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Esophageal Achalasia is treated with surgical Heller Cardiomyotomy and Dor Fundoplication: A Rare Case Report

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KEYWORDS
Achalasia; Dysphagia; Surgical heller cardiomyotomy; Dor fundoplication

ABSTRACT
Achalasia is a rare esophageal motility disorder characterized by failure of lower esophageal sphincter (LES) relaxation and absent peristalsis. With an annual incidence of 1/100,000, it causes dysphagia, regurgitation, and weight loss. This report discusses a case managed successfully with surgery. A 47-year-old male presented with progressive dysphagia to solids and liquids, regurgitation, chest tightness, and 5 kg weight loss over one year. Unresponsive to proton pump inhibitors, endoscopy revealed distal esophageal narrowing. The case highlights the common misdiagnosis of achalasia as GERD, which can lead to significant delay in appropriate treatment. The gold standard for definitive management, especially in fit patients, is laparoscopic or open Heller cardiomyotomy. This procedure involves cutting the dysfunctional esophageal muscle fibers to relieve the obstruction. It is complemented by a Dor (anterior) fundoplication, a partial wrap that helps prevent postoperative gastroesophageal reflux, a common complication of the myotomy alone. Surgical intervention with Heller cardiomyotomy and Dor fundoplication is an effective and definitive treatment for esophageal achalasia. It successfully relieves obstructive symptoms and prevents iatrogenic reflux, leading to significant clinical improvement, as evidenced by a postoperative Eckardt score of 0 in this patient. A high index of suspicion is necessary for timely diagnosis and to improve patient outcomes.

INTRODUCTION

Esophageal achalasia is a rare esophageal motility disorder in which the lower esophageal sphincter (LES) fails to relax (Wilkinson & Halland, 2020). The global annual incidence of achalasia is approximately 1 in 100,000 individuals, with an overall prevalence of 9 to 10 in 100,000 individuals (Gaber et al., 2022). Patients with achalasia typically experience symptoms of dysphagia for solids and liquids, regurgitation, heartburn, chest pain, and nocturnal cough (Pesce et al., 2023).

Dysphagia is present in more than 90% of patients with achalasia (Pandolfino & Gawron, 2015). Achalasia is caused by degeneration of the vagus nerve fibers and the myenteric nerve plexus located in the LES (Rieder et al., 2020). This degeneration leads to the loss of inhibitory neurons containing nitric oxide synthase and vasoactive intestinal peptide in the esophageal myenteric plexus (Chalazonitis & Rao, 2018). However, the exact cause underlying this degenerative process remains elusive, although various hypotheses include viral infection, genetic susceptibility, and autoimmune responses (Casanova & Abel, 2021).

Diagnostic support is crucial to confirm the diagnosis of achalasia, as the symptoms are nonspecific and difficult to differentiate (Niebisch et al., 2017). Barium esophagography is the best initial test, demonstrating classic findings such as a "bird's beak" appearance and lack of peristalsis(Barlow et al., 2019) . Upper endoscopy is also used to rule out malignancy (Zullo et al., 2019). Esophageal manometry remains the gold standard (Pham et al., 2020). Various treatments are available to relieve symptoms and reduce outflow resistance (Lerman, 2015). Pharmacological options include phosphodiesterase-5 inhibitors, calcium channel blockers, and (Kloner et al., 2018). Surgical therapy includes laparoscopic or laparotomy and surgical cardiomyotomy with fundoplication (Rebecchi et al., 2018).

Here, we present a case report of a 45-year-old man with esophageal achalasia treated with surgical Heller cardiomyotomy and Dor fundoplication (Uppal & Wang, 2016). The purpose of this study is to describe the clinical presentation, diagnostic approach, and surgical management of a patient with esophageal achalasia, highlighting the effectiveness of Heller cardiomyotomy combined with Dor fundoplication (Doubova et al., 2021). This report aims to contribute to the medical literature by providing insights into the management of this rare condition and emphasizing the importance of timely surgical intervention to improve patient outcomes and quality of life (Dumbuya et al., 2025).

