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Case Report

Laparoscopic Heller myotomy in sigmoid megaesophagus: function preserving surgery as an alternative to esophagectomy in end stage achalasia

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ABSTRACT

End-stage achalasia with sigmoid megaesophagus represents a therapeutic challenge. Esophageal resection has traditionally been favored, although it is associated with significant morbidity. Laparoscopic Heller myotomy has emerged as a function-preserving alternative in selected patients; however, its use in cases with severe respiratory symptoms and moderate esophageal angulation has not been widely documented. Herein, we report the case of a 43-year-old male with progressive dysphagia, regurgitation, unintentional weight loss, and severe respiratory symptoms (dyspnea and orthopnea). The preoperative Eckardt score was 10 (severe achalasia). Imaging studies demonstrated sigmoid megaesophagus (grade IV) with an esophageal angulation of 75°. Laparoscopic Heller myotomy with intraoperative endoscopy was performed, and 300 mL of retained food content was aspirated. The patient had a favorable postoperative course, with immediate symptom resolution and an Eckardt score of 1 at two months. The hiatal release with anchoring technique (pull down) was not required due to the manageable angulation. Laparoscopic Heller myotomy is an effective option for the management of selected patients with end-stage achalasia and sigmoid megaesophagus, even in the presence of severe respiratory symptoms and moderate esophageal angulation. Appropriate patient selection and meticulous surgical technique allow esophageal preservation with low morbidity and excellent clinical outcomes.

Keywords: Achalasia, Sigmoid megaesophagus, Heller myotomy, Eckardt score, Esophageal preservation

INTRODUCTION

Achalasia is a low prevalence esophageal motility disorder, with an estimated annual incidence of 1.6 per 100,000 individuals.[1] When left untreated or refractory to initial management, it may progress to an end stage form characterized by a massively dilated, sigmoid shaped esophagus, a condition known as megaesophagus.[2]

This advanced morphological stage, particularly the "sigmoid" or "redundant" esophagus, presents unique technical challenges due to esophageal tortuosity and food retention, which historically led to the perception that the esophagus is "irreparable".[3] The optimal surgical management of end stage achalasia with sigmoid megaesophagus remains controversial. Although esophageal resection has traditionally been the standard approach, it is associated with significant morbidity, including anastomotic leakage, delayed gastric emptying, and a mortality rate of 2–5%.[2,5] Emerging evidence challenges this paradigm, suggesting that laparoscopic Heller myotomy can achieve adequate symptomatic relief even in patients with a sigmoid esophagus, particularly when combined with intraoperative esophageal cleaning.[6,7]

The Eckardt score constitutes the international standard for assessing the severity of achalasia and the response to

treatment, and it is widely used in case series and comparative studies. Likewise, esophageal angulation, especially angles greater or less than 90°, has been reported as a prognostic parameter in preoperative evaluation. The myotomy technique with hiatal release and esophageal anchoring (pull down) has demonstrated better outcomes in patients with severe angulation (>90°).[4]

We present the case of a patient with end stage achalasia and sigmoid megaesophagus successfully managed with conventional laparoscopic Heller myotomy, highlighting the importance of preoperative quantification using validated scales.

CASE REPORT

We present the case of a 43-year-old male with no history of allergies, chronic diseases, or toxic habits. His only relevant surgical antecedent was a vasectomy performed eight years earlier. The clinical picture began five years before consultation, characterized by heartburn, regurgitation, chest pain, and dry cough, with self-medication using antacids that provided partial and transient improvement. During the last year, a significant exacerbation was documented, with the development of progressive dysphagia for both solids and liquids, exertional dyspnea, respiratory difficulty during conversation, and orthopnea with a choking sensation secondary to nocturnal reflux, which forced him to sleep in a seated position.

According to the Eckardt score, the patient had a score of 10 (dysphagia: 3, regurgitation: 3, weight loss: 3, chest pain: 1), corresponding to severe achalasia.

