CASE REPORTS

Distal oesophageal spasm secondary to eosinophilic oesophagitis in a child: response to diet therapy

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ABSTRACT

We report a case of a school-age child with symptomatic distal oesophageal spasm (DES), clinical dysphagia and typical feature in high-resolution oesophageal manometry secondary to eosinophilic oesophagitis (EoE). His symptoms resolved with normalisation of oesophageal manometry after standard treatment of EoE. DES is mainly an adult disorder and rarely affect children; to the best of our knowledge, this is the first reported case in a child that document full recovery after treating the underlying EoE.

INTRODUCTION

Eosinophilic oesophagitis (EoE) is a chronic, immune-mediated disease characterised clinically by symptoms related to oesophageal dysfunction and histologically by eosinophil predominant inflammation of at least 15 eosinophils per high-power field (HPHF).¹ Children with EoE may present with a variety of non-specific symptoms such as feeding difficulties, failure to thrive and reflux like symptoms, while adolescents usually present with dysphagia and food bolus impaction.²

Distal oesophageal spasm (DES) is a major disorder of peristalsis diagnosed by oesophageal manometry and defined by Chicago classification as the occurrence of premature oesophageal contractions in at least 20% of swallows in the presence of normal lower oesophageal sphincter (LOS) relaxation.³ It commonly presents with dysphagia and/or non-cardiac chest pain. DES is an uncommon motility disorder that usually affects adults with average age of presentation at 60 years.

Up to half of adult patients with EoE are reported to have oesophageal motor disorders as detected by oesophageal manometry. Although children with EoE can also develop oesophageal dysmotility,

there is paucity in literature in formal manometric assessment.⁴ We report a case of a young child with DES secondary to EoE, a complete resolution of symptoms and normalisation of oesophageal manometry after EoE treatment.

A 6-year-old child with aberrant origin of the right subclavian artery and left aortic arch presented with difficulty in swallowing solid food and cough while eating. The symptoms were initially attributed to vascular malformation and he was extensively investigated by Barium study, MRI and bronchoscopy. Results showed non compressed trachea and patent airways. There was mild indentation of the upper oesophagus in the barium but no contrast holdup, no proximal dilatation and the distal two-third of the oesophagus were normal. He underwent an endoscopy and high-resolution oesophageal impedance manometry (36 channels HRiM MMS). Endoscopy while on acid suppression showed linear furrows, and biopsies (six biopsies from upper, mid and lower oesophagus) showed basal cell hyperplasia, spongiosis, elongation of the papillae and increased eosinophils infiltrate >15 PHPF (16–33 in all levels). Oesophageal manometry showed normal LOS and reduced distal latency (DL) <3.8 in >20% of contractions, high distal contractile integral (DCI) >4500 (figure 1) and multi-peaked contractions, features typically seen in DES. He was started on six items exclusion diet (cow's milk protein, wheat, egg, soy, peanut/tree nuts and fish).^{1,4} Parents reported good symptoms response after 2 weeks and complete resolution of all swallowing difficulties after 6 weeks. Repeat endoscopy and oesophageal manometry 4 months later showed normal oesophageal motility (figure 2) and normal endoscopic

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Figure 1 Wet swallow distal latency 3.4.

mucosal appearance; histology were mostly normal in all but single biopsy which showed increased eosinophil counts (33).

DISCUSSION

EoE is an immune-mediated condition characterised by the presence of at least 15 eosinophils PHPF and symptoms of oesophageal dysfunction.¹ It is not an uncommon disorder with rising incidence and prevalence in paediatric population. EoE can be associated with atopic disease and it thought to result from adaptive immune response to food allergens. One of the hallmark symptom of EoE is oesophageal dysfunction and impaired oesophageal motility. In school-age children, the most common presentation of EoE is food refusal, preference for soft or liquid diet and eating slowly. Abdominal pain and vomiting can also dominate symptoms in this age group, while food impaction and dysphagia are usually common in older children and adolescent similar to adults.

Figure 2 Wet swallow normal.

This difference in range of symptoms in different age group may simply reflect varied abilities to express and communicate symptoms between children and adults or a lifestyle and eating adaptation along the years. Treatment modalities of EoE include food elimination (empiric six food elimination diet) or topical steroid therapy. Both treatment options are effective with reported clinical and histological remission in both children and adults.¹

DES is a rare motility disorder of the oesophageal smooth muscles. It was first described as manometric abnormality by Creamer *et al* in 1958 and thought to affect the entire oesophagus and hence erroneously called diffuse oesophageal spasm; however, it became clear that the disorder mainly affect the distal part of the oesophagus and the current and more accurate nomenclature of DES was adopted. It is mainly an adult disorder that present with dysphagia and chest pain; it has rarely been described in paediatric. DES is often idiopathic, but can be secondary to gastro-oesophageal reflux disease GORD.

Although contrast studies can suggest the presence of oesophageal spasm and features such as corkscrew oesophagus has been described, oesophageal manometry is considered the gold standard to diagnose DES. It is diagnostic criterions according to Chicago classification require the occurrence of premature contractions (simultaneous contractions in conventional manometry) in at least 20% of swallows in the presence of normal LOS relaxation pressure (normal integrated relaxation pressure). Premature contractions typically has DL <4.5 s in adults, although paediatric values may be shorter.⁵ Multi-peaked particularly triple-peaked contractions are rare but considered to be salient feature of oesophageal spasm. Elevated DCI is no longer a diagnostic criterion for DES.

Unfortunately, therapeutic interventions are not always effective in treating DES. Medications such as nitrates, calcium channels blockers and tricyclic antidepressants were tried with variable symptomatic response. Peroral endoscopic myotomy is more effective in achalasia than DES and thus not routinely recommended as treatment option. While surgical myotomy was reported in small case series to improve the dysphagia and chest pain in patient with DES, more evidence is required before widespread recommendation.⁵

Different patterns of oesophageal motor disorders were described in adult patients with EoE including hypomotility and hyper motility disorders. Panoesophageal pressurisation appears to be the most common disorder in EoE. Nennstiel *et al* described a complete resolution of oesophageal motility disorders in adult patient with EoE after treating the eosinophilic inflammation. Nurko *et al* have described an abnormal motility patterns in children with EoE studied by four pressure transducer manometry catheter.

To the best of our knowledge, this is the first report of a child with a confirmed DES secondary to EoE with complete clinical and manometric resolution of symptoms after standard EoE treatment.

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