METHOD

Case Report

A 47-year-old man came to the hospital with complaints of difficulty swallowing that had been felt since 1 year ago and had worsened since 1 week ago, the patient had difficulty swallowing liquid foods, the stomach felt full and bloated, nauseous, vomiting and felt a bit tight, the patient also complained of losing weight of about 5 kg after experiencing complaints. The patient's medical history had been treated by a general doctor and was said to have GERD, had taken stomach acid-reducing medication but there was no change, the patient had a history of hypertension and regularly took amlodipine 5 mg, From the physical examination, it was found that the consciousness was compos mentis, blood pressure 130/80, Pulse 80x / minute, RR 20x / minute Spo2 99% room air, From the General status examination, the results were within normal limits. Supporting examinations were carried out, chest X-rays were within normal limits, complete blood and clinical chemistry were within normal limits, the patient also underwent an endoscopy, the results showed that the lower 1/3 of the esophageal lumen was narrowed even though esophageal insufflation had been performed. The patient was treated with RL 20 tpm, amlodipine 1x5mg, lansoprazole 2x1 vial. Next, the patient was planned to undergo Heller Cardiomyotomy and Dor fundoplication with General anesthesia, with pre-op ceftriaxone 2 grams.

Figure 1. Endoscopy shows the lower 1/3 of the esophageal lumen is narrowed.

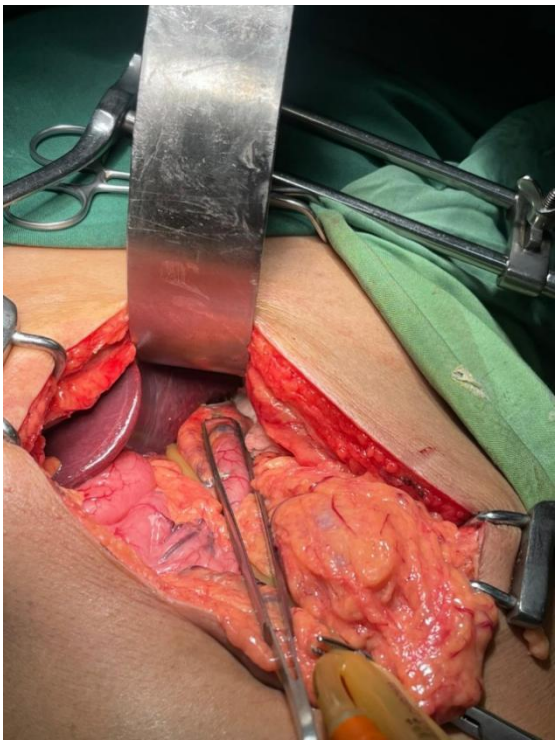

Figure 2. Surgical Heller Cardiomiotomy performed

Source: Authors' documentation

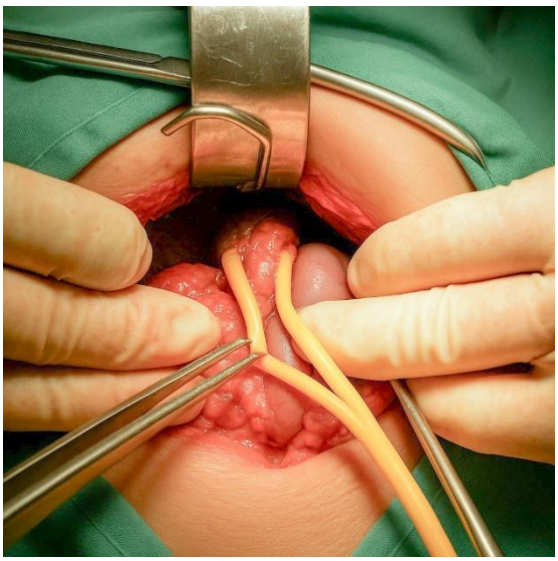

Figure 3. Dor Fundoplication performed

Source: Authors' documentation

Result and Discussion

Achalasia is a rare disorder affecting the esophagus and disrupting the movement of food into the stomach, where the lower esophageal sphincter (LES) cannot relax. Achalasia can

manifest in a variety of ways that negatively impact quality of life. Achalasia can be categorized into four categories based on the diameter and length of the esophagus: a diameter <4 cm is grade 1, a diameter of 4–6 cm is grade 2, a diameter >6 cm is grade 3, and grade 4 is a sigmoid esophagus. The hallmark symptom of achalasia is progressive difficulty swallowing solids and liquids. Approximately 27% to 42% of patients with achalasia complain of heartburn, which is often misdiagnosed as gastroesophageal reflux disease, which is treated with proton pump inhibitors (PPIs).

Misdiagnosis can lead to significant delays in diagnosing achalasia until the patient experiences persistent symptoms, necessitating further diagnostic evaluation. Dysphagia and regurgitation are common in all age groups, but younger patients can also experience chest pain and heartburn, while obese patients (BMI ≥30) complain of coughing and choking. In addition, complaints that can arise include weight loss, such as having food stuck in their neck, accompanied by a burning sensation in the chest and chest pain, In this patient, the patient complained of difficulty swallowing liquid food which had been felt since 1 year ago and had worsened since 1 week ago, the patient had difficulty swallowing liquid food, the stomach felt full and bloated, nauseous, vomiting and felt a bit tight, the patient also complained of experiencing weight loss of around 5 kg after experiencing the complaint and did not improve with PPI treatment.

