Retrograde Gastroesophageal Intussusception: Initial Presenting Feature of Achalasia in a Teenager

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This is the author's manuscript of the article published in final edited form as:

Morocho, B., Bose, P., Grayson, B., Croffie, J. M., & Billmire, D. F. (2019). Retrograde gastroesophageal intussusception: Initial presenting feature of achalasia in a teenager. Journal of Pediatric Surgery Case Reports, 50, 101300. https://doi.org/10.1016/j.epsc.2019.101300

Abstract

A 16-year-old Caucasian male presented with acute vomiting and dysphagia. Imaging studies revealed retrograde gastroesophageal intussusception (RGEI), which reduced prior to diagnostic laparoscopy. No clear etiology for RGEI was identified at that time, so further surgical intervention was deferred. He returned several months later with persistent dysphagia. Imaging, endoscopy, and endoluminal function imaging probe then diagnosed achalasia. He underwent a second laparoscopy for Heller myotomy and Dor fundoplication. This is the first report of RGEI preceding a diagnosis of achalasia.

Keywords: retrograde esophageal intussusception, achalasia, Heller myotomy

Retrograde gastroesophageal intussusception (RGEI)[1] is a rare condition. Occurrence has been associated with achalasia and history of manipulation of the gastroesophageal (GE) junction. We present an interesting case of an adolescent male who presented with emesis and dysphagia and was found to have RGEI. This intussusception preceded a diagnosis of achalasia.

Case Report

A 16-year-old Caucasian male presented with persistent forceful, nonbilious emesis and dysphagia that began two weeks earlier. He noted a history of nocturnal emesis, heartburn, dysphagia to solids and liquids, and a 6-kg weight loss. He denied long term dysphagia. Two days prior to his presentation, he developed hematemesis. He was empirically treated by his Primary Care Provider with famotidine and ondansetron without improvement. His past medical history was remarkable for juvenile myoclonic epilepsy managed with valproic acid and lamotrigine. His surgical history included hypospadias repair, bilateral inguinal hernia repair, and tonsillectomy. Family history was negative for any chronic gastrointestinal disorders. Social history was remarkable for strength training and negative for smoking or illicit drug use. Upon hospital admission, his vital signs were stable, and his abdominal exam was unremarkable.

A barium esophagram showed complete obstruction in the mid to distal esophagus, from what was suspected then to be a food impaction (Figure 1). Upper endoscopy revealed a partially obstructing, circumferential mass in the lower third of the esophagus (Figure 2). The endoscope was advanced on one side of the lesion into the stomach. Upon entering the stomach, the mass appeared to arise from gastric mucosa. No biopsy was attempted due to bleeding concerns.

CT angiography of the chest and abdomen post-endoscopy revealed a gastroesophageal intussusception with the gastric fundus within the esophageal lumen (Figure 3). He was taken to the operating room for diagnostic laparoscopy and endoscopy. After induction of general anesthesia, an

[1] RGEI = retrograde gastroesophageal intussusception

orogastric tube was passed easily to a depth of 55-cm with return of dark fluid. Laparoscopic evaluation revealed the stomach within the peritoneal cavity and the RGEI was reduced. The hiatus was of normal size. The stomach appeared pink and well perfused. The fundus was found to be anatomically normal with a thickened wall and serosal petechiae. Intraoperative endoscopy showed that the esophageal mass was no longer present. No lead points or masses were found, and the ligamentous attachments of the stomach were also normal. No further surgical interventions were done as a clear etiology for the intussusception could not be found. He was discharged home on a regular diet 4 days post operatively.

