

Thoracoscopic esophagectomy with gastric pull-up in megaesophagus secondary to terminal achalasia

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Abstract

In terminal achalasia, the presence of a megaesophagus with loss of axis in its distal third or persistence of symptoms despite multiple previous treatments, the esophagectomy by thoracoscopy with reconstruction by laparoscopic gastric ascent and cervical anastomosis (McKeown or 3-field esophagectomy) is a feasible and safe option that has managed to reduce morbimortality secondary to the intervention. We present the case of a 69-year-old patient with megaesophagus secondary to terminal achalasia and describe the surgical management and post-operative evolution.

Keywords: Terminal achalasia. Megaesophagus. Thoracoscopic esophagectomy. Gastric pull-up. McKeown.

Introduction

Achalasia is a chronic, progressive, idiopathic esophageal motor disorder characterized by the loss of primary peristalsis and the inability of the lower esophageal sphincter to relax in response to swallowing. A decrease in the number of ganglion cells in the myenteric plexuses has been observed in several surgical specimens, which preferentially affects the inhibitory neurons that produce nitric oxide that influences the relaxation of esophageal smooth muscle^{1,2}.

It has an estimated annual incidence of 1.6 cases/100,000/year. The most common symptoms are dysphagia, regurgitation, retrosternal pain, weight loss, and respiratory symptoms that can lead to aspiration pneumonia. An incidence of achalasia squamous cell carcinoma is estimated at 312.4 cases/100,000 patient-years².

Treatment aims to reduce the pressure gradient through the lower esophageal sphincter and improve gravitational esophageal emptying to alleviate symptoms. Pneumatic dilation and surgical myotomy are the first-line treatment modalities. In the absence of treatment or inadequate management, approximately 5% of patients develop terminal disease, which is characterized by progressive dilation with increased tortuosity of the esophagus^{3,4}. Esophagectomy is indicated in patients with persistent or recurrent achalasia after the failure of less invasive treatments and who have developed a tortuous and dilated esophagus with deviation of its axis^{5,6}.

Case presentation

A 69-year-old woman was taken to the emergency room of our institution for presenting painless and

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Figure 1. **A:** coronal section of computed tomography showing tortuous dilated esophagus with loss of axis with respect to the diaphragmatic hiatus and narrowing at the level of the esophagogastric junction. **B:** axial section showing markedly dilated megaesophagus occupying a large part of the left hemithorax. **C:** esophagogastric series with water-soluble contrast medium showing dilated esophagus of sigmoid aspect with filiform passage of the contrast medium into the stomach. Note the loss of the vertical axis.

progressive dysphagia to solid and liquid foods that began 11 years ago, which leaves her free evolution until total intolerance to the oral route. Dyspnea and unintentional weight loss of approximately 16 kg in 2 years were added, along with the appearance of a mass in the left cervical region, which is why she was evaluated by the otorhinolaryngology service, who requested a chest X-ray in which there was evidence of a mediastinal tumor with a hydro-aerial level. A thoraco-abdominal tomography is indicated in which a dilated esophagus with a diameter of up to 14.3 mm is observed at the retrocardiac level from the pharyngeal portion to the esophageal-gastric junction where its luminal diameter is abruptly reduced, the interior occupied with gastro-food residue throughout the trajectory causing anterior displacement and extrinsic compression of the trachea and anterior displacement of the cardiac silhouette. Corresponding to megaesophagus (Fig. 1A and B).

Physical examination showed oxygen saturation of 84% in room air, tachycardia, cachectic complexion with a body mass index of 18.6 kg/m² and at the left cervical level, a tumor of approximately 7 × 5 cm, soft, not painful. A 20 Fr nasogastric tube was placed for aspiration and emptying of food remains in the esophagus and to perform panendoscopy, however, after 3 attempts, it was not possible to complete the study due to the presence of pronounced angulation of the esophagus in its middle third that made it difficult for the endoscope to pass, observing significant esophageal dilation and esophagitis due to stasis.



