Eosinophilic Oesophagitis in Adults: Case History and Literature Review

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Abstract: Eosinophilic oesophagitis has become one of the common clinical presentations encountered by gastroenterologists and histopathologists. Despite the huge strides in the understanding of pathogenesis, diagnosis and treatment of the condition over the past decade, data regarding clear management guidelines are still limited. Presented is the case of a 28 years old female who was referred to our endoscopy department with worsening dysphagia. The diagnosis of Eosinophilic oesophagitis was confirmed. This article discusses the clinical presentation, diagnosis and summarise the outlines of management in adults.

Keywords: Eosinophilic oesophagitis, clinical presentation, diagnosis, treatment, Food allergy.

INTRODUCTION

Cases of probable eosinophilic oesophagitis were first reported in late 1960 [1], In 1978, Landres *et al*. reported an isolated case of severe achalasia in a patient with oesophageal esionphilia [2], however, it was not until 1993 when the clinicopathological features of eosinophilic oesophagitis were first described. Intraepithelial oesophageal eosinophils (IEE) were found in patients presenting with dysphagia in absence of other pathology [3]. In 1995, it was reported as an allergic disease that responded to dietary elimination [4]. Since then, eosinophilic oesophagitis has become more common with increased prevalence. In USA, the estimated prevalence is at 50 to 100 per 100,000 of general population, which is similar to estimate prevalence of ulcerative colitis [5].

CASE HISTORY

28-years-old Caucasian female with history of multiple food allergies, referred by her GP for open access OGD with worsening dysphagia for 2 years and recent episodes of food impaction. She had a barium swallow and meal 6 months prior to referral, which showed only proximal oesophagitis (Figure **1**) that did not respond to proton pump inhibitor therapy PPI. Her only past medical history is asthma and hay fever. The OGD showed severe oesophageal narrowing and trachealisation with difficulty passing standard gastroscope beyond 27 cm (Figure **2**). Multiple biopsies form the proximal and mid oesophagus were obtained.

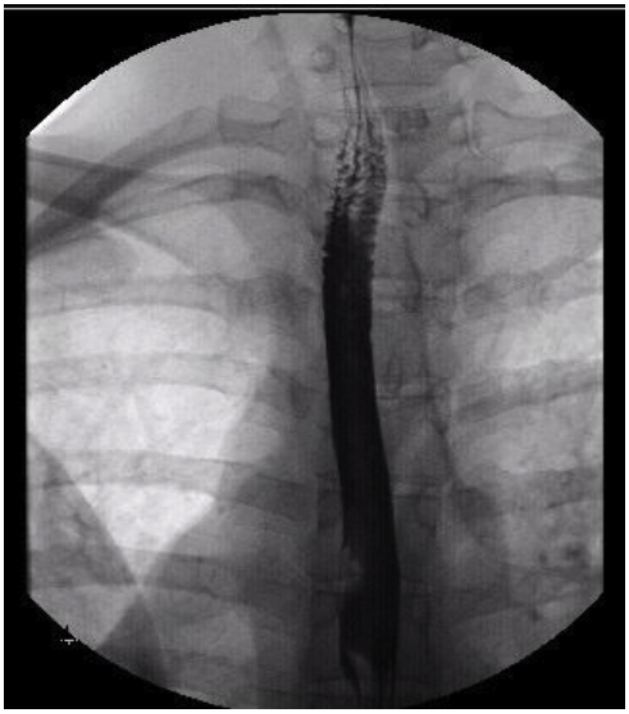

Figure 1: Barium swallow and meal: proximal oesophagitis and no stricture.

Histopathology confirmed squamous epithelium with prominent number of eosinophils with micro abscesses (Figure **3a** and **3b**). She was commenced on Fluticasone inhalers 250 microgram 2 puffs Twice a day (to swallow) and offered a follow up appointment in outpatient clinic to assess the response to the treatment. She reported only partial response. Unfortunately 6 weeks later, she was admitted *via* emergency department with difficulty swallowing. Whilst inpatient she was commenced on oral budesonide MR 9 mg. In view of the severity of her symptoms she was advised elemental diet and referred to the immunology clinic to look for food triggers. Skin

