CLINICAL VIGNETTE

End-Stage Achalasia Complicated by Massive Esophageal Bezoar Saved by Rigid Esophagoscopy

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Case

A 90-year-old male with a long history of achalasia and multiple medical comorbidities, presented with aspiration pneumonia and worsening dysphagia for four days. The patient was initially diagnosed with achalasia over 40 years ago which was confirmed by manometry at an outside hospital. He underwent a Heller myotomy without fundoplication over 15 years ago, at which point he reports that his esophagus was already "sigmoid-shaped." Post-operatively, there was a temporary relief of symptoms, but they slowly returned, requiring repeated upper endoscopies over the years. Prior procedures included multiple pneumatic dilations, botox injections, and disimpactions, the last one over a year ago. Surgical options including Redo myotomy versus total esophagectomy had been discussed at in the past. The patient was non-compliant with a recommended pureed diet. He reported multiple episodes of self-induced vomiting when his symptoms progressed.

He presented to the ED after attempts of self-vomiting were no longer helping. He had been recently admitted for aspiration pneumonia, COPD exascerbation and atrial fibrillation one month prior. He was discharged home on oral antibiotics, steroids, and diuretics, with persistent respiratory symptoms after discharge. Vital signs included temperature of 36.8, BP 129/95, HR 99, and oxygen saturation of 97% on room air. His physical exam was remarkable for crackles in the left anterior chest and bilateral lower lung fields. His weight on presentation was 63.5 kg which was stable from a month prior. Labs revealed a mild leukocytosis of 10.03 x10E3/uL (ref 4.16-9.95 x10E3/uL), stable normocytic anemia with hemoglobin of 11.2 g/dL (ref 13.5 – 17.1 g/dL), notable albumin of 2.7 g/dL (ref 3.9 – 5.0 g/dL), otherwise normal platelets, electrolytes, liver function tests, and cardiac enzymes.

A computed tomography (CT) scan of the thorax showed a massively dilated patulous esophagus completely filled with solid material through cervical esophagus (see Figure 1). A chest x-ray from his prior hospitalization is included for comparison. (see Figure 2). In retrospect, this is likely what caused his first episode of aspiration with subsequent lack of improvement due to poor oral medication delivery. Conservative management with nasogastric tube (NGT) decompression failed due to clogging. He was kept nothing per oral (NPO) for the next 3 days, however repeat CT scan showed no improvement.

At this point, given a stable cardiovascular status and worsening nutritional status, he was scheduled for endoscopic decompression on day four of hospitalization. General anesthesia assisted with intubation and airway protection. Upon entry past the upper esophageal sphincter (UES), a large column of food debris was encountered, consistent with a massive esophageal phytobezoar. We attempted to gently pass the scope around this to find the gastroesophageal junction (GEJ), but at 40cm, the scope started looping and the GEJ could still not be found. An overtube was placed and attempted suctioning through a large oral gastric tube was unsuccessful. Previous reports have described using large volume saline lavage and suctioning to help remove retained food. However, we did not feel comfortable injecting a large volume of saline given the proximity of the bezoar to the UES and his poor pulmonary reserve should he suffer any further aspiration.

Over three hours were spent making multiple passes with a large Roth net to remove pieces of the large phytobezoar. Continued attempts with flexible endoscope likely would have taken an additional 12-24 hours. The patient remained intubated and thoracic surgery was consulted to assist with decompression using a rigid esophagoscopy given its larger therapeutic channel (up to 9mm versus 3.7mm). In the operating room, the rigid endoscope was able to slowly remove more food with forceps and suctioning. At 42cm, the GEJ still could not be found, but enough food had been cleared to allow easier visualization with a flexible endoscope. Re-evaluation with a flexible endoscope found a widely patent lower esophageal sphincter amongst the remaining debris.

At this point, additional attempts to fully clear the esophagus seemed futile. The best option for the patient was to place a percutaneous endoscopic gastrostomy (PEG) tube to ensure adequate nutrition and medication delivery going forward. The patient was then extubated without complication and tolerated tube feeds by the next day. He is not a candidate for esophagectomy given his age and comorbidities. Repeat attempts at myotomy or PD are likely futile given his retention is mostly secondary to aperistalsis and esophageal deformity. If his esophagus is able to clear of food over time, reintroduction with a liquid (or blenderized at most) diet for pleasure may be considered.

Discussion

We present a unique and extreme case of end-stage achalasia leading to mega esophagus with massive esophageal food retention forming a phytobezoar.

Achalasia remains the most described primary motility disorder of the esophagus. Despite definitive therapeutic procedures such as myotomy and serial pneumatic dilations, 10-15% of patients will still have progression of disease.[1] This can lead to end-stage achalasia often described in literature as mega esophagus—a massively dilated esophagus—or a sigmoid shaped esophagus.[2] When this happens, all options should be exhausted before recommending total esophagectomy, which up to 5% of patients will end up requiring.[3] Our patient had already underwent myotomy, multiple dilations, and botox injections, none of which would have provided any benefit in this setting.