Figure 2. Semiprone decubitus position. Delimitation of the posterior axillary line and placement of the trocars in the 4th, 6th, and 8th intercostal spaces.

An esophagogastric series with water-soluble contrast was performed, where elongation of the esophagus was observed with a tortuous path (with a “sigmoid” appearance) and gradual luminal decrease at the level of its distal third, acquiring a bird’s beak appearance that barely allowed the filiform passage of the contrast medium to the stomach, suggesting achalasia (Fig. 1C). Without the possibility of performing esophageal manometry, it was decided to perform a serological test to rule out Chagas disease, which was negative.

At the biochemical level, protein-calorie malnutrition and hydroelectrolyte imbalance were evident, so a central venous catheter was placed to initiate total parenteral nutrition for 2 weeks before the surgical procedure.

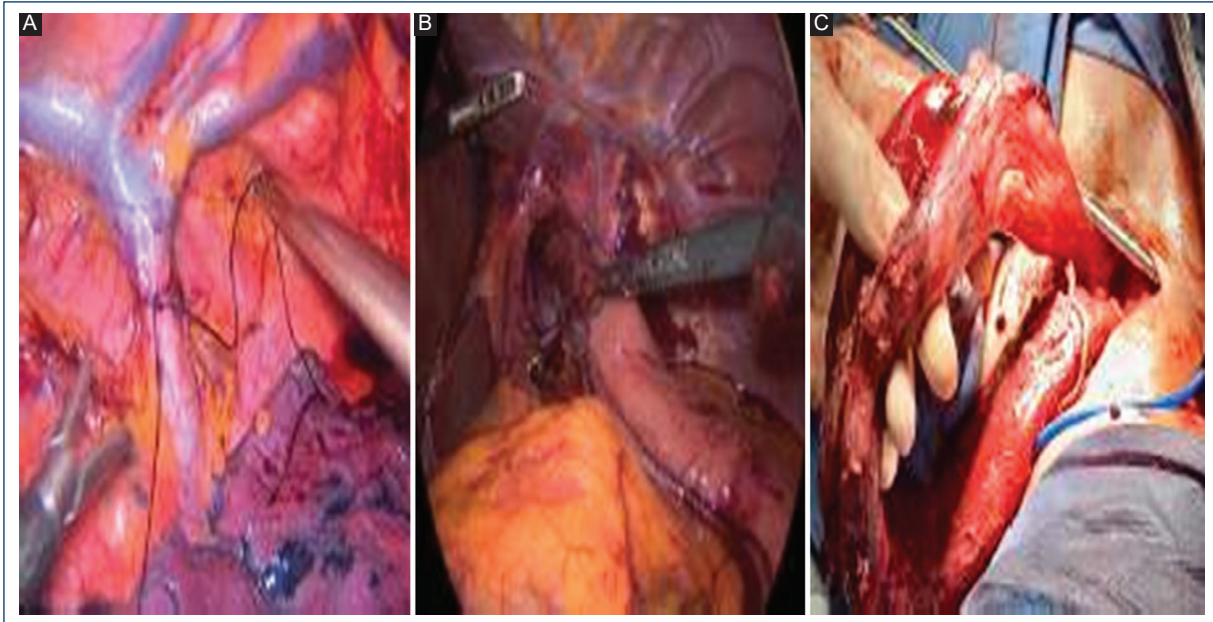


Figure 3. **A:** ligation of the azygos vein during thoracoscopy to allow complete dissection of the esophagus. **B:** formed gastric tube approximately 4 cm wide being directed toward the diaphragmatic hiatus. During the ascent, it must be ensured that the stapling line is kept on the right side avoiding its torsion. **C:** dilated esophagus exteriorized by left cervical incision.

In view of the imaging findings and having evaluated and ruled out other treatment alternatives, it was decided to proceed with esophagectomy and gastric ascent using a minimally invasive approach.