It is important to note that these respiratory symptoms were secondary to mediastinal occupation by the megaesophagus and not to an underlying septic or infectious condition, a finding that has been previously described in the literature.[8]

On physical examination, the patient was hemodynamically stable, afebrile, and showed no signs of systemic inflammatory response. Pulmonary auscultation revealed decreased air entry in the right apical region, correlating with the mediastinal occupation observed on the chest radiograph (Figure 1).

The patient also reported decreased appetite and unintentional weight loss. Admission laboratory tests showed all parameters within normal ranges, including a complete blood count without leukocytosis, normal C reactive protein, and negative procalcitonin, which supported the absence of an infectious process.

High resolution esophageal manometry revealed ten failed swallows, all accompanied by panesophageal pressurization. Catheter coiling at the level of the esophagogastric junction (mirror image sign) was observed, a phenomenon highly suggestive of megaesophagus, which made it impossible to measure the integrated relaxation pressure of the lower esophageal sphincter.

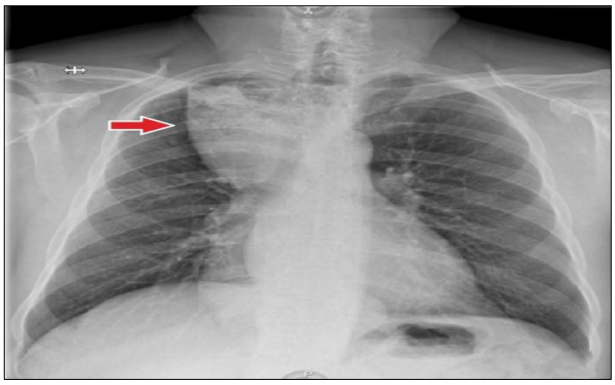

Figure 1: Plain chest radiograph showing esophageal dilation and upward displacement into the right hemithorax (arrow).

Motility of the upper esophageal third (striated muscle) was preserved, and the upper esophageal sphincter showed adequate coordination with the pharynx. The water-soluble contrast esophagogastroduodenal series demonstrated a patulous esophagus with a significant reservoir and a characteristic inverted sigmoid configuration in the right hemithorax (Figure 2), findings consistent with advanced megaesophagus (grade IV), according to the morphological classification described in the literature.[9]

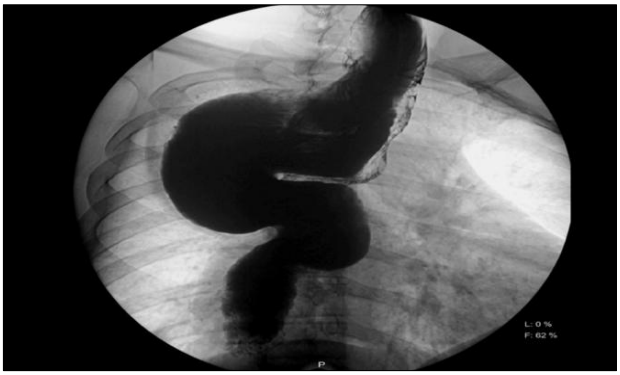

Figure 2: Water soluble contrast esophagogastroduodenal series demonstrating a dilated and redundant esophagus with a sigmoid configuration projected into the right hemithorax.

Esophageal angulation was estimated at 75° based on the esophagogastroduodenal series image, confirming a high-grade sigmoid esophagus. Serology for *Trypanosoma cruzi* was negative, ruling out a Chagasic etiology, which is relevant in our context since *Chagasic achalasia* may have a different clinical behavior.[10]

Given the diagnosis of end stage achalasia with symptomatic megaesophagus, the case was presented at a

multidisciplinary session. In accordance with current evidence supporting esophageal preservation in advanced disease, and after a comprehensive patient evaluation, laparoscopic Heller myotomy with intraoperative endoscopy was determined to be the most appropriate intervention.[3,4,7]

Intraoperative findings confirmed a redundant, dilated esophagus approximately 5 cm in diameter, with a narrowing zone at the level of the esophagogastric junction. During intraoperative endoscopy, abundant retained food material was observed. A total of 300 ml of fluid was aspirated; however, solid debris adherent to the esophageal wall prevented the endoscope from traversing the sigmoid segment. Insufflated air passed successfully into the gastric cavity, demonstrating partial communication at the esophagogastric junction. The myotomy was extended approximately 5 cm proximally along the esophagus and 3 cm distally onto the stomach (Figure 3).

