

Eosinophilic Esophagitis: A Case Study of a 12-Year-Old Boy

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ABSTRACT

Eosinophilic esophagitis is an underrecognized chronic condition in which symptoms of gastroesophageal reflux are accompanied by eosinophilia in the esophagus. This disorder affects both children and adults and is associated with a history of other atopic disorders, particularly food allergies. This case study discusses the epidemiology of eosinophilic esophagitis and the proper procedures for diagnosis, referral, and management using the dietary restrictions and pharmacologic treatment included in the clinical recommendations issued by the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition; the American Academy of Allergy, Asthma, and Immunology; and the American College of Gastroenterologists. *J Pediatr Health Care.* (2017) ■, ■-■.

KEY WORDS

Abdominal pain, eosinophilic esophagitis, food allergy, pediatric case study, recurrent vomiting

INTRODUCTION

Eosinophilic esophagitis (EoE) is an inflammatory disease that affects the esophagus. This disorder is characterized by gastroesophageal reflux disease (GERD)-like symptoms and the presence of eosinophils in the esophagus.

Although EoE can affect all populations, most cases occur in males, with a 3:1 male-to-female ratio (Furuta & Katzka, 2015). There is also evidence that supports an association between EoE and other atopic disorders. Up to 60% of children with EoE also suffer from other atopic disorders such as food allergies, atopic dermatitis, allergic rhinitis, and asthma (Soon, Butzner, Kaplan, & deBruyn, 2013). In addition to an association with atopic disorders, there may also be a genetic component, as there is strong evidence of family history in this disorder (Furuta & Katzka, 2015). Most recently, research points to a connection between EoE and connective tissue disorders such as Ehlers-Danlos and Marfan syndrome (Liacouras, Spergel, & Gober, 2014).

The estimated prevalence of EoE ranges from 1 to 4 per 10,000 individuals (Carr & Watson, 2011). In recent years, EoE has been more commonly diagnosed by gastroenterologists and allergists/immunologists; however, the lack of awareness in primary care can lead to missed diagnoses. As many as 1 out of every 10 patients who have been diagnosed with GERD actually have EoE (Liacouras, 2011). This case study discusses a 12-year-old boy with signs and symptoms of EoE and summarizes diagnosis and treatment options.

CASE PRESENTATION

Chief Complaint

A 12-year-old boy and his mother presented to a primary care clinic with complaints of worsening abdominal pain, food getting stuck in his throat, and choking with occasional vomiting.

History of Present Illness

The child's mother reported that the abdominal pain started 1 month ago and was followed by episodes of choking and vomiting. Originally, the mother believed the child had an episode of acute gastroenteritis, and he was kept home from school for a few days and placed

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on a bland diet. The abdominal pain and nausea continued intermittently throughout the day with varying severity, despite rest and diet changes. The patient described the pain as located in the epigastric area radiating to his throat with a resultant sour taste in his mouth. Over-the-counter calcium carbonate and ranitidine were used to attempt to alleviate the discomfort, but neither intervention reduced the pain. He had noticed that the pain worsened after meals. The pain did not cause nighttime awakening.

The child also reported several episodes of choking and vomiting while eating that occurred five times over the past month. He vomited partially digested food during two of the episodes. With the vomiting episodes, the child reported that while eating he felt food suddenly get stuck, which caused him to cough and choke. When questioned about his eating habits, his mother reported that although he eats regular meals, it takes him what she feels is an unusually long time to finish eating. The mother also reported that her son meticulously chews his food and that he takes a drink after every bite. There had been no recent exposure to illness, and his mother reported that his immunizations were up to date.

Review of Systems

No weight loss had occurred as a result of the vomiting, but his mother did report a decreased appetite after the episodes. The child denied hematemesis or bilious vomiting and any episodes of diarrhea or constipation. He stated that he had a soft, formed bowel movement one to two times per day with no hematochezia reported. The child denied a history of cough, wheezing, and shortness of breath. No new or recent rashes or lesions were reported.

Past Medical History

The patient had a history of atopy, primarily food allergies (peanuts and tree nuts), and atopic dermatitis. He had an allergic reaction, which included generalized hives and some wheezing, to a peanut butter cookie at 1 year old. He was treated in the emergency department and has avoided peanuts and tree nuts since the episode. His mother reported that allergy testing was never done. There was no history of any prior gastrointestinal disorders. At birth

the child was circumcised and underwent a right orchiopexy at 16 months of age. His medications included mometasone 0.1% ointment as needed for atopic dermatitis, cetirizine 10 mg as needed for allergen exposure, and injectable epinephrine as needed for anaphylaxis.