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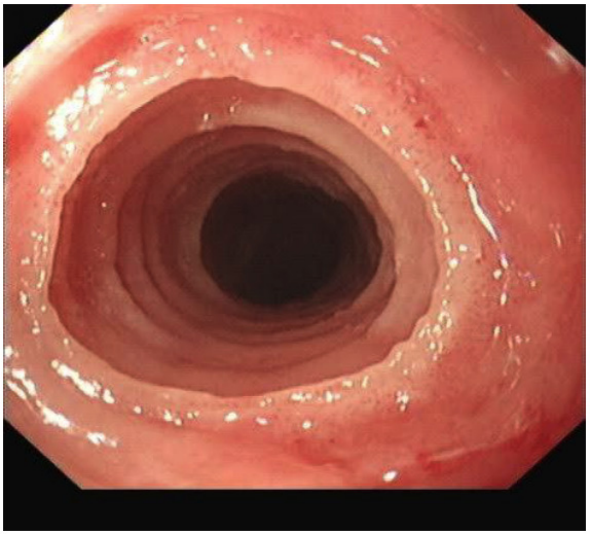
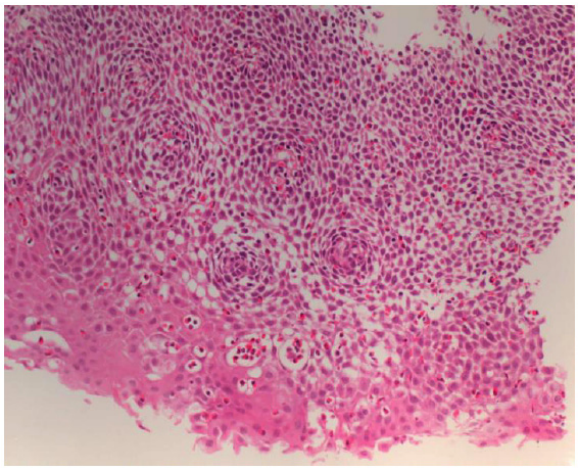


Figure 2: Index OGD: Severe Oesophageal trachealization and stricture.



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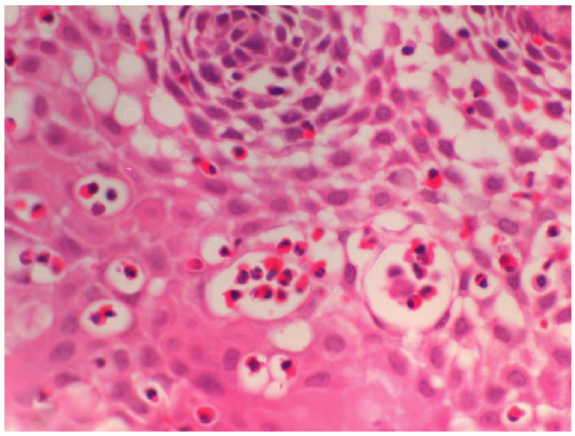

b

Figure 3: a. Histopathology: Esinophilic Oesophagitis.

b. Histopathology: 15-20 Esionphils per High power field.

prick test was arranged and she was found to be sensitive to Profilins (allergen widely present in fruits, nuts and latex). She responded well to elemental diet (Elemental 028 Extra liquid, 200 ml, 9 per day). A repeated OGD 3 months form the index diagnosis showed featureless oesophagus but no stricture (Figure **4**). Histopathology of repeated Oesophageal biopsy showed only hyperplastic squamous mucosa with few intraepithelial lymphocytes suggestive of reflux oesophagitis (Figure **5**). The patient preferred to continue with elemental diet. Dilatation has been recommended if she experienced further recurrence. She remains under regular follow up by dietician, immunologist and the gastroenterology team.

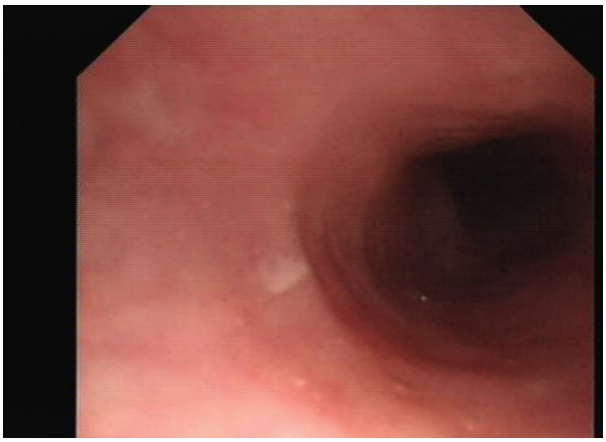

Figure 4: Repeated OGD in 3 month: Featureless but no stricture.

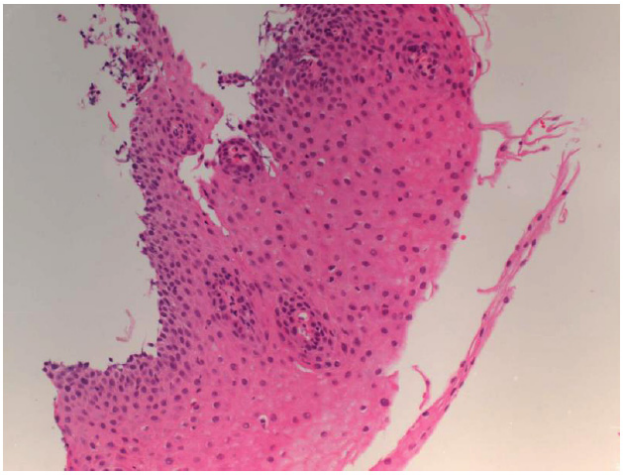

Figure 5: Histopathology: few intraepithelial lymphocytes, no esinophils.

