



# Achalasia as Unusual Indication of Esophagectomy: A Case Report

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## **Authors' contributions**

*This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.*

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**Case Report**

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## **ABSTRACT**

Achalasia is a rare disease. Treatment options in delayed stages like sigmoid esophagus are heller myotomy and esophagectomy. This case report aims to show the anatomy variations, strength, and outcomes of esophagectomy with and hybrid technique.

**Presentation of Case:** A 69-year-old man presented with a long history of dysphagia, regurgitation, and smoking history. An esophagectomy was done. A common complication presented (fistula) and other not so common; pyloric obstruction. Then the patient improved with conservative management.

**Discussion:** the purpose of this review is to compare the different treatment options for end-stage achalasia, one of them is esophagectomy which involves a certain degree of difficulty compared to other indications for esophagectomy. Esophagectomy in patients with achalasia seems a safe procedure. Others treatment options used for end-stage achalasia or for failure from other treatment options for achalasia like Heller's myotomy, pneumatic dilatations or POEM.

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**Conclusion:** Treatment options for end-stage achalasia range from conventional treatment (endoscopic, heller myotomy) to esophageal resection, with very specific indications. Morphological alterations in end-stage achalasia are a surgical challenge. Knowledge of postoperative complications and their treatment has important implications for the right management.

**Keywords:** Achalasia; esophagectomy; minimally invasive approach; three-hole esophagectomy; left aberrant hepatic artery.

## 1. INTRODUCTION

Achalasia is a rare esophageal motor disorder with a prevalence of 8 cases per 100,000 inhabitants [1]. In which there is loss of peristalsis predominantly in the distal esophagus as well as loss of relaxation of the lower esophageal sphincter (LES). The etiology is unknown, however, the most accepted theory proposes that there is a decrease in inhibitory ganglion cells of the myenteric plexus of the esophagus. Consequently producing an imbalance in neurotransmitters and motor alterations such as decreased peristalsis, increased esophageal body pressure, and hypertonia of the LES due to the absence of its relaxation [2].

There are multiple treatments, from pharmacological treatments based on nitrates, botulinum toxin and calcium antagonists which cause smooth muscle relaxation and decrease LES pressure, nevertheless, the main disadvantage of these treatments is that they are temporary [3].

Esophageal dilatation is another treatment modality with an efficacy of 65-85% in a short period time. On the other hand, this treatment has a long-term efficacy of 40 %. Additionally, gastroesophageal reflux disease presents as a complication [4].

The standard surgical treatment is cardiomyotomy, which consists of an anterior myotomy of the fibers of the LES, and the association with an anti-reflux procedure (180° fundoplication either anterior or posterior) with an effectiveness of up to 80% and 65% at 20 years [5, 6]. Several studies have shown superior efficacy in endoscopic treatment with botulinum toxin [7].

Finally, esophagectomy is considered only in the presence of a tortuous esophagus, sigmoid esophagus, megaesophagus, or failure in a patient treated surgically with a myotomy procedure [8]. While there are a few reports of

esophagectomy secondary to achalasia we decided to present this report as an uncommon indication for esophagectomy as well as the strength, outcomes, and limitations of the surgical technique.

## 2. CASE REPORT

A 69 year-old man presented with a long history of dysphagia, regurgitation, and smoking history. No loss of weight or nodal mass in CT. Manometric assessment reports a pattern of I of the Chicago classification, esophageal endoscopy reveals intrinsic narrowing and hiatal hernia type III . It is important to mention that the patient never had previous medical care nor received management of early stage achalasia.

Contrasted radiography of the upper intestinal tract shows a distal filling defect and a sigmoid esophagus (Fig. 1). Previously the intervention laboratory test revealed leukocytes 50000 /mcl, hemoglobin 17 g/dl, hematocrit 51 %, platelets 340000/mcl, glucose 119 mg/dl, urea 3 mg/dl, creatinine 80 mg/dl.

**Operative technique:** Under endotracheal intubation, and general anesthesia, the patient was placed in the left lateral position. An incision of 5 mm was made in the right thorax at the level of the 5° intercostal space, at the anterior axillary line an optic trocar was placed, neumotórax started up to 8 mmHg, two other trocars were set for the assistant, dissection of the esophagus was started in the parietal pleural by advanced monopolar energy, the dissection was followed dorsally (Fig. 2). The azygos vein was clipped, then it was cut.