Initially, thoracoscopic esophagectomy was performed in the semi-prone position (Fig. 2) observing distortion of the anatomy at the thoracic level with an esophagus approximately 10 cm dilated with increased vascularity and periesophageal inflammation, as well as multiple adhesions to the right lung and mediastinal pleura. An adequate dissection was performed from the thoracic operculum to the diaphragmatic hiatus (Fig. 3A). During the second stage and after placing the patient in dorsal decubitus in a French position, laparoscopy was performed to begin with the dissection of the gastrocolic ligament, preserving the right gastroepiploic arch that will supply the gastric tube. By means of an articulated linear stapler, a gastric tube of approximately 4 cm wide with the greatest curvature is formed (Fig. 3B). At the same time, a left cervical incision of approximately 5 cm was made using a third approach to access and dissect the cervical esophagus (Fig. 3C). Once the gastric tube has been made and the abdominal hiatal dissection has been completed, the cervical esophagus is tractioned, externalizing the thoracic megaesophagus through the incision in the neck until the neoesophagus

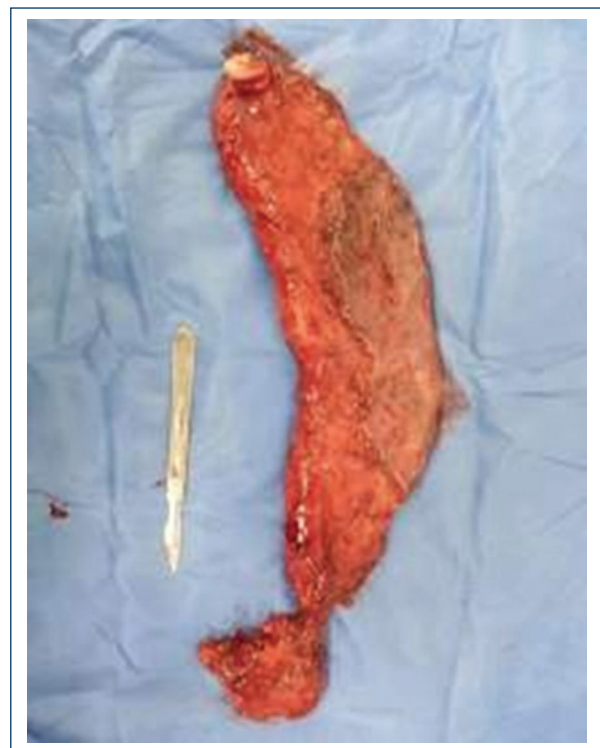


Figure 4. Surgical specimen.

and its gastric remnant, which had not yet been sectioned, are visible (Fig. 3C). Mechanical antiperistaltic lateral cervical esophagogastric anastomosis was

performed. Finally, a Penrose drain is placed leading to the mediastinum posterior to the cervical anastomosis. The resected specimen showed an esophagus dilated throughout its entire length (Fig. 4). The procedure lasted a total of approximately 7 h and 50 min with a bleed of 100 cc. The histopathological study was compatible with achalasia, revealing mild lymph node degeneration + acute ulcerated esophagitis with accentuated epithelial regenerative changes and histological data suggestive of acute esophagitis.

The patient remained in intensive care for 72 h, completing the post-operative period without complications. Total parenteral nutrition was continued until oral feeding was ensured, which was started with clear liquids on the 3rd day, with no evidence of leakage, a polymeric diet was started on the fourth day and progressed to porridge on the 11th post-operative day, having adequate tolerance. She was finally discharged on the 13th post-operative day. After 8 months of follow-up, the patient had adequate oral intake and a weight gain of 5 kg.

Discussion

Patients with achalasia have a 7-140 times higher risk than the general population of developing esophageal carcinoma⁷. It takes approximately 10-20 years from the onset of the first symptoms for a neoplastic transformation². Esophagectomy in terminal achalasia is indicated in patients with a tortuous and dilated esophagus with axis deviation. Extrapolation from surgical series suggests that this process can last between 12 and 15 years^{3,8}. Another indication is the persistence of long-term disabling symptoms despite multiple endoscopic and/or laparoscopic interventions. In a series of 385 patients with achalasia who underwent esophagectomy after having previously undergone unsuccessful Heller myotomy, the symptoms reported were regurgitation (100%), dysphagia (83%), chest pain (67%), and recurrent episodes of pneumonia and/or chronic cough (50%)⁹. In addition, the success of a myotomy is substantially reduced when the esophagus is tortuous in its supradiaphragmatic course, as is the case in the case presented^{5,9}.