Seven months later, he was hospitalized again for acute throat pain, dysphagia, and nonbilious, nonbloody emesis. He endorsed monthly episodes of dysphagia and emesis of chewed food contents since his initial evaluation. Repeat CT scan of his chest showed a dilated esophagus without evidence of RGEI. Repeat esophagram showed a dilated esophagus with minimal contrast passing through the GE junction (Figure 4). He had a second upper endoscopy that showed significantly dilated esophagus and inflamed mucosa at the GE junction. High resolution esophageal manometry showed increased mean residual LES pressure of 23.3mmHg (normal < 15mmHg). No esophageal peristalsis was visualized for 90% of his instructed swallows. These findings were suggestive of achalasia type 2 with 40% panesophageal pressurization. He was referred for endoluminal functional imaging probe (Endoflip), where the narrowest GE junction cross sectional area (CSA) at 20ml, 30ml, and 40ml distension were measured as 20mm^2, 19mm^2, and 27mm^2 respectively (normal medians of 38mm^2, 94mm^2, and 264mm^2 respectively). GE junction distensibility was impaired with a distensibility index (CSA/intrabag pressure) at 50ml distension of 1.5mm^2/mmHg (normal >2.9mm^2/mmHg), which supported the diagnosis of achalasia.

The family agreed to proceed with surgical management for achalasia with a laparoscopic Heller myotomy and Dor fundoplication. Intraoperatively, the hiatus showed evidence of prior inflammation with subsequent adhesions. A 6-cm myotomy was performed, extending 2-cm onto the gastric wall, with

endoscopic confirmation that the lower esophageal sphincter opened freely with insufflation. An anterior 180-degree Dor fundoplication was performed with two parallel rows of stitches affixing the fundoplication to the myotomy and the crura, the latter at the superior most sutures. The patient was discharged home on a full liquid diet on post-operative day 1. He reported resolution of vomiting with meals on follow up 1 month post operatively. He continued to report intermittent heartburn, for which he takes a pantoprazole 40mg once daily, and mild dysphagia to solid foods like meat.

Discussion

This teenage boy experienced RGEI prior to diagnosis and treatment of achalasia, which is very unusual from a physiologic standpoint. Achalasia is an esophageal disorder involving high resting LES pressure, impaired esophageal peristalsis, and impaired LES relaxation.[1]. We posit that increased intraabdominal pressure associated with vomiting overcame the high LES sphincter tone, which allowed for the stomach to move retrograde into the esophagus. However, due to high LES tone, the stomach remained trapped in the esophagus unable to reduce back to its normal position thereafter.

Of the reported cases of RGEI in humans, a majority have occurred in patients with previously treated achalasia. It is proposed that treatments for achalasia increase risk of RGEI, since the GE junction is enlarged.[2] In one reported case, a 22-year-old male with achalasia and history of peroral endoscopic myotomy suffered a RGEI, and ultimately was managed with reduction and anterior gastropexy.[3] In another case, a 43-year-old male with a history of achalasia with esophageal dilation required esophagogastric myotomy and Nissen fundoplication for RGEI.[4] In addition to interventions for achalasia, Gowen et al.[5] proposed risk factors for RGEI including eating disorders, duodenal obstruction, physical exertion, peptic ulcer disease, and hyperemesis gravidarum in pregnancy. A 40-year-old woman with a history of Heller myotomy and Dor fundoplication for achalasia presented with RGEI from hyperemesis gravidarum.[6] Only one case of RGEI has been reported in a child without an existing esophageal condition. Lukish et al.[7] reported on a 3-year-old previously healthy child with abdominal

pain and vomiting, who had evidence of esophageal obstruction on esophagram and a subsequent CT showing an RGEI. He underwent trans-thoracic esophagomyotomy for management.

To the best of our knowledge, this is the only case of a patient who was diagnosed with achalasia after occurrence of RGEI, and not as a consequence of treatment for achalasia. The finding of normal anatomy at laparoscopy with negative history of symptoms at initial presentation provided the rationale for avoiding additional procedures such as fundoplication or gastropexy at that time. Closer follow up after his initial presentation, with consideration for esophageal manometry or esophagram might have allowed earlier diagnosis of achalasia which is a recognized co-morbidity of this rare problem.

Additional Information

Patient Consent: This report does not contain any information that could lead to identification of the patient, therefore consent was not obtained.

Funding: No funding or grant support

Authorship: All authors attest that they meet the current ICMJE criteria for Authorship.