In end-stage achalasia, the esophagus is aperistaltic and essentially paralyzed. Food retention can be common from aperistalsis and the distorted shape regardless of LES pressure. To our knowledge, this is the first report of esophageal food retention leading to such massive phytobezoar extending the entire length of the esophagus. There are a lack of reports and guidance on how to management such extreme complications encountered in end-stage patients such as this one.

Esophageal bezoars are rare and mostly described in case reports of primary esophageal dysmotility such as achalasia.[4,5] Therapeutic management can be challenging short of surgery, especially for severe cases extending the length of the esophagus. Esophagectomy was considered, as it could have treated his massive esophageal bezoar along with his end-stage achalasia. However given his age and comorbidities, he was not a good candidate for a surgery that carries a high risk of post-operative complications.

We initially attempted decompression using a flexible endoscope, which ultimately proved futile, limited by the small size of the instrument channel (3.7mm). Our surgical colleagues, using a rigid esophagoscopy were more successful with decompression. Gastroenterologists are no longer trained in rigid endoscopy, which is most commonly used for esophageal foreign body removal. Esophagoscopes have a larger therapeutic instrument channel (up to 9mm) and is equally, if not more, effective than flexible esophagoscopy. However, rigid procedures require general anesthesia, have greater perforation risk (up to 3%), and more frequent post-procedure dysphagia.[6,7] Thus in general, esophagoscopy use is reserved for cases that fail initial attempts with flexible endoscopy.

In our case, the rigid endoscope allowed easier suctioning and removal of larger pieces of food. This slow and tedious task allowed eventual advancement of the rigid esophagoscope to near the GEJ. Being able to switch back to a flexible endoscope at the appropriate time allowed for identification of the GEJ and advancement into the stomach. Lastly, this allowed placing an endoscopic gastrostomy tube, avoiding need for laparoscopic tube placement.

This was an extreme manifestation of end-stage achalasia and how to manage a massive esophageal bezoar if surgery is not an option. Flexibility and ability to transition between flexible and rigid endoscopies was paramount.

Figures

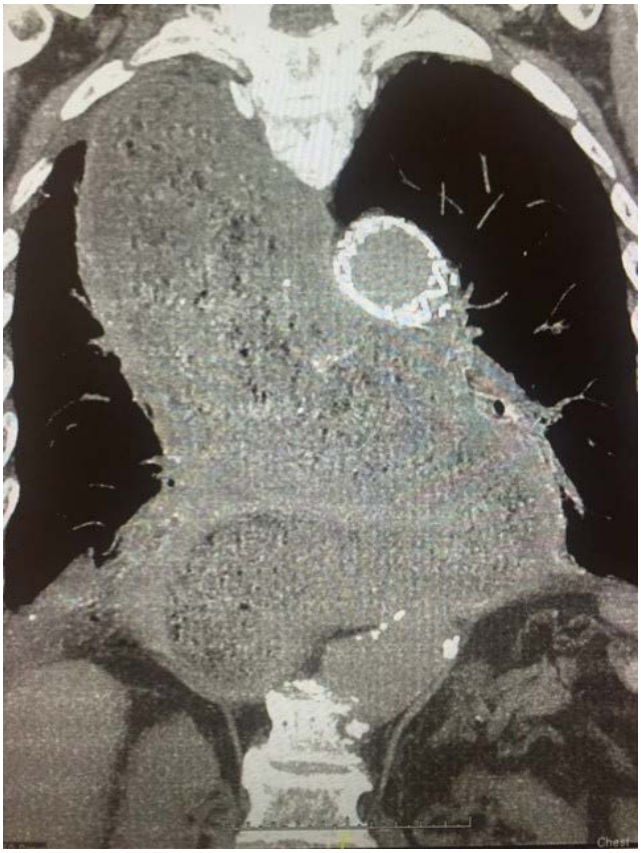

Figure 1. CT chest showing massively dilated esophagus with retained food involving entire esophageal lumen.

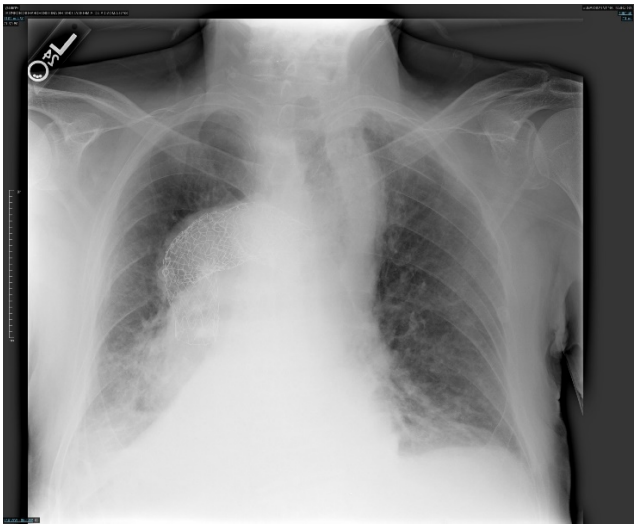
Figure 2. CXR from prior admission

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