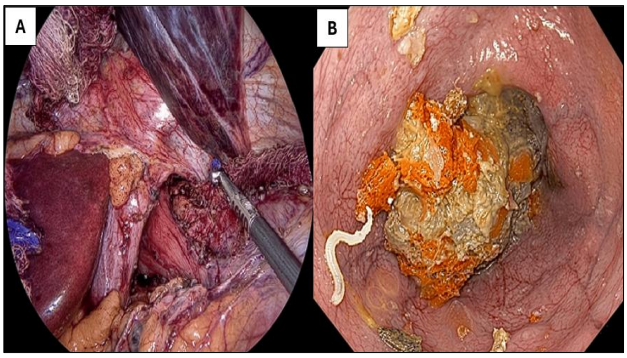

Figure 3: Intraoperative findings (A) laparoscopic view of the Heller myotomy, and (B) intraoperative endoscopy revealing abundant retained food debris within the sigmoid esophagus.

In this case, an extensive myotomy (5 cm proximal, 3 cm distal) was chosen without additional hiatal release or esophageal anchoring (pull down technique), because the angulation was manageable (75°) and esophageal mobilization was sufficient to allow adequate exposure. The patient tolerated the procedure well, with no intraoperative or postoperative complications. Oral intake was initiated on the first postoperative day, with immediate resolution of dysphagia and regurgitation. He was discharged on the second postoperative day. At two month follow up, the patient remained asymptomatic, with significant improvement in respiratory symptoms (dyspnea and orthopnea resolved), partial recovery of body weight, and an Eckardt score of 1 (occasional dysphagia), demonstrating an excellent clinical response.

DISCUSSION

The management of end-stage achalasia with sigmoid megaesophagus remains controversial, and while esophagectomy has been the traditional standard, it is associated with significant morbidity including anastomotic leakage, delayed gastric emptying, and a mortality rate of 2-5%.[2,5] Our case adds to the growing evidence that laparoscopic Heller myotomy can be a valid, safe, and function-preserving alternative in selected patients.[3,4,7] The use of the Eckardt score was instrumental in objectively quantifying the patient's severe preoperative status (score of 10) and his excellent response to surgery (score of 1 at two months), a degree of improvement comparable to that reported in major series.[4] A key finding was the estimation of esophageal angulation at 75°. This moderate angulation (<90°) allowed us to opt for a conventional extensive myotomy (5 cm proximal, 3 cm distal), without the need for the more complex pull-down technique (hiatal release and anchoring), which is reserved for patients with severe angulation (>90°).[4] The decision was justified by the favorable intraoperative outcome. The patient's severe respiratory symptoms (dyspnea, orthopnea), which were initially worrisome, were confirmed to be secondary to mediastinal occupation by the megaesophagus and resolved completely after surgery, a finding previously described but worth emphasizing.[8] The main limitation of our report is the absence of postoperative imaging to objectively document the correction of the esophageal angulation; however, the sustained clinical improvement validates the success of the procedure. In conclusion, for patients with sigmoid megaesophagus and moderate angulation (<90°), laparoscopic Heller myotomy should be considered the first surgical option before resorting to the more morbid esophagectomy.

CONCLUSION

Laparoscopic Heller myotomy constitutes a valid, safe, and less morbid alternative to esophagectomy in selected patients with end stage achalasia and sigmoid megaesophagus. This case demonstrates that even in the presence of significant esophageal redundancy (angulation of 75°) and severe respiratory compromise, a function preserving approach is feasible with favorable outcomes, provided that adequate patient selection and meticulous surgical technique are performed. The Eckardt score proved to be a useful tool for quantifying preoperative severity and postoperative response. The absence of postoperative imaging studies constitutes a limitation that should be addressed in future research. Heller myotomy should be considered the first surgical option before resorting to esophagectomy in patients with sigmoid megaesophagus and moderate angulation.

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