The following authors have no financial disclosures: BM, PB, BG, JMC, DB

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Figures

Figure 1. Esophagram at presentation
Description: Barium esophagram with mid to distal esophageal obstruction

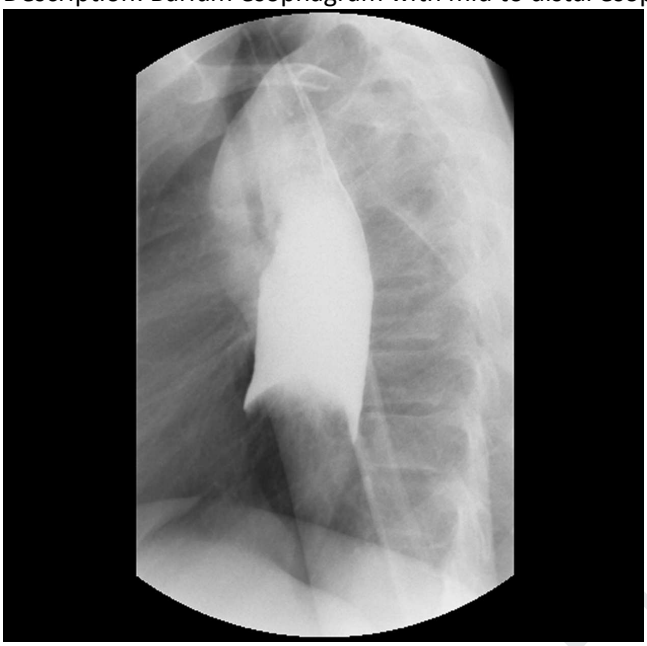


Figure 2. RGEI from endoscopic perspective
Description: Endoscopic images of gastric mucosa intussuscepting into the distal esophagus

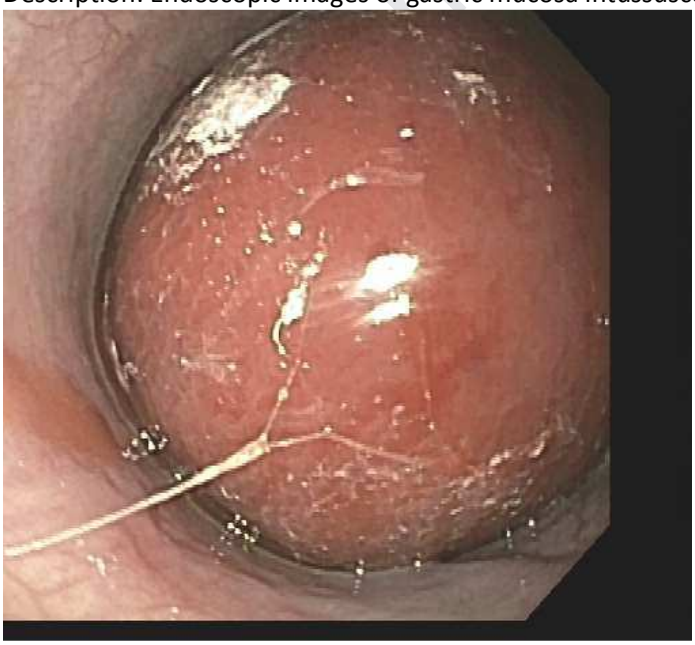


Figure 3. RGEI on CT
Description: CT scan showing RGEI; coronal (top) and sagittal (bottom)

