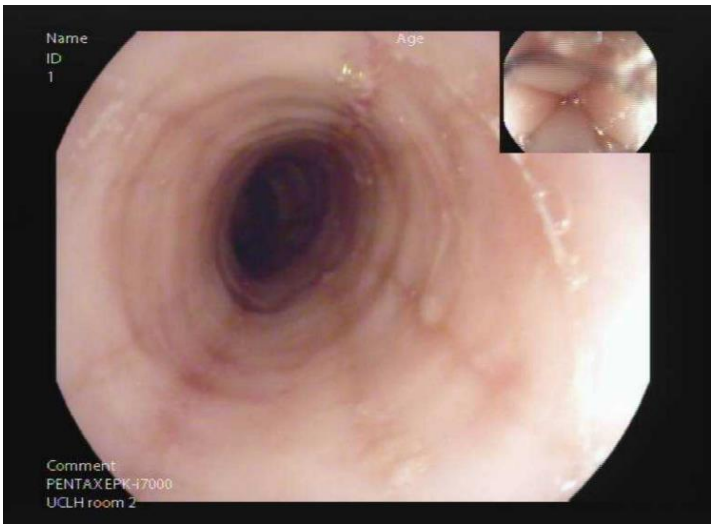
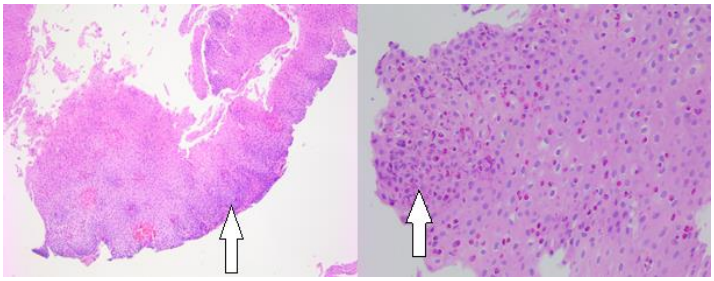


Figure 2: *Gastroscopy showing longitudinal furrowing & trachealisation of the oesophagus.*



Figures 3a*: Eosinophilic oesophageal inflammation in eosinophilic oesophagitis.*

Figure 3b: *High-power view showing large numbers of eosinophils accumulating preferentially towards the luminal surface.*

Discussion

EoE is an increasingly recognised condition with a rapidly increasing incidence [1-4]. In adults, dysphagia and GORD type symptoms predominate. Endoscopic examination may often be normal [1-3]. Although typical endoscopic findings include longitudinal furrows, concentric rings (trachealisation), white exudates and loss of vascular markings [5]. Biopsies demonstrating more than 15 eosinophils/hpf are required for diagnosis [4]. The practical management of EOE can be challenging due to minimal data on optimal length of treatment and long-term outcomes. There is emerging evidence that maintenance treatment with topical corticosteroids, particularly in cases of oesophageal perforation, may slow progression and reduce further complications. Long-standing uncontrolled eosinophilic inflammation of the oesophagus can lead to cellular hyperplasia, mucosal fragility, and wall re-modelling and stricture formation [6]. The resultant narrowing of the oesophagus pre-disposes to food bolus impaction which can lead to secondary perforation. Endoscopic dilation of strictures can lead to iatrogenic perforation. Our case illustrates both the variability in the presentation of EOE and the significance of the vulnerability of the oesophageal wall. The case demonstrates an important consequence of uncontrolled chronic oesophageal inflammation. Oesophageal perforation, or Boerhaave's syndrome, can lead to significant consequences such as mediastinitis, abscesses, infection of the spinal cord and empyema which may lead to organ failure and death. Clinicians should remain aware of EoE as a potential cause of oesophageal perforation even when, as in our case, there are no preceding oesophageal symptoms.

Conflicts of Interest

All authors declare no conflict of interest related to this study.

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