A young man with progressive dysphagia

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CASE REPORT

In 2008, a 27-year-old Caucasian male presented with dysphagia which had been present for over two years. His symptoms had been progressive over the last few months with solid food impaction, which resolved spontaneously, several times a week. His weight had remained stable and he did not complain of pyrosis, acid reflux or chest pain. An upper gastro-intestinal (GI) tract endoscopy had already been performed in 2006, but a clear aetiology had not been established. Proton pump inhibitor (PPI) treatment had not relieved these symptoms. Medical history revealed an allergic reaction of unknown origin at the age of six with angio-oedema of buccal and genital mucosa. In addition he reported a strong family history of allergic rhinitis, from which he also suffers. An upper GI tract endoscopy was performed and revealed a characteristic finding (figure 1A and 1B).

WHAT IS YOUR DIAGNOSIS?

See page 203 for the answer to this photo quiz.

Figure 1A. Endoscopic picture of upper oesophagus showing concentric fixed ring-like mucosal deformation or 'trachea-like' aspect

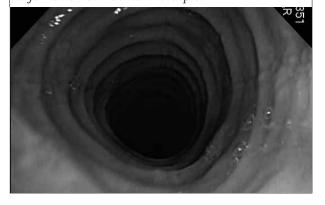
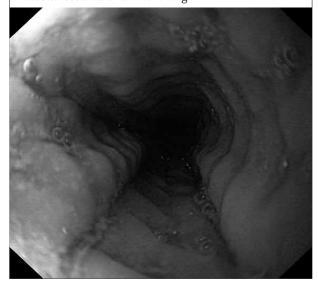


Figure 1B. Endoscopic picture of mid oesophagus showing oesophageal furrowing with longitudinal lines and nodular aspect of mucosa, indicative of mucosal oedema and thickening



ANSWER TO PHOTO QUIZ (ON PAGE 202)

A YOUNG MAN WITH PROGRESSIVE DYSPHAGIA

DIAGNOSIS

At endoscopy fixed rings and longitudinal furrowing were seen in the upper and mid oesophagus (figure 1). Biopsy of upper oesophageal tissue revealed dense eosinophilic infiltration (240 eosinophils per high-power field (HPF) of 0.2 mm²) (figure 2).

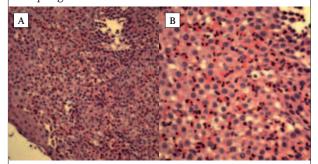
Laboratory examination showed marked elevation of serum IgE (802 IU/ml), without eosinophilia. Serum allergy testing was positive for several food allergens (milk, wheat and peanuts), aerogenic allergens (grass and tree pollen) and dust mites.

These macroscopic findings, the eosinophilic infiltrates, the lack of response after two months of PPI treatment and the diminution of symptoms following initiation of topical steroid therapy (swallowing fluticasone propionate 250 $\mu g/$ puff, two puffs twice daily) confirmed the diagnosis.

EO has been described for over three decades, but has become an entity of increasing interest because of its apparent rising prevalence with an incidence of around 1:10,000 persons/year. EO predominantly affects children and young adults. Men are more commonly affected than women.^{1,2}

Common presenting symptoms in adults are dysphagia, food impaction and gastro-oesophageal reflux disease

Figure 2. Light microscopic picture of upper oesophageal tissue



Marked infiltration of eosinophils, layered from submucosa to superficial epithelium, and microabscess formation (clustering of 4 or more eosinophils) (haematoxylin and eosin stain; original magnification A) x 100 and B) x 200).

(GERD) symptoms. Its natural history has not been clarified, but no effect on life expectancy or development of (pre)malignant oesophageal disease has been reported. 1,2 A diagnosis can be made on endoscopic and microscopic findings, after exclusion of other illnesses associated with oesophageal eosinophilia, such as eosinophilic gastroenteritis, GERD and Crohn's disease. At endoscopy longitudinal furrowing, whitish exudates, fixed rings and strictures can be seen in the proximal, mid or distal oesophagus. The lack of macroscopic abnormalities, however, does not exclude the disease. Therefore, in patients with typical symptoms but without endoscopic abnormalities, biopsies of normal mucosa should be considered.2 Microscopic features associated with EO are >15 intraepithelial eosinophils per HPF, eosinophilic microabscess formation, superficial epithelial layering of eosinophils and basal zone hyperplasia.¹ Peripheral eosinophilia, high total IgE and food specific IgE have been described. An association with allergic rhinitis, eczema and asthma has also been recognised.1

Recommended treatment strategies for EO include diet therapy with specific food restrictions and topical or systemic steroids.²⁻³ Oesophageal strictures, a complication of EO, might be safely dilated, although oesophageal perforation has been described.⁴

This case illustrates the significant burden of this disease and emphasises the importance of improved awareness.

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