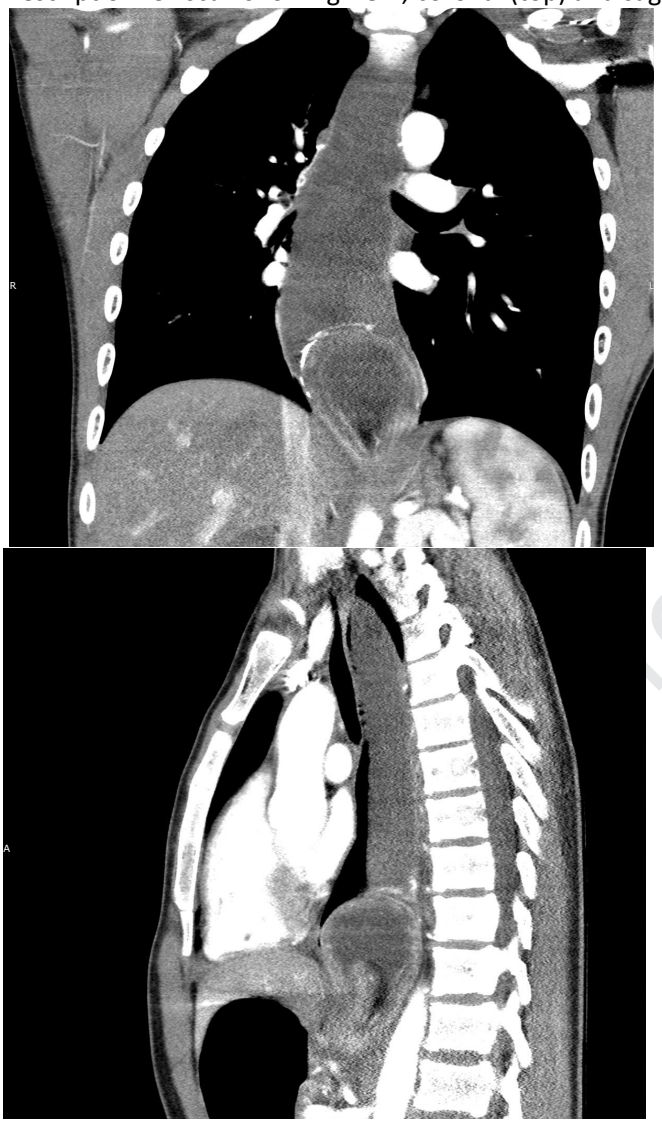
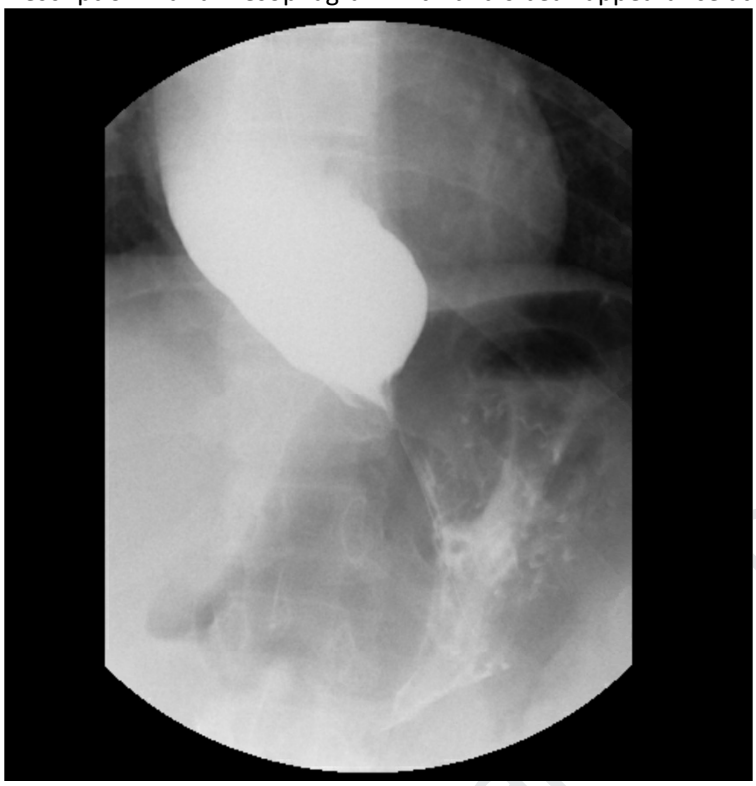


Figure 4: Esophagram at second presentation
Description: Barium esophagram with bird's beak appearance at GE junction



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Conflict of Interest

The following authors have no financial disclosures: BM, PB, BG, JMC, DB