Family History

A positive family history of atopy was reported. The child's mother reported having food (peanuts and tree nut) and environmental allergies and that his father had a history of asthma. The boy had one sibling who also has food allergies (peanuts) and atopic dermatitis. The mother explained that she has dealt with reflux symptoms most of her adult life and currently takes over-the-counter ranitidine on occasion for heartburn. There is a history of GERD noted in both maternal and paternal grandparents.

Personal/Social History

The child lived at home with mother, father, and a younger sister. There were no pets living in the home, and his parents took special care to avoid his allergen triggers. The mother reported having a good support system at home, with both sets of grandparents living nearby. The child attended school regularly and got along with his peers. The mother reported that school had a special peanut-free area for students in the cafeteria and that they had an allergy action plan and injectable epinephrine available. The child also carried his injectable epinephrine with him outside of school for emergencies but had never had to use it. The child denied the use of tobacco, alcohol, or illegal drugs.

Physical Examination

Upon examination, the child was well appearing and was not in acute distress. Vital signs were age appropriate. He was alert and oriented and responded appropriately to questions. No wheezes, rubs, or crackles were auscultated. The child's abdomen was soft, non-distended, and without lesions or masses. No pain or guarding was elicited with deep and light palpation. No hepatomegaly or splenomegaly was noted. Strength and sensation were equal in the bilateral upper and lower extremities.

CASE STUDY QUESTIONS

1. What differential diagnoses would you consider for this child?
2. What would be included in your initial treatment plan for this child?
3. After an 8-week trial of a proton pump inhibitor (PPI), the patient returns to the office with no improvement in symptoms. How does this change your treatment plan?
4. What are the diagnostic criteria for EoE?
5. What are the clinical recommendations for the management of EoE?

CASE STUDY ANSWERS

1. What differential diagnoses would you consider for this child?

When a child presents with symptoms of gastroesophageal reflux, causes of esophagitis should be considered in the differential diagnoses. Causes of gastroesophageal reflux symptoms and esophagitis in adolescent patients can include GERD, EoE, Crohn's disease, bulimia, pill-induced esophagitis, caustic ingestion, Barrett's esophagus, graft-versus-host disease, connective tissue disorders, bullous skin diseases, lymphoma, and infections with pathogens such as *Candida albicans*, Herpes simplex, and *Cytomegalovirus* species (Vandeplas et al., 2009). Because episodes of vomiting are also a concerning symptom, differentials for vomiting should also be considered. These include gastroparesis, gastroenteritis, food allergy, EoE, intracranial hemorrhage or mass, metabolic disorders, ingestion of toxic substances, self-induced vomiting, and child abuse or neglect (Vandeplas et al., 2009). Although many of these differentials would require further diagnostic testing to rule out, the history and physical examination of the patient suggest beginning with gastrointestinal causes, especially those associated with esophagitis.

2. What would be included in your initial treatment plan for this child?

Because differentiation between EoE and GERD can be difficult initially, the first step in treatment is to rule out GERD with a PPI trial. According to the clinical practice guidelines released by the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN; Vandeplas et al., 2009), the initial treatment plan for a child presenting with symptoms of gastroesophageal reflux would be to initiate lifestyle changes and begin pharmacologic therapy with an acid-buffering agent, mucosal surface barrier, or gastric antisecretory agent (Vandeplas et al., 2009). Two of the most commonly prescribed drug classes for GERD are histamine-2 receptor antagonists and PPIs. According to the NASPGHAN 2009 practice guidelines for GERD, PPIs are superior in providing symptom relief of erosive esophagitis and GERD symptoms (Vandeplas et al., 2009). This recommendation accompanied by the patient's history of atopy would be reason to begin medication therapy with a PPI. Omeprazole, lansoprazole, and esomeprazole are all approved for use in children and would be appropriate choices for treatment (Vandeplas et al., 2009).

Lifestyle changes appropriate for adolescents include avoidance of triggers such as smoking, caffeine, and acidic foods, elevating the head of the bed, and left lateral positioning while sleeping (Vandeplas et al., 2009). Although these recommendations have been thoroughly studied only in adults, it is assumed that they will help relieve symptoms in the adolescent

population as well. Foods such as caffeine, chocolate, and acidic foods are common triggers and should be eliminated or avoided if they provoke symptoms in children (Vandeplas et al., 2009). Children should also avoid tobacco smoke exposure because this can also be an aggravating factor (Vandeplas et al., 2009). The current NASPGHAN guidelines recommend a 4-week lifestyle medication and PPI trial in children and adolescents and children with chronic reflux symptoms (Vandeplas et al., 2009), but the American Academy of Allergy, Asthma and Immunology and NASPGHAN guidelines on the management of EoE state that a PPI trial of 6 to 8 weeks is useful before referral to a specialist for a biopsy (Greenhawt, Aceves, Spergel, & Rothenbert, 2013; Papadopoulou et al., 2014).