Radiological supporting examinations that should be performed to diagnose patients with suspected esophageal achalasia include: 1) Plain chest x-ray: in this examination, signs of inhalation pneumonia, dilation of the esophagus behind the heart may be found. 2) Esophagram: in this examination, a dilated and elongated esophagus with a narrowed "bird's beak" appearance at the cardia can be seen. 3) Esophagoscopy: in the examination, food remains were found in the dilated esophagus accompanied by esophagitis. The LES was found to be narrowed and to rule out differential diagnoses such as esophageal carcinoma or other diseases. From this case, it is strongly suspected that the patient was diagnosed with esophageal achalasia from the appropriate history and also proven by endoscopic examination which showed a narrowed 1/3 of the lower lumen of the esophagus.

The goal of achalasia therapy is to relieve symptoms by eliminating outflow resistance, caused by a non-relaxed lower esophageal sphincter (LES). Pharmacological treatment can be used to reduce LES pressure with calcium channel blockers and nitrates, but only about 10% of these therapies provide benefits to patients and are usually given to elderly patients who cannot tolerate surgery. In addition, there is treatment with botulinum toxin injections into the LES to block acetylcholine receptors and restore the balance between excitatory and inhibitory neurotransmitters, but only provides limited results and often patients require repeated toxin injections. Surgical therapy is performed with Heller cardiomyotomy and dor fundoplication. In this operation, the esophageal muscle in the valve area between the esophagus and stomach is incised until it reaches the esophageal submucosa, followed by a dor fundoplication procedure, which then pulls the gastric fundus forward to the esophagus (anterior) 180 degrees to prevent complaints of gastroesophageal reflux postoperatively. Postoperative complications can occur persistent obstruction, this is caused by ineffective peristalsis in long esophageal

achalasia or incomplete myotomy. Myotomy to the stomach wall can cause gastroesophageal reflux. Postoperative evaluation is carried out referring to the Ekcard Score looking at patient complaints after surgery and to assess the success of the operation.

Achalasia Severity: Eckardt Score

	Score for each symptom/sign			
Symptom/Sign	0	1	2	3
Recent weight loss (Kg)	none	< 5	5-10	>10
Dysphagia	none	occasional	daily	each meal
Chest pain	none	occasional	daily	several times/day
Regurgitation	none	occasional	daily	each meal

Figure 4. Schematic illustration of Heller Cardiomyotomy and Dor Fundoplication

Eckardt symptom score	Clinical stage	Category
0 to 1	0 (zero)	Remission
2 to 3	I (one)	Remission
4 to 6	II (two)	Treatment failure
7 to 12	III (three)	Treatment failure

Figure 5. Post-operative patient monitoring and outcome assessment
Source: Patient's Medical Record

Post-operatively, the patient was treated with IVFD totusol 28 tpm, fasting for 6 hours, liquid diet, ceftriaxone 2x1 gr, lansoprazole 2x30 mg iv, pain loss 3x1 gram. Post-operative follow-up The patient's complaints improved while in the treatment room. The patient had no complaints, no nausea, no vomiting, no shortness of breath, and no chest pain. There was no pain swallowing with an Eckardt score of 0.

CONCLUSION

Esophageal achalasia is a rare motility disorder characterized by the inability of the lower esophageal sphincter (LES) to relax, leading to symptoms such as dysphagia, regurgitation, and chest pain. The primary treatment goal is to relieve outflow obstruction at the LES to improve esophageal emptying. While nonsurgical treatments like pneumatic dilation and pharmacotherapy (calcium channel blockers, nitrates) can provide symptom relief, surgical cardiomyotomy with the Heller procedure combined with fundoplication remains the preferred definitive intervention, especially for durable symptom control and reducing reflux. Future

research should focus on optimizing surgical techniques and comparing long-term outcomes between laparoscopic Heller myotomy with fundoplication and emerging minimally invasive methods such as peroral endoscopic myotomy (POEM), with attention to quality of life and reflux management post-procedure.

REFERENCES

Barlow, J. M., Craig, D. A., Lowe, V. J., & MacCarty, R. L. (2019). Radiology of the esophagus: Barium, computed tomography scan, positron emission tomography scan, magnetic resonance imaging. In Shackelford's Surgery of the Alimentary Tract, 2 Volume Set (pp. 57–84). Elsevier.