The transhiatal approach can prevent the development of serious pulmonary complications and accelerate post-operative recovery; however, insufficient visualization of the posterior mediastinum, limited space for manipulation due to a large esophageal dilation, long operating time and great blood loss due to the difficulty in performing adequate hemostasis, are disadvantages that are considered to impact morbidity and mortality. The choice in favor of triple approach esophagectomy,

also known as McKeown, is associated with fewer lethal complications in the event of anastomotic leakage because cervical exposure allows an anastomosis to be performed with less technical difficulty¹⁰⁻¹². A dietary adjustment before surgery with a clear liquid diet for 2-5 days is recommended to reduce the amount of esophageal content retained¹³. Likewise, the anesthesiologist should perform the rapid sequence intubation technique with cricoid pressure or intubation in a semi-upright or fully upright position to avoid bronchoaspiration of the contents that could remain in the esophagus¹⁴.

In a systematic review of patients undergoing esophagectomy for terminal achalasia, bringing together a total of 1307 patients, a post-operative morbidity was found between 19% and 50%, finding that the two most frequent complications were pneumonia and anastomotic leakage, a mortality of 0-5.4% and a hospital stay between 10 and 16 days. It was also observed that 27.5% of patients required endoscopic dilation of the anastomosis within 1 year¹⁵. The risk of respiratory infection after surgery can be reduced by three times if a minimally invasive approach is used. It has been seen that up to 40% of patients experience some type of cardiac arrhythmia after esophagectomy, most frequently atrial fibrillation, which is often asymptomatic, however, this can be a symptom of anastomotic leakage^{14,15}. Chylothorax is a rare complication that can occur in 2% of all esophagectomies but can increase by up to 13% in achalasia, probably due to enlargement of the lymphatic ducts and their displacement due to distortion of the esophageal anatomy. Recurrent laryngeal nerve injury during cervical dissection may be greater in up to 20% of patients with achalasia and megaesophagus⁸. Up to 46% of patients may experience postoperative dysphagia, most of them due to stenosis of the anastomosis, which usually responds to endoscopic pneumatic dilations¹⁵.

Regarding the choice of esophageal substitute, gastric interposition is considered to be of choice because it requires a single anastomosis and has a more constant pattern of blood supply to the stomach compared to the colon. In colonic interposition, three anastomoses are required, and long interpositions run a greater risk of ischemia, which can predispose to the formation of fistulas and stenosis. Duct redundancy can cause mechanical dysfunction and reoperation^{3,15}. It is estimated that the majority of patients (75 and 100%) have managed to follow an unrestricted diet after the procedure. The success of surgery deteriorates progressively over time, with a failure rate of 20% at 10 years and up to 35% at 20 years¹⁶.

In this case, the choice of esophagectomy by transhiatal approach is ruled out as an option due to the

formation of dense adhesions between the dilated esophagus and the mediastinum, as well as the presence of hypertrophied bronchial and aortic branches that would have hindered blind mobilization of the esophagus. Similarly, excessive dilation not only of the distal esophagus but also of the proximal esophagus would make its transhiatal mediastinal dissection extremely difficult with a higher rate of bleeding. For these reasons, the thoracoscopic approach turned out to be a feasible, safe option with a lower risk of complications for the patient.

Conclusion

Thoracoscopic esophagectomy can be performed safely, with acceptable morbidity, low mortality, and satisfactory dietary function. The results obtained by this minimally invasive approach for patients with megaesophagus are good when performed by experienced surgeons.

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Conflicts of interest

The authors declare no conflicts of interest.

Ethical declarations

Protection of humans and animals. The authors declare that no experiments involving humans or animals were conducted for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from patients, and received approval from the Ethics Committee. The SAGER guidelines were followed according to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing of this manuscript.

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