3. After an 8-week trial of a PPI and the recommended lifestyle changes, the patient returns to the office with no improvement in symptoms. How does this change your treatment plan?

If symptoms persist after a 6- to 8-week trial of a PPI, or symptoms recur after discontinuing the medication, referral should be considered (Liacouras, 2011). The patient's history of atopy, food allergies, and persistent symptoms despite a PPI trial should all be directing the primary care provider to the diagnosis of EoE. Patients who are suspected of having EoE will benefit from a multidisciplinary approach. The role of the primary care provider at this point is to recognize the signs and symptoms of the disease and make the correct referrals. Referrals to both an allergist and a gastroenterologist are necessary and important as EoE is often co-managed by both specialties because of the complexity of the disease (Greenhawt et al., 2013).

4. What are the diagnostic criteria for EoE?

GERD symptoms accompanied by a history of atopy and PPI trial failure should make a clinician highly suspect a diagnosis of EoE. Unfortunately, EoE cannot be easily diagnosed by relying on physical examination results or laboratory test values. A clinician must therefore rely on symptoms and esophageal assessment and biopsy via endoscopy for diagnosis (Carr & Watson, 2011). According to the guidelines on diagnosing and managing EoE released by the American College of Gastroenterologists, the only current reliable tool for diagnosis of EoE is an endoscopic biopsy (Dellon et al., 2013). To confirm diagnosis, two to four biopsy samples should be obtained from different locations of the proximal and distal esophagus (Papadopoulou et al., 2014). These biopsy samples should preferably be obtained after an 8-week trial of a PPI. Although eosinophils are typically seen throughout the gastrointestinal tract in healthy children, they are usually absent in the esophagus (Carr & Watson, 2011). According to the Management Guidelines of

Eosinophilic Esophagitis in Children released by NASPGHAN, 15 or more eosinophils must be seen per high power field in at least one of the esophageal biopsy samples (Papadopoulos et al., 2014).

Although a PPI trial is not mandatory before esophageal biopsies are obtained, NASPGHAN guidelines recommend doing a second biopsy after starting a PPI on children whose biopsies were performed before a PPI trial was initiated. Diagnosis can then be confirmed with a second esophageal biopsy, and other differentials can also be confidently identified or ruled out (Papadopoulos et al., 2014). Endoscopic assessment of the esophagus is also a useful tool in the diagnosis of EoE. Endoscopic findings that closely correlate with the diagnosis of EoE include linear furrowing, concentric rings, Schatzki ring (a narrowing of the lower esophagus), white exudate, small-caliber esophagus, and linear mucosal tears with the introduction of the endoscope (Carr & Watson, 2011). Endoscopic assessment should not be used alone because in up to 30% of patients with EoE, the esophagus can appear completely normal without any of the aforementioned findings (Vandeplas et al., 2009).

5. What are the clinical recommendations for the management of EoE?

There are two agreed-on treatment options for management of EoE: trigger avoidance through elimination diets and pharmacologic treatment with corticosteroids (Carr & Watson, 2011). Treatment can begin with either dietary restrictions or topical corticosteroids and depends upon patient and family preference (Greenhawt et al., 2013). Decisions regarding treatment should be discussed with the patient and the parents to determine which therapy would have the best adherence in their particular situation. Treatment goals not only include symptom relief but also focus on inflammation control and restoration and preservation of esophageal function (Furuta & Katzka, 2015).

