An Adolescent with Vomiting and Weight Loss

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Citation:

1. Abstract
Pediatric Achalasia is a rare condition in children, often presents with common symptoms of vomiting and weight loss. We present a case of a 15-year-old female with persistent vomiting and weight loss who was initially diagnosed with eosinophilic esophagitis (EoE) due to endoscopic findings and elevated eosinophils on esophageal histology. After clinical worsening with EoE therapy, a broader workup showed classic bird beak on upper gastrointestinal series and subsequent esophageal manometry confirmed a diagnosis of achalasia. Our case report reinforces the need to review the differential diagnosis of esophageal eosinophilia from common disorders such as gastroesophageal reflux disease and EoE to rare conditions such as achalasia.

2. Case Report
A 12-year-old previously healthy African-American female presents to the Pediatric Gastroenterology Clinic for a two-month history of non-bloody, non-bilious vomiting, and unintentional weight loss. These episodes of vomiting occurred up to eight times each day without relation to mealtimes. She tolerated liquids well but had difficulty swallowing solids. She reported associated fatigue, nausea, and decreased activity. She denied heartburn, acid brash, chest pain, epigastric pain, chronic cough, recurrent throat clearing, excessive water intake to help with swallowing, or neurologic symptoms. She sought medical care at the local emergency department multiple times and was prescribed a proton pump inhibitor (PPI) and ondansetron, without symptomatic improvement. Pediatric psychiatry was consulted to evaluate for clinical depression and possible eating disorder. Physical exam was remarkable for a weight of 36.3 kg (11th percentile), and a documented 25-pound weight loss over the past 6 months. The remainder of the examination was benign, without any focal findings. Laboratory evaluation, including complete blood count with differential, celiac panel, hepatic panel, amylase, lipase erythrocyte sedimentation rate, and c-reactive protein were within normal limits. An upper endoscopy (EGD) revealed linear furrowing in the esophagus with mucosal biopsies, consistent with active esophagitis with 2-10 eosinophils/high powered field (hpf) in the distal and 1-10 eosinophils/hpf in the proximal esophagus. The LES was able to be traversed without resistance. She was treated for a diagnosis eosinophilic esophagitis (EoE) instead of gastro-esophageal reflux disease (GERD) due to her pre-treatment with PPI and the increased eosinophils in the proximal esophagus. Her treatment included fluticasone, 2 puffs swallowed BID, omeprazole 20 mg administered orally once daily, and an empiric milk protein-free diet.

Two after treatment initiation the patient was hospitalized for fever and abdominal pain. A computed tomography (CT) scan of the abdomen showed esophageal dilation with a narrowing of the distal esophagus/gastro-esophageal junction. An esophagram demonstrated the classic beaked distal esophagus found in achalasia (Figure 1). Repeat EGD revealed significant esophagitis and white residue. Histopathology was consistent with a diagnosis of candida esophagitis and she was prescribed fluconazole. An esophageal manometry study was conducted, revealing normal resting pressure of the lower esophageal sphincter (LES), increased residual pressure of 15.6mmHg, and an absence of peristaltic contractions with wet swallows, consistent with a diagnosis of achalasia.

Ultimately, she underwent a laparoscopic Heller myotomy with Dor fundoplication. Post-operative imaging showed improvement in the dilatation of the esophagus. A follow-up nine months post-operatively showed complete resolution of emesis and dysphagia. Her weight had stabilized at 52.9 kg (58th percentile).
3. Discussion

Achalasia has a reported global prevalence of 12.6 per 100,000 persons per year, with 20,000 to 30,000 cases per year in the United States and a peak incidence between 30-60 years of age [2]. Pediatric achalasia is even rarer, with less than 5% of patients reporting symptoms before 15 years of age [3]. The etiology of achalasia is thought to be due to a loss of inhibitory neurons in the Myenteric plexus in the distal esophagus and the LES.

Clinical manifestations of achalasia in children and adolescents include vomiting, dysphagia, chronic cough, and weight loss. The symptoms of achalasia often mimic other more common disease processes, including GERD, EoE, Helicobacter pylori gastritis, peptic ulcer disease, inflammatory bowel disease, celiac disease, asthma, thereby delaying the diagnosis [4]. In adolescents, it is also crucial to consider psychiatric disorders, including anorexia/bulimia and depression, only after anatomical/metabolic causes have been ruled out.

The patient presented in this report was treated for depression, GERD, and EoE before being diagnosed with achalasia. In fact, up to half of the patients diagnosed with achalasia report initial treatments with antihistamines, histamine 2-blockers, or PPIs [4]. In one case, a boy was treated for GERD for 10 years before the diagnosis of achalasia was made [3]. Another subset of achalasia cases presenting as dyspnea on exertion and nocturnal cough were treated as asthma [5]. Thus, it is unsurprising to note that our patient’s time to diagnosis was similar to the 7 to 31 months reported in the literature [6].

Early imaging results which may suggest the diagnosis of achalasia include chest x-ray findings showing the absence of air in the stomach, dilated esophagus with air-fluid levels, and an esophagram with dilation and beaking at the gastro-esophageal junction. EGD findings in achalasia may show a dilated esophagus that contains residual food or fluid, sometimes in large quantities, and there may also be some difficulty in the endoscope traversing the LES. While esophagrams have high sensitivity (>90%) and positive predictive value (96%) for the diagnosis of achalasia, the gold standard test is esophageal manometry demonstrating a lack of esophageal peristalsis.

Achalasia has no defined cure and treatment includes symptomatic management with pneumatic dilation and esophageal myotomy. Less effective medication therapy includes botulinum toxin, nitrates, silde-modaffil, and calcium channel blockers. Our patient underwent the Heller myotomy with Dor fundoplication, which has a 90% short-term success rate [10]. Complications of the procedure include recurrent dysphagia, aspiration risk, and GERD.

In our patient, given her clinical worsening after EoE therapy and diagnostic confirmation of achalasia it is unlikely that she dysphagia due to EoE. Presumably, her eosinophilic infiltration of the esophagus occurred as a result of achalasia with chronic food stasis in the esophagus and subsequent inflammation of the esophageal mucosa. Patients with persistent esosis and esophagitis who do not respond to therapies for GERD or EoE should prompt revisiting the differential diagnosis for esophageal eosinophilia such as achalasia.

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5. Author Contributions

JA wrote the manuscript and participated with PDW in revising the manuscript critically for important intellectual content. All authors approved the final version of the manuscript to be published.

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