The abdominal dissection was started by dissecting the greater curvature of the stomach and the lesser curvature, preserving the right gastroepiploic artery. We found a left aberrant hepatic artery, so we stopped the dissection at this point, the gastric tube was created with 3 linear staplers of 60 mm. 4.8 mm. We decided to use 200 units of botulinum toxin in the pylorus as

a drainage procedure. Lateral incision to the sternocleidomastoid muscle, dissection until the esophagus is located then esophagus is extracted (Fig. 3).

We decided to convert to a laparotomy because both sides of the future anastomosis were tense. Kocher maneuver was made to reach a larger gastric tube. An enteral access to nutrition was made with a tube jejunostomy created for enteral nutrition.

Hand sewn anastomosis was performed with PDS 3-0 in one fashion running suture and Connel-mayo suture (Fig. 4). Pen rose drainage was placed in the cervical surgical bed, and two closed drainages in the thorax and abdomen were placed. Abdominal wall closure was performed conventionally. An endopleural tube was placed in the right pleural cavity, continuous suction was started. A chest x-ray of the thorax 24 hours later showed a gastric tube placed in the mediastinum without complications. An apparent complex collection was observed in the right thorax, a thorax CT-Scan was obtained, and a dilated gastric tube was observed secondary to a closed pyloric and delayed gastric emptying, decompression was started with a nasogastric tube.

On the 13th postoperative day esophageal fistula was diagnosed the patient continued decompression of the gastric tube. The patient was discharged on the 15th postoperative day and was followed weekly until the cervical drainage was removed. Accidentally, the jejunostomy tube was removed one month later, because the above, contrast study was done, and it revealed the correct passing of contrast beyond the pylorus.

### 3. DISCUSSION

As mentioned above, achalasia is a rare motor disorder that had a palliative treatment in all the options of management. The indication of esophagectomy in a long-term disease with anatomical changes is offered in approximately 5 % of the patients [9].

The long-term changes in patients with achalasia are the sigmoid esophagus, a tortuous supradiaphragmatic segment, clinically with delayed emptying. Our patient presented with a similar presentation.

In 2012 a classification was proposed regarding the morphology of the esophagus, this divides it

into 3 types: I (<3 cm ), II (3.5< 6 cm ), and III (6 cm ). It takes into account the radiographic findings, measuring the maximum transverse diameter and the angulation it creates in the esophageal axis. [10]. Moreover, the sigmoid esophagus is defined as having a diameter of 10 cm [11].

In Latin America the indication for esophagectomy tends to raise, which is considered a safe procedure in benign causes of esophagectomy, one of the largest reported series was published by Pinotti et. al [12]. The etiology of the sigmoid esophagus in this series of patients was Chagas disease, with similar clinical characteristics as our patient but the pathology differs.

There is literature supporting heller myotomy in patients with megaesophagus [13]. However, in patients with a diameter greater than 6-7 cm in the upper third of the esophagus, beneath 55 years of age, severe mucosal inflammation and moderate to severe dysphagia. The outcomes showed that the reflux is attributed to the Heller myotomy, conversely, the dysphagia improves with surgery, except for 27% of patients that were submitted to myotomy of heller with a preoperative diagnosis of sigmoid esophagus, which suggests the resection of the esophagus from the beginning [14].

Performing peroral endoscopic myotomy is more complicated due to morphological changes, fibrosis and limited space in the submucosal tunnel, perforation of the mucosa can easily occur in the dissection process, making it such a challenging procedure. Reporting long-term symptom relief in over 96 % cases and without serious complications. However, these results are from a tertiary level hospital with experienced endoscopist [15].

In the context of patients with megaesophagus or sigmoid esophagus as in our patient, resection and reconstruction are recommended [16,17]. During the resection of the esophagus, it was difficult to dissect it from the pulmonary ligament due to its esophagic diameter, exposing the irrigation coming from the aorta, achieve adequate vascular control and extraction of the surgical specimen through the cervical incision. There are recommendations which suggest an incision in posterior axillary line in the 7<sup>th</sup> intercostal space with a wound protector but it is used to remove the surgical specimen in the Ivor Lewis a esophagectomy [18].

The use of Esophagectomy is the best treatment to release esophageal obstruction, dismissing pulmonary complications in patients with achalasia and megaesophagus, This treatment is by far superior to myotomy or cardioplasty.

