



## Type III Achalasia Cardia with Epiphrenic Diverticulum Treated By Subtotal Esophagectomy

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### ABSTRACT

*A 48 Year old male patient who is a known case of achalasia cardia presented with recurrent dysphagia 3 ½ years after successful endoscopic pneumatic dilation. On evaluation he was found to have type III achalasia cardia with sigmoid-shaped mega-esophagus and epiphrenic diverticulum. It was decided that the best treatment option would be subtotal esophagectomy, which the patient underwent. The postoperative period was uneventful and the patient was started on oral fluids on postoperative day 7. He was discharged on postoperative day 10*

**KEYWORDS:** *achalasia cardia, megaesophagus, epiphrenic diverticulum, esophagectomy.*

### INTRODUCTION

Vigorous achalasia (type III achalasia), first reported by Sanderson et al. in 1967, is an esophageal motility disorder characterized by both non-peristaltic simultaneous contractions, as seen in diffuse esophageal spasm (DES), and the incomplete relaxation of the lower esophageal sphincter (LES), as seen in achalasia.<sup>(1)</sup>

Esophageal diverticulum is an uncommon entity. They are classified according to their location into proximal, middle and distal types. Epiphrenic diverticula is a disease of the lower end of esophagus situated 4 to 10 cm above the cardia and is about 10% of all esophageal diverticula. It is a disorder due to primary motility dysfunction of the esophagus.<sup>(2)</sup> Achalasia, diffuse esophageal spasm, hypertensive lower esophageal sphincter, and nonspecific motor abnormalities have all been

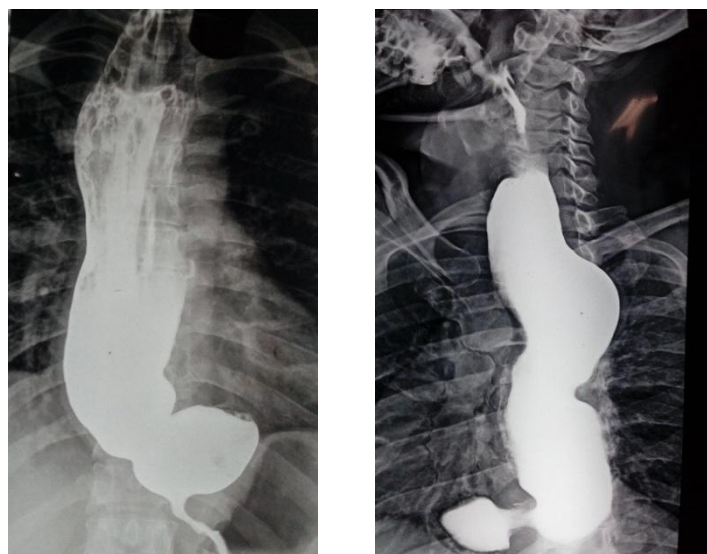
seen in as many as two-thirds of patients with epiphrenic diverticula.<sup>(3)</sup>

### CASE REPORT

This is a 48 year old male patient who is a diagnosed case of achalasia cardia in 2012 and had undergone endoscopic pneumatic dilation at that time. He was asymptomatic for the past 3 ½ years. Now he presented with dysphagia for the past 6 months. It was gradual in onset and progressive. The dysphagia is more for solids compared to liquids. Moreover for ease of passage of solid foods he needs to drink water. He gives history of occasional retrosternal chest pain. History of regurgitation of food is present. Regurgitation occurs predominantly in supine position and it disturbs his sleep. There is history of unquantified weight loss. There is no history of weakness, fasciculation, autonomic symptoms

On physical examination, he is moderately built and well nourished with a pulse rate of 80/minute and blood pressure of 120/70 mm of Hg. There is no pallor, icterus, cyanosis, clubbing, lymphadenopathy or pedal edema. Examination of chest and abdomen are within normal limits. Other systems are within normal limits.

He was evaluated with barium swallow, manometry and esophago gastro duodenoscopy. Barium swallow (5/7/2016) revealed dilated esophagus with lower esophageal diverticulum



**Figure 1:** Barium swallow, **Figure 2:** Barium swallow, 2016

Manometry (9/7/16) was done. It showed elevated basal LES pressure, elevated IRP, absent esophageal peristalsis and presence of premature contractions. There was pan-esophageal pressurization on all swallows. These findings were consistent with achalasia cardia type III – spastic achalasia cardia

Esophago gastro duodenoscopy (19/7/16) showed dilated esophagus with decreased peristalsis. There was a diverticulum at lower end close to the GE junction. GE junction was eccentrically placed and tight

In view of type III achalasia cardia with lower esophageal diverticulum, it was decided to treat the patient surgically

He underwent subtotal esophagectomy on 25/10/2016. Thoracoscopic mobilization of

esophagus was performed. This was followed by laparotomy and creation of gastric conduit. Through incision in left neck, cervical esophagus was divided. The specimen was delivered out through abdomen. Gastric conduit was tunneled through posterior mediastinum followed by anastomosis in the neck



**Figure 3:** Esophagectomy specimen showing diverticulum



**Figure 4 :** Specimen cut open to show the mouth of the diverticulum

## DISCUSSION

Achalasia is a primary motility disorder of the esophagus characterized by lack of peristalsis and failure of the lower esophageal sphincter (LES) to relax appropriately in response to swallowing.<sup>(4)</sup> It is a chronic benign disease that is a common cause of dysphagia, yet its cause remains poorly understood. To this day, the cornerstone of treatment of achalasia remains relief of the

functional obstruction at the level of the gastro-esophageal junction.