DISCUSSION

This case history illustrates a difficult case of eosinophilic oesophagitis. She is a Caucasian with atopy who presented with dysphagia and food impaction. The OGD findings were striking. Pharmacolological treatment was unsatisfactory and

she only responded to elemental diet. Clear management guidelines are important in treatment of such patients.

The updated consensus about Eosinophilic oesophagitis (2011) defined the condition as chronic, immune/antigen-mediated disease characterized clinically by symptoms related to oesophageal dysfunction and histologically by eosinophil-predominant inflammation [6].

Eosinophilic oesophagitis is more common in males (male to female ratio 3:1) [6], typically Caucasian with atopy. It usually present in childhood or third/forth decades of life.

In adults, the most common presentation is dysphagia to solids and food impaction [7]. Others symptoms include: chest pain, heartburn, upper abdominal pain, odynophagia and vomiting. None of the symptoms is pathognomonic and most of the patients end with the diagnosis of gastro-oesophageal reflux disease. Endoscopy is recommended after failure of a trial of PPI.

Diagnosis is based on clinical presentation of oesophageal dysfunction and histological criteria from oesophageal biopsy (Table **1**).

Table 1: Criteria for Diagnosing Esinophilic Oesophagitis

Symptoms of oesophageal dysfunction.
≥15 eosinophils per HPF.
Eosinophilia limited to the oesophagus.
Other causes of oesophageal eosinophilia excluded

At endoscopy, the possible mucosal abnormalities of the oesophagus has recently been referred to as "the eosinophilic oesophagitis endoscopic reference score": EREFS, which stand for: Exudate, Rings, Edema, Furrows and Strictures. None of these features are specific and in isolation are insufficient to make a diagnosis [8]. The endoscopic appearance of the oesophagus can be completely normal in 10% of patients [4].

Oesophageal biopsy should be obtained if the condition is suspected. It should also be considered in all patients presenting with dysphagia but had normal looking oesophagus or gastro-oesophageal reflux disease (GORD) that is not responsive to PPI.

As eosinophilic oesophagitis is patchy disease, minimums of 2-4 biopsies are recommended from the proximal and distal end of the oesophagus. More biopsies increase the diagnostic yield. Simultaneous biopsy from gastric antrum and duodenum is recommended to exclude other potential causes of GI esinophilia (Table **2**, other causes of oesophageal esinophilia).

Table 2: Other Conditions with Oesophageal Esinophilia

Eosinophilic gastrointestinal diseases.
Coeliac disease.
Hypereosinophilic syndrome.
Achalasia.
Connective tissue diseases.
Crohn's disease.
Drug hypersensitivity.
PPI-responsive esophageal eosinophilia.

The minimum diagnostic threshold is ≥ 15 intraepithelial eosinophils per high-power field (HPF) in the oesophageal biopsy specimen [6]. Other supportive histological features include: esionphils micro-abscesses, basal zone hyperplasia and extracelluar esinophilic granules [9].

PPI trial has been the first line of pharmacological treatment for patient with oesophageal symptoms with response to treatment indicating GORD. Recent studies shown that some patients with oesophageal esnionphila and no evidence of GORD, respond well to treatment with PPI, PPI-responsive oesophageal esinophilia (PPI-REE) [10]. It is therefore recommended that all patient with symptomatic oesophageal esinophilia receive a trial of PPI treatment, 20-40 mg twice daily for 8 weeks [11].

Having excluded PPI-REE, Topical steroids are the first line of treatment. These include fluticasone inhalers and neubilised viscous budesonide – to be swallowed- for 8 weeks. They result in reduction of the esinophilic count with some symptom resolution, however there is a high chance of recurrence once treatment stopped. Candidal oesophagitis has been reported in 5 – 30 % of cases [12]. Systemic steroids like prednisolone can be used if no response to topical treatment and rapid symptoms resolution is needed. Their use is limited due to high rate of relapse and side effect profile [13].