Regarding the technical difficulties that can be encountered in a gastric pull-up, the most common anatomical variant is the aberrant left hepatic artery, a branch of the left gastric artery. The patient presented an aberrant left hepatic artery, so it was decided to left in place, not knowing the implications that could occur, such as distal ischemia of the gastric tube, and hepatic ischemia, the last mentioned complication has already been described previously [19,20]. The incidence of hepatic ischemia is as high as 6.3%. The presence of an aberrant left hepatic artery is reported in 4.3-22.4% [19].



**Fig. 1. Contrasted radiographic study showing dilated esophagus in all its portions**



**Fig. 2. Esophageal dissection**



**Fig. 3. Surgical specimen, resected esophagus from its cervical portion to the esophagogastric junction**



**Fig. 4. Gastric-esophageal hand sewn anastomosis in 1 layer with PDS-3-**

Outcomes in esophagectomies for other causes versus achalasia have the same overall results; mortality, readmission, reoperation, and overall complications are not statistically significantly associated in patients undergoing esophagectomy for achalasia and non-achalasia patients [21]. The patient remained hospitalized for 15 days.

Anastomotic leakage is one of the most feared complications. [22], is present in 6.6-17 % of anastomoses at the cervical level. An anastomotic leak is defined as a "full-thickness gastrointestinal defect involving the esophagus, anastomosis, staple line, or conduit, regardless of presentation or method of identification" [23].

The incidence of anastomotic leakage is reported to be 2% in the minimally invasive Ivor Lewis approach and 6.6% in the Mckeown approach. [24] Another large series of patients (n=501) reported an incidence of 11.8 % overall, leakage was predominant in the minimally invasive approach with a rate of 20 % [25].

In the particular case of our patient, conservative management was decided since it was fitted in the classification of Lerut et al. We detected it by radiological studies [23]. The clinical evolution did not show any inflammatory response and the upper intestinal tract contrasts did not show collections in cervical anatomic planes. Some risk factors have been identified concerning the patient's medical history: malnutrition (albumin <3.0 g/dL), obesity (body mass index [BMI] > 30 kg/m<sup>2</sup>) or underweight patients (BMI < 18.5 kg/m<sup>2</sup>), heart failure, hypertension, diabetes, renal failure, steroids, and tobacco use [26].

Regarding the surgical technique and anastomotic leaks, it is debated whether there are factors that influence the leakage of the anastomosis, it is more common as mentioned above in cervical leaks, requiring a longer segment of the stomach and the site where the anastomosis is performed, which is at the level of the gastric fundus, a site that presents some restriction of blood supply [26]. Despite preserving the left gastric artery in this case, having a double irrigation tube, a leak occurred. Currently, there are no studies comparing the results and the rate of leakage in patients operated on with minimally invasive esophagectomy techniques in comparison with the open or hybrid approach as well as in favor of or against hand-sewn or mechanical anastomosis in the patient with achalasia, the

paper only reports the incidence 1 patient in series of 26 patients [15]. The maximum evidence we have is in oncologic patients in whom it has been demonstrated that there is no difference between the technical factors: hand-sewn anastomosis versus EGA, minimally invasive approach versus open esophagectomy, or the route of the ascent of the gastric tube [26, 27].

Although there is evidence comparing non-draining versus intrapyloric Botulinum Toxin injection versus pyloroplasty, pyloroplasty is not associated with short-term complications, only with long-term morbidity. Intervention at the level of the pylorus may be necessary. Despite the use of intrapyloric botulinum toxin injection, the patient developed delayed gastric emptying [28].

#### **4. CONCLUSION**

Treatment options for end-stage achalasia range from conventional treatment (endoscopic, heller myotomy) to esophageal resection, with very specific indications. It is important to know the anatomical variations that can be found in end-stage achalasia esophagectomy with the aim to reduce complications. Knowledge of postoperative complications and their treatment has implications for a right management. Although there is not much evidence of the results of esophagectomies for advanced-stage achalasia, the current evidence shows esophagectomy a safe procedure.

#### **CONSENT**

As per international standard or university standard, Participants' written consent has been collected and preserved by the author(s).

#### **ETHICAL APPROVALS**

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

#### **COMPETING INTERESTS**

Authors have declared that no competing interests exist.

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