Gastroesophageal Surgery Case Scenario

A 71-Year-Old Woman with Achalasia

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PRESENTATION OF CASE

A 71-year-old woman with longstanding history of episodic non-bilious projectile vomiting (mostly undigested food and saliva) presented to Massachusetts General Hospital given the worsening of her symptoms and food intolerance during the previous six months. Over that time frame, she noted a sensation of food becoming stuck in her throat and progressive difficulty swallowing. This difficulty was initially related to solids and then progressively to liquids, which severely limited her food intake. She lost 16 pounds since the symptoms started to worsen.

Her past medical history was notable for hypothyroidism, hypertension and depression. Her medications included thyroid medication, vitamins and omeprazole. She lived alone and did not smoke or drink alcohol. Her family history is non-contributory.

She underwent an extensive workup, where an esophagogastroduodenoscopy (EGD) showed dilated lower third of the esophagus and a benign-appearing stricture. The barium swallow demonstrated dilated mid-distal esophagus with stenosis at the gastroesophageal junction (Figure 1), and the esophageal manometry study was confirmatory for Chicago classification Type II achalasia, a disease where the esophagus does not function properly, resulting in impaired passage of food into the stomach.

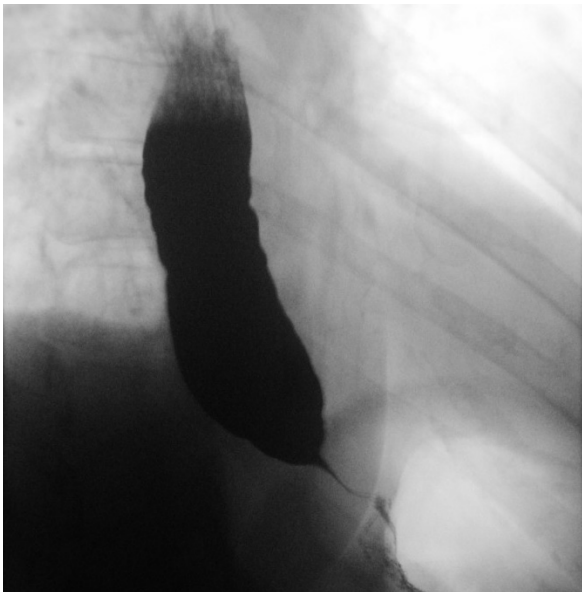

FIGURE 1. Preoperative barium swallow showing dilation of the esophagus with distal tapering of barium, which resembles the classic "bird's beak" radiologic appearance of achalasia.

She underwent a novel minimally invasive procedure called per oral endoscopic myotomy (POEM) at Mass General. During POEM, the surgeon uses a specially designed endoscopic tool and makes a small incision in the inner lining (mucosa) of the esophagus. This allows the surgeon to create a submucosal tunnel to reach the sphincter muscle that is impaired by the disease. The muscle fibers that block the passage of food are divided (Figure 2). Once the blockage caused by the malfunctioning sphincter muscle has been relieved, the endoscope is removed and the incision is closed with clips.

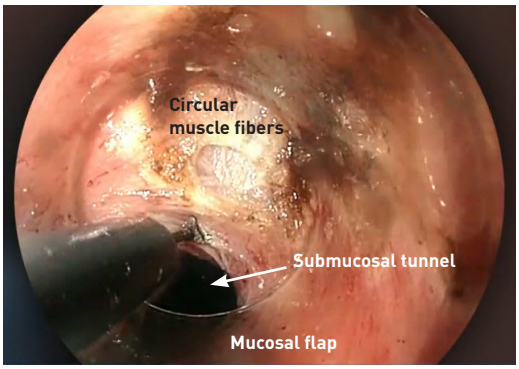

FIGURE 2. Surgical anatomy of POEM.

FIGURE 3. Postoperative barium swallow showing effective myotomy and decreased esophageal diameter. The barium passes freely through the lower esophageal sphincter into the stomach, confirming the relief of the obstruction.