The defining pathologic feature of achalasia is progressive inflammation and selective loss of inhibitory myenteric neurons in Auerbach plexus of esophagus. This results in failure of relaxation of the LES and aperistalsis of the esophageal body with subsequent functional obstruction at the level of the gastroesophageal junction and gradual dilation of the esophagus. There is progressive esophageal dilation above the LES ranging from minimal dilation in early achalasia to an esophageal diameter of 10 – 14 cm in patients with long-standing disease.<sup>(5)</sup>

The cardinal symptom of achalasia is dysphagia. As the esophagus dilates, it becomes a reservoir of undigested food. Because of stasis in the esophagus, regurgitation occurs in 76% of patients and 52% complain of heartburn<sup>(6)</sup>

Once the diagnosis of achalasia is suspected, a contrast esophagram should be obtained. This test often shows distal esophageal narrowing (bird beak), an air-fluid level, slow emptying of barium, and esophageal dilation. A sigmoid esophagus or hiatal hernia may also be present.<sup>(6)</sup>

The criterion standard for diagnosis of achalasia is esophageal manometry.<sup>(7)</sup> The defining manometric abnormality is lack of peristalsis of the esophageal body, as manifested by simultaneous, non-propulsive contractions. Impaired LES relaxation with swallowing is seen in 87% of patients. Vigorous achalasia is defined as the presence of non-propulsive contractions with an amplitude greater than 37 mm Hg.<sup>(8)</sup>

A classification scheme has recently been developed based on high-resolution manometry of 73 patients with achalasia. These patients demonstrated impaired relaxation at the esophago-gastric junction but were found to vary significantly with regard to esophageal pressurization. Type 1 (classic) patients had minimal to no esophageal pressurization, type II had pan-esophageal pressurization, and type III (vigorous) had distal spasm<sup>(9)</sup>

Sigmoid-shaped megaesophagus represents the end stage of the disease, when the failure of LES relaxation and aperistalsis of the esophageal body leads to progressive dilation and elongation of the gullet, which may acquire a sigmoid-shaped form.<sup>(10)</sup>

The optimal treatment option is an ongoing matter of debate. Unless new conclusive data prove otherwise, laparoscopic heller myotomy is considered the most durable treatment for achalasia. However, pneumatic dilation is the first choice for non-operative treatment and is more cost effective<sup>(11)</sup>

Endoscopic pneumatic dilation aims to disrupt the LES by forcefully dilating it with an air-filled balloon.<sup>(12)</sup> Eckard et al found 60% Of patients symptom free after 1 year, but more than half of their patients had recurrent symptoms after 5 years.<sup>(13)</sup> Parkman et al showed that 40% of patients treated with a single dilation require a second dilaton after 5 years.<sup>(14)</sup> The patient being reported here had undergone pneumatic dilation as the initial treatment. But he developed recurrent dysphagia 3 ½ years after the procedure

Epiphrenic diverticula represent Pulsion diverticula secondary to increased intra esophageal pressure. They generally project from the right posterior wall of the esophagus. Most epiphrenic diverticula are found in middle-aged or elderly patients, and male patients have a slight preponderance.

With the advent of manometric studies, it has become evident that functional obstruction of the distal end of the esophagus may be not only the cause of the diverticulum but also a major cause of symptoms. Achalasia, diffuse esophageal spasm, hypertensive lower esophageal sphincter, and nonspecific motor abnormalities have all been seen in as many as two-thirds of patients with epiphrenic diverticula.<sup>(3)</sup> It is inferred that increased motor activity and abnormal lower esophageal sphincter relaxation produce zones of increased intraluminal pressure through which outpouchings occur.<sup>(15)</sup>

The symptoms most commonly reported in patients with epiphrenic diverticula are dysphagia and regurgitation. Dysphagia is sometimes associated with esophageal obstruction. Regurgitation of undigested food is frequently of large volume, frequently occurs at night, and is often precipitated by a change in position.<sup>(16,17)</sup>

The diagnosis of epiphrenic diverticulum is established by a barium swallow study. The diverticulum appears as a round structure with a diameter of 1 to 5cm. Giant diverticula are rarely seen but can be larger than 10 cm. The size and position of the diverticulum neck can be precisely assessed with esophagoscopy. Standard esophageal manometry is crucial to assess the presence of an underlying motility disorder.

Patients with minimal symptoms should be managed conservatively. If symptoms are incapacitating or recurrent respiratory complications from aspiration are reported or suspected, surgical therapy is mandatory.<sup>(18)</sup>

The standard and extended heller myotomies are the major procedures for treatment of achalasia. Currently, an esophagectomy is only used in extremely rare cases of end-stage disease in any motility disorder including achalasia that has failed all other therapies and in which the patient is experiencing severe symptoms of sequelae.<sup>(19)</sup>

Our patient presented with recurrent dysphagia 3 ½ years after pneumatic dilation and on evaluation was found to have type III achalasia cardia with sigmoid- shaped megaesophagus. He was also found to have developed an epiphrenic diverticulum. He was treated successfully with subtotal esophagectomy.

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