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CASE REPORT

Case Report and Therapeutic Approach for Achalasia Cardia

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Abstract: Achalasia cardia is an uncommon esophageal motility disorder characterized by the impaired relaxation of the lower esophageal sphincter (LES) and the absence of peristaltic contractions in the esophageal body. This report presents a case of a 42-year-old woman with a prolonged history of dysphagia and regurgitation of solids and liquids. The patient's clinical history included a 10-year history of neck swelling, hypothyroidism, and diabetes mellitus. Physical examination and laboratory investigations revealed normal findings, except for elevated blood glucose, thyroid hormone levels, and thyroid-stimulating hormone (TSH) levels. An esophagogram showed proximal dilation of the distal esophagus with a contrast fluid level, and endoscopy revealed a tight LES, which was difficult to pass through. Based on these findings, the diagnosis of achalasia cardia was established. Initial medical management with nitrates and proton pump inhibitors did not provide significant improvement. Subsequently, the patient underwent laparoscopic Heller's myotomy with fundoplication, which successfully resolved her dysphagia symptoms. This case report highlights the diagnostic challenges and the importance of a multidisciplinary approach in managing achalasia cardia. It emphasizes the role of surgical interventions, such as Heller's myotomy with fundoplication, in alleviating symptoms and restoring esophageal function when conservative medical management fails.

Keywords: Achalasia cardia; Dysphagia; Esophageal motility disorder; Heller's myotomy; Fundoplication

1. Introduction

Achalasia cardia is a rare esophageal motility disorder characterized by the impaired relaxation of the lower esophageal sphincter (LES) and the absence of peristaltic contractions in the esophageal body. This condition leads to a functional obstruction of the esophagogastric junction, resulting in difficulty swallowing (dysphagia) and regurgitation of undigested food and liquids. [1,2] The exact etiology of achalasia cardia remains unclear, but several theories have been proposed. One hypothesis suggests that the degeneration of the myenteric plexus and vagus nerve fibers innervating the LES may play a crucial role in the pathogenesis of the disease. This neuronal degeneration can lead to the impaired relaxation of the LES and the subsequent loss of peristaltic contractions in the esophageal body. [3] Another proposed mechanism involves autoimmune factors, where an autoimmune response against the neural structures of the esophagus may trigger the degenerative process. Additionally, viral infections, such as the varicella-zoster virus or herpes simplex virus, have been implicated as potential triggers for the development of achalasia cardia in some cases. [4]

Achalasia cardia is a progressive disorder, and if left untreated, it can lead to significant morbidity and complications. As the disease progresses, the esophagus may become dilated and tortuous, increasing the risk of stasis, fermentation, and potential aspiration of retained food and liquids.[5] Other complications may include weight loss, malnutrition, esophageal ulceration, and an increased risk of developing esophageal cancer. The diagnosis of achalasia cardia is typically based on a combination of clinical symptoms, radiological findings, and esophageal manometry. Barium swallow studies can reveal a dilated esophagus with a narrow, bird-beak appearance of the distal esophagus due to the non-relaxing LES. Endoscopy may show a dilated esophagus with retained food and liquids, as well as a tightly closed LES. However, esophageal manometry remains the gold standard for diagnosing achalasia cardia, as it can demonstrate the characteristic findings of aperistalsis in the esophageal body and impaired relaxation of the LES. The management of achalasia cardia involves a multidisciplinary approach, with treatment options ranging from pharmacological interventions to endoscopic procedures and surgical interventions. While medications such as nitrates and calcium channel blockers can provide temporary relief by relaxing the LES, their efficacy is often limited, and they do not address the underlying pathophysiology of the disease. [6]

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2. Case presentation

A 42-year-old woman presented to the general surgery ward of a tertiary care hospital with a prolonged history of dysphagia for both solids and liquids, accompanied by regurgitation of liquids. The patient reported experiencing similar symptoms since childhood, which had gradually worsened over time. In addition to her primary complaint, the patient's medical history revealed the presence of a neck swelling for the past 10 years, hypothyroidism for 5 years, and diabetes mellitus for 3 years. Physical examination findings were largely unremarkable, with a blood pressure of 120/80 mmHg, a pulse rate of 78 beats per minute, and an oxygen saturation level of 98%. Systemic examination did not reveal any significant abnormalities.