Immediately after the POEM procedure, the patient was able to swallow normally, and the postoperative swallow study showed barium passing freely through the lower esophageal sphincter into the stomach, confirming the relief of the obstruction (Figure 3). She was discharged home the next day.

DISCUSSION OF MANAGEMENT

This patient had esophageal achalasia, which is a primary motility disorder characterized by impaired lower esophageal sphincter (LES) relaxation and absence of esophageal peristalsis leading to impaired transit of the food bolus from the esophagus into the stomach[1]. The symptoms may include dysphagia, regurgitation, respiratory complications (nocturnal aspiration), chest pain and weight loss. Achalasia can also be mistaken for gastroesophageal reflux disease (GERD). Achalasia occurs equally in men and women with incidence of 1 in 100,000 individuals and prevalence of 10 in 100,000. The majority of cases occur in individuals between 30 and 60 years of age[2,3].

Diagnosis of Achalasia

The first step in establishing the diagnosis of achalasia is to exclude a benign or malignant obstruction using radiology and endoscopy. In early stages of the disease, those tests may be normal. However, in more advanced cases, the esophagus is usually dilated with retained food and saliva. Barium studies show a classic “bird’s beak” appearance due to the non-relaxing LES[4] (Figure 1).

The gold standard diagnostic tool is esophageal manometry, which demonstrates decreased or absence of peristalsis, and an increase in the resting tone of the LES with incomplete relaxation[5,6]. High-resolution manometry provides further diagnostic sub-classifications:

- Type I (classic achalasia)
- Type II (achalasia with compression or compartmentalization in the distal esophagus)
- Type III (spastic achalasia)

Treatment of Achalasia

Because the pathogenesis of achalasia is unknown, a curative treatment is not currently available[7]. The palliative treatment options are geared toward reducing the pressure at the LES, thus allowing easier passage of food and liquids into the stomach, relieving the primary symptoms of dysphagia and regurgitation, improving esophageal emptying and preventing the progressive dilatation of the esophagus in the majority of cases.

Treatment modalities include:

- Pharmacological therapy, which has only a minor role in the treatment of esophageal achalasia as it is the least effective option. However, it is useful to help alleviate symptoms while the patient waits for a definite treatment[8, 9]
- Endoscopic injection of botulinum toxin (Botox®), which provides temporary relief from dysphagia. However, it requires new administration of Botox injections every 6-12 months[10]. Additionally, there is evidence that multiple Botox injections into the LES could make a subsequent surgical myotomy more challenging[11]. Considering these limitations, the use of Botox injections should be restricted to patients with comorbidities who are not candidates for laparoscopic Heller myotomy and POEM or who need temporary relief of swallowing problems before undergoing a more definitive procedure
- Pneumatic dilatation (PD), which uses air pressure balloons to dilate intraluminally and disrupt the muscle fibers of the LES. PD is considered the most effective non-surgical treatment for achalasia[12]. However, it is associated with potential severe complications such as esophageal perforation (1.9%) and often requires multiple treatments to achieve a durable result[13, 14]
- Laparoscopic Heller myotomy (LHM), which is the gold standard treatment for achalasia. The technique has evolved from open surgery to a purely laparoscopic approach[15]. It has a much lower morbidity and faster recovery when compared with open and thoracoscopic surgery. The mortality rate is very low (0.1%), and the success rates are very high, averaging 89% (range 76-100%)[16, 17]
- Per oral endoscopic myotomy (POEM), which is a new, minimally invasive surgical technique to treat achalasia[18]. It was developed in Japan in 2008 and then introduced into the United States in 2010[19]. The procedure is purely endoscopic. Occasionally, a tiny incision (5 mm or less) in the abdomen or chest is needed to release carbon dioxide used during the endoscopic procedure. The postoperative pain is minimal, which may lead to a faster recovery. POEM is as effective as LHM in relieving dysphagia; however, opening the LES valve without the addition of a fundoplication permits a significantly higher rate of postoperative acid reflux than LHM. Therefore, patients undergoing POEM need to take a proton pump inhibitor for at least a year postoperatively[20, 21]

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