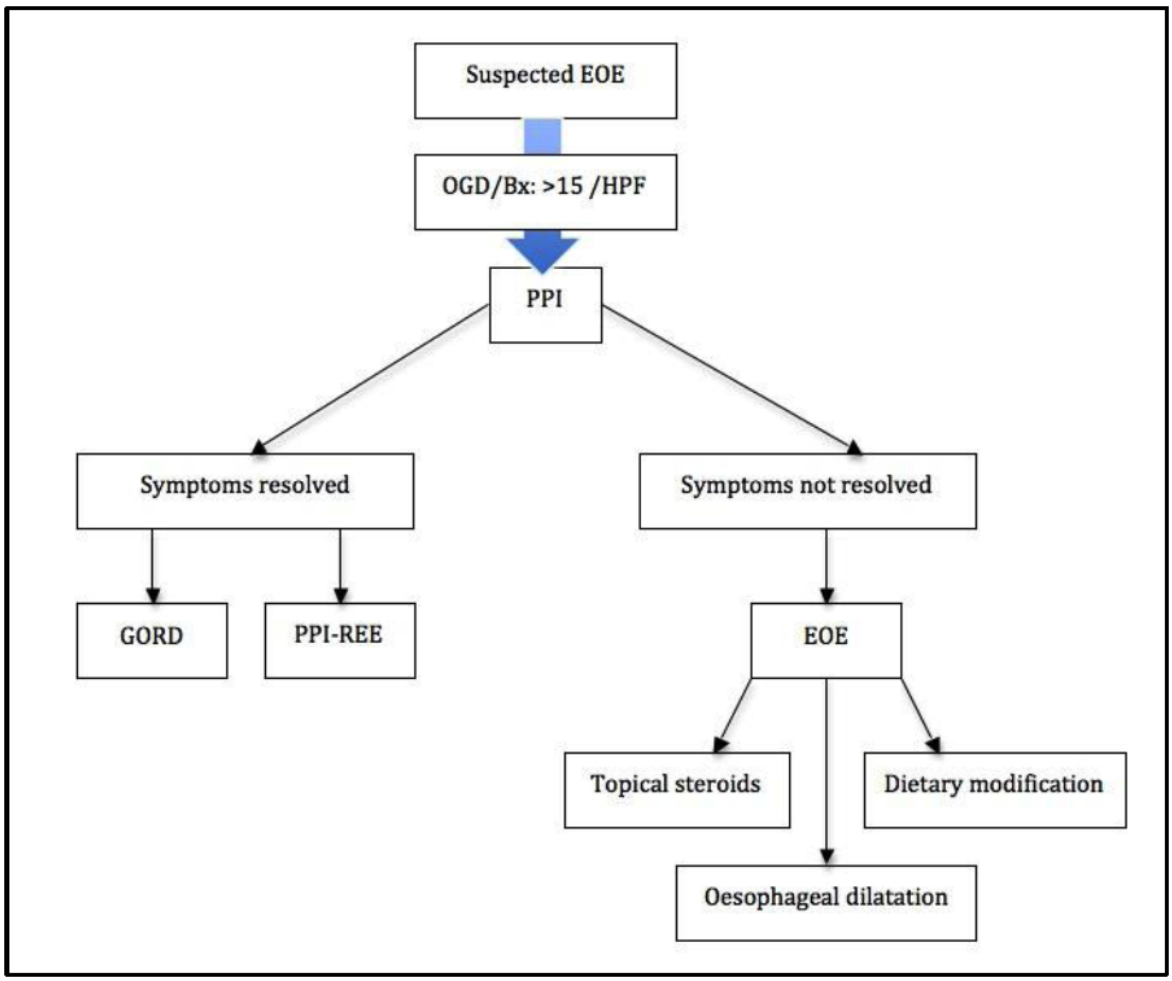

Figure 6: EOE Management flow chart.

Food allergy is reported in 15% to 43% of patients with eosinophilic oesophagitis [14]. Diet therapy to find food that trigger the esinophilia should be considered. There are 3 approaches to diet therapy, which have demonstrated symptomatic and histologic resolution in paediatric patients in uncontrolled studies [15]. Food elimination has not been as widely studied in adults. The decision to use a specific dietary approach should be tailored to individual patient needs and available resources. Direct elimination diet is based on result of skin prick testing and has 46% success rate. Empirical 6-food elimination diet (soy, egg, milk, wheat, nuts, and seafood) has 72% success rate. Elemental diet using amino acid based formula has 91% success rate.

Although oesophageal dilatation is a very effective treatment when strictures develop, it is generally considered when other forms of treatment failed. There is no preferred method of dilation (balloon vs buge), however aiming for 15-18 mm diameter appear to provide good relief of dysphagia. This may need few sessions to achieve [16]. The most common complication post-dilatation is chest pain occurring in 75% [17]. Recent studies shown that the risk of perforation is 0.3% in experienced centres [18].

SUMMARY AND CONCLUSION

The heterogeneous nature of eosinophilic oesophagitis and lack of specific symptoms make it difficult to predict. Therefore Eosinophilic oesophagitis should be considered as one of the main differential diagnoses in any patient presenting with a history of intermittent or continuous dysphagia or GORD not responding to PPI. Once the condition is confirmed by histology, the treatment should be tailored to the patient, starting by topical steroids and follow the chart (Figure **6**).

CONFLICT OF INTEREST

The authors declare that no financial or other conflict of interest exists in relation to the content of the article.

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