Laboratory investigations showed elevated levels of fasting blood sugar (149 mg/dL) and postprandial blood sugar (213 mg/dL), suggesting suboptimal glycemic control. Thyroid function tests revealed a low triiodothyronine (T3) level of 0.7 nmol/L, a high thyroxine (T4) level of 151.35 nmol/L, and an elevated thyroid-stimulating hormone (TSH) level of 8.51 mIU/L, indicative of hypothyroidism. Other laboratory parameters, including lipid profile, liver function tests, and renal function tests, were within normal ranges. The patient's initial presentation and clinical history raised suspicion of an underlying esophageal motility disorder, prompting further investigations to establish a definitive diagnosis and guide appropriate management.[1,7]

3. Investigations

To investigate the patient's dysphagia and regurgitation symptoms, several diagnostic tests were performed. An esophagogram, also known as a barium swallow study, revealed proximal dilation of the distal esophagus with a contrast fluid level, suggesting obstruction at the lower esophageal sphincter (LES) level. Furthermore, an upper endoscopy was conducted, which demonstrated a tight LES (Figure 1) that was difficult to traverse with the endoscope. This finding, combined with the esophagogram results, strongly supported the suspected diagnosis of achalasia cardia. Although endoscopic and radiographic findings were suggestive of achalasia, esophageal manometry was performed to confirm the diagnosis definitively. [8,9] Esophageal manometry is considered the gold standard for diagnosing achalasia cardia, as it can demonstrate the characteristic features of aperistalsis in the esophageal body and impaired relaxation of the LES during swallowing. [10,11]

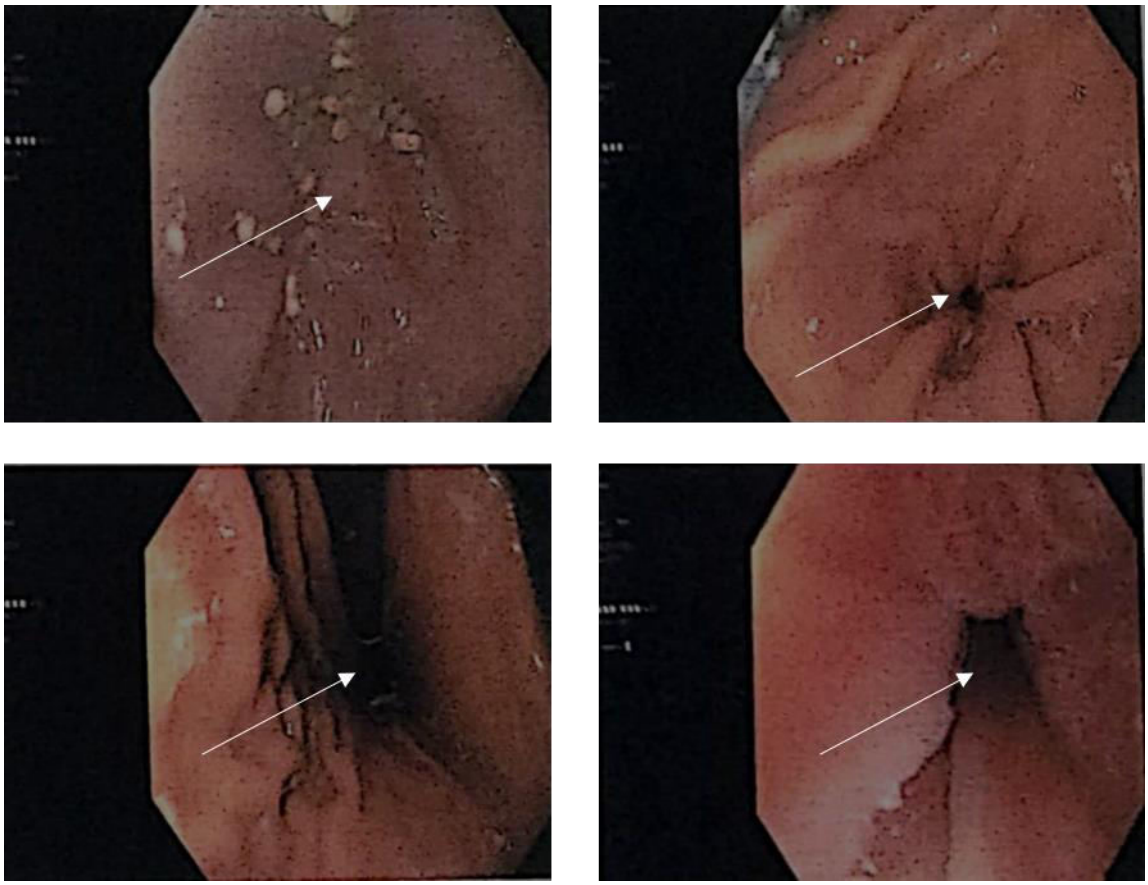

Figure 1. Endoscopy showing dilatation on the lower third of the esophagus

4. Treatment

Based on the clinical presentation, radiological findings, and endoscopic evaluation, the patient was diagnosed with achalasia cardia. Initial management involved conservative medical treatment with nitroglycerin (2.6 mg once daily) and esomeprazole magnesium (40 mg once daily) to alleviate symptoms and reduce lower esophageal sphincter pressure. Additionally, the patient was prescribed metformin (500 mg once daily) for diabetes management and thyronorm (100 mcg once daily) for hypothyroidism. Despite medical therapy, the patient's condition did not improve significantly, and her dysphagia persisted. Consequently, a decision was made to pursue surgical intervention to address the underlying pathology and restore esophageal function.