Casanova, J.-L., & Abel, L. (2021). Mechanisms of viral inflammation and disease in humans. Science, 374(6571), 1080–1086.

Chalazonitis, A., & Rao, M. (2018). Enteric nervous system manifestations of neurodegenerative disease. Brain Research, 1693, 207–213.

Doubova, M., Gowing, S., Robaidi, H., Gilbert, S., Maziak, D. E., Shamji, F. M., Sundaresan, R. S., Villeneuve, P. J., & Seely, A. J. E. (2021). Long-term symptom control after laparoscopic Heller myotomy and Dor fundoplication for achalasia. The Annals of Thoracic Surgery, 111(5), 1717–1723.

Dumbuya, J. S., Zeng, C., Deng, L., Li, Y., Chen, X., Ahmad, B., & Lu, J. (2025). The impact of rare diseases on the quality of life in paediatric patients: current status. Frontiers in Public Health, 13, 1531583.

Gaber, C. E., Eluri, S., Cotton, C. C., Strassle, P. D., Farrell, T. M., Lund, J. L., & Dellon, E. S. (2022). Epidemiologic and economic burden of achalasia in the United States. Clinical Gastroenterology and Hepatology, 20(2), 342–352.

Hamad, M. D., Ramanathan, R., & Bestoun, H. (2017). 2017 Scientific Session of the Society of American Gastrointestinal and Endoscopic Surgeons (SAGES) Houston, Texas, USA, 22–25 March 2017. Surg Endosc, 31, S68–S107.

Kahrilas, P. J., Bredenoord, A. J., Fox, M., Gyawali, C. P., Roman, S., Smout, A. J. P. M., Pandolfino, J. E., & Function, I. W. G. for D. of G. M. and. (2017). Advances in the management of oesophageal motility disorders in the era of high-resolution manometry: a focus on achalasia syndromes. Nature Reviews Gastroenterology & Hepatology, 14(11), 677–688.

Kloner, R. A., Goggin, P., Goldstein, I., Hackett, G., Kirby, M. G., Osterloh, I., Parker, J. D., & Sadovsky, R. (2018). A new perspective on the nitrate–phosphodiesterase type 5 inhibitor interaction. Journal of Cardiovascular Pharmacology and Therapeutics, 23(5), 375–386.

Lerman, B. B. (2015). Mechanism, diagnosis, and treatment of outflow tract tachycardia. Nature Reviews Cardiology, 12(10), 597–608.

Niebisch, S., Hadzijusufovic, E., Mehdorn, M., Müller, M., Scheuermann, U., Lyros, O., Schulz, H. G., Jansen-Winkeln, B., Lang, H., & Gockel, I. (2017). Achalasia—an unnecessary long way to diagnosis. Diseases of the Esophagus, 30(5), 1–6.

Pandolfino, J. E., & Gawron, A. J. (2015). Achalasia: a systematic review. Jama, 313(18),

1841–1852.

Pesce, M., Pagliaro, M., Sarnelli, G., & Sweis, R. (2023). Modern achalasia: diagnosis, classification, and treatment. Journal of Neurogastroenterology and Motility, 29(4), 419.

Pham, T., Telias, I., & Beitler, J. R. (2020). Esophageal manometry. Respiratory Care, 65(6), 772–792.

Rebecchi, F., Allaix, M. E., Schlottmann, F., Patti, M. G., & Morino, M. (2018). Laparoscopic Heller myotomy and fundoplication: what is the evidence? The American Surgeon, 84(4), 481–488.

Ribolsi, M., Andrisani, G., Di Matteo, F. M., & Cicala, M. (2023). Achalasia, from diagnosis to treatment. Expert Review of Gastroenterology & Hepatology, 17(1), 21–30.

Rieder, E., Fernandez-Becker, N. Q., Sarosiek, J., Guillaume, A., Azagury, D. E., & Clarke, J. O. (2020). Achalasia: physiology and diagnosis. Annals of the New York Academy of Sciences, 1482(1), 85–94.

Uppal, D. S., & Wang, A. Y. (2016). Update on the endoscopic treatments for achalasia. World Journal of Gastroenterology, 22(39), 8670.

Wilkinson, J. M., & Halland, M. (2020). Esophageal motility disorders. American Family Physician, 102(5), 291–296.

Zullo, A., Manta, R., De Francesco, V., Fiorini, G., Hassan, C., & Vaira, D. (2019). Diagnostic yield of upper endoscopy according to appropriateness: a systematic review. Digestive and Liver Disease, 51(3), 335–339.