Pharmacologic Management

The mainstay of pharmacologic treatments for EoE is topical corticosteroids. Fluticasone is available as a meter-dose inhaler that is puffed directly into the mouth and can be gulped or swallowed (Dellon et al., 2013). Budesonide is available as a nebulizer solution that can be mixed with a sugar substitute to create a slurry called viscous budesonide. After mixing, the viscous budesonide is swallowed and coats the esophagus (Dellon et al., 2013). Patients should not eat or drink for 30 minutes after applying the topical steroids (Carr & Watson, 2011). Both swallowed fluticasone and viscous budesonide have been shown to improve symptoms and control inflammation and are both considered first-line treatments (Dellon et al., 2013). A follow-up endoscopic examination should be done after 6 to 8 weeks of therapy to ensure histologic response along with symptomatic relief (Carr &

Watson, 2011). Systemic corticosteroids are also effective in the treatment of EoE, but because of the chronic nature of the disease, there is risk of complications due to long-term steroid use (Dellon et al., 2013). Other medications are also being studied for potential treatment options in the future and include mast-cell inhibitors, leukotriene inhibitors, reslizumab, and omalizumab (Dellon et al., 2013).

Dietary Management

Dietary elimination of triggers is accomplished by one of three diet choices: using a total elimination diet, targeted elimination diet, or empiric elimination diet (Greenhawt et al., 2013). The total elimination diet consists of being fed solely by an elemental or amino acid-based formula (Greenhawt et al., 2013). Although the elemental formula diet has been found to be the most effective, it can also greatly diminish the quality of life. The formula is very costly and can require the placement of a feeding tube for administration (Dellon et al., 2013). Although this may be feasible for some infants and younger children, it is often not a good choice for older children and adolescents. Targeted and empiric elimination diets are often tried before using elemental formula (Carr & Watson, 2011). The targeted elimination diet uses allergy testing by skin prick test or atopy patch test to determine which foods may be causing symptoms (Greenhawt et al., 2013). These foods are then eliminated from the diet. The empiric elimination diet eliminates the six most common food allergens (milk, egg, soy, wheat, peanut/tree nuts, fish/shellfish) from the patients' diet without any skin prick or blood testing. Both targeted elimination and empiric elimination have been shown to be effective for approximately 75% of patients with EoE (Carr & Watson, 2011).

With all three elimination diets, reintroduction of foods is required 4 to 8 weeks after remission to determine which foods are eliciting the symptoms (Dellon et al., 2013). Foods are gradually reintroduced one at a time from least allergenic foods to most allergenic foods (Papadopoulos et al., 2014). A repeat endoscopic examination should be done after the reintroduction of every three to five foods to ensure that there is no recurrence of inflammation (Carr & Watson, 2011). If recurrence of symptoms occurs after reintroduction of a food, it may need to be eliminated indefinitely (Papadopoulos et al., 2014).

Dietary restrictions and daily medications can present difficulties in everyday life for patients suffering from EoE. Nonadherence rates of medication therapy were found to be around 30% in a study conducted by Hommel, Franciosi, Hente, Ahrens, and Rothenberg (2012). Dietary therapy nonadherence rates were reported to be slightly higher, at around 33% (Hommel et al., 2012). It is very important for health care providers to regularly assess how treatment plans are working and

determine any barriers that may be in place due to the high nonadherence rates and chronic nature of EoE. It is also important to develop an alternative plan if adherence to a particular therapy is not conducive to the patient's lifestyle.

Other Management Considerations

Esophageal dilation is also an accepted form of treatment for EoE, but it does not address the cause of the inflammation (Carr & Watson, 2011). It is most often reserved for treatments of adults or adolescents or for those patients with remodeling of the esophagus that persists after therapy (Carr & Watson, 2011).

Recent research suggests that there is also a major psychosocial component to EoE, with 69% of children with the disorder reporting difficulties with sleep, school, anxiety, depression, and with social interactions (Liacouras et al., 2014). In addition, another study found that caregivers of children with EoE reported stress from caring for their child (Liacouras et al., 2014). When taking these studies into consideration, a psychosocial evaluation of these children, along with support for the family and caregivers, is a necessity in the management of EoE (Liacouras et al., 2014).

CONCLUSION

EoE is a chronic, recurring disease affecting the esophagus in which inflammation and symptoms are likely to recur with cessation of treatment. Although there have been some reports of spontaneous remission in some patients, EoE is most likely a lifelong condition. Maintenance therapy of topical corticosteroids or dietary restrictions should be considered for all patients to prevent symptom recurrence (Dellon et al., 2013). Complications include esophageal strictures, perforation, and malnutrition (Furuta & Katzka, 2015). There have been no reports of malignancy as a result of EoE (Furuta & Katzka, 2015). The primary care provider plays an essential role in the recognition and diagnosis of EoE. By recognizing risk factors, signs, and symptoms and

providing support through a multidisciplinary team with referral to allergy, gastroenterology, psychology, and dietary/nutrition specialists, the primary care provider is ensuring the best possible outcomes when it comes to long-term management goals for patients with EoE.

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