The patient underwent laparoscopic Heller's myotomy with fundoplication, a minimally invasive surgical procedure aimed at relieving the obstruction caused by the non-relaxing LES. During the procedure, the LES muscle fibers are cut (myotomy) to eliminate the functional obstruction, and a partial fundoplication is performed to prevent gastroesophageal reflux. Heller's myotomy is considered the standard surgical treatment for achalasia cardia, as it effectively addresses the underlying pathophysiology by disrupting the non-relaxing LES and allowing for improved passage of food and liquids into the stomach. [12-14] The addition of fundoplication helps to prevent the potential complication of gastroesophageal reflux disease (GERD) after myotomy. The surgical procedure was performed without any major complications, and the patient tolerated the operation well. Postoperatively, the patient was prescribed antibiotics (cefixime 200 mg twice daily), a proton pump inhibitor (esomeprazole magnesium 40 mg once daily), thyroxine (100 mcg once daily), sucralfate suspension (10 ml twice daily), and analgesics (tramadol 37.5 mg + acetaminophen 325 mg) as needed for pain management.

5. Outcome

Following the laparoscopic Heller's myotomy with fundoplication, the patient's dysphagia symptoms resolved promptly, indicating a successful surgical outcome. The patient was able to tolerate oral intake without significant difficulty, marking a significant improvement in her quality of life. The patient remained hospitalized for 15 days postoperatively for monitoring and management of any potential complications. During this period, her recovery was uneventful, and she was discharged with a comprehensive medication regimen and instructions for follow-up. At the time of discharge, the patient was prescribed cefixime (200 mg twice daily) as an antibiotic, esomeprazole magnesium (40 mg once daily) as a proton pump inhibitor, thyroxine (100 mcg once daily) for hypothyroidism management, sucralfate suspension (10 ml twice daily) for gastric mucosal protection, and tramadol (37.5 mg) with acetaminophen (325 mg) as needed for pain relief. The patient was advised to attend regular follow-up visits at the outpatient clinic on a monthly basis to monitor her progress, manage any residual symptoms, and address any potential complications that may arise.

6. Discussion

Achalasia cardia is a rare esophageal motility disorder that poses significant diagnostic and therapeutic challenges. [15, 16] The case presented here highlights the importance of a thorough clinical evaluation, appropriate diagnostic investigations, and a multidisciplinary approach to managing this condition effectively. The patient's presenting symptoms of dysphagia and regurgitation, coupled with the radiological and endoscopic findings, were suggestive of an esophageal motility disorder. However, esophageal manometry played a crucial role in confirming the diagnosis of achalasia cardia by demonstrating the characteristic features of aperistalsis and impaired LES relaxation. Initial medical management with nitrates and proton pump inhibitors aimed to provide symptomatic relief and reduce LES pressure. However, these conservative measures proved ineffective in this case, highlighting the limitations of medical therapy in managing advanced or refractory cases of achalasia cardia.

Surgical intervention, specifically laparoscopic Heller's myotomy with fundoplication, emerged as the definitive treatment option for this patient. Heller's myotomy addresses the underlying pathophysiology by disrupting the non-relaxing LES, allowing for improved passage of food and liquids into the stomach. [17,18] The addition of fundoplication helps mitigate the risk of postoperative gastroesophageal reflux disease (GERD), a common complication after myotomy. The successful resolution of the patient's dysphagia following the surgical procedure underscores the importance of timely and appropriate surgical intervention in cases of achalasia cardia that do not respond to conservative medical management. This case also highlights the value of a multidisciplinary team approach, involving gastroenterologists, surgeons, and other healthcare professionals, in ensuring optimal patient outcomes. [19, 20] While the exact etiology of achalasia cardia remains unclear, several theories have been proposed, including neuronal degeneration, autoimmune factors, and viral infections. However, further research is needed to fully understand the underlying pathogenic mechanisms and explore potential preventive or therapeutic strategies targeting these mechanisms. It is essential to recognize that achalasia cardia is a progressive disorder, and if left untreated, it can lead to significant morbidity and complications, such as esophageal dilation, stasis, aspiration, malnutrition, and an increased risk of esophageal cancer. Early diagnosis and prompt management are crucial to prevent these potential complications and improve the patient's quality of life.

7. Conclusion

Achalasia cardia is a rare and challenging esophageal motility disorder that requires a comprehensive diagnostic approach and multidisciplinary management. This case report illustrates the importance of surgical intervention, specifically laparoscopic Heller's myotomy with fundoplication, in cases refractory to conservative medical therapy. Early recognition and prompt treatment are essential to prevent potential complications and improve the patient's quality of life. Continued research efforts are needed to further elucidate the underlying pathogenic mechanisms and explore novel therapeutic strategies for this debilitating condition

Compliance with ethical standards

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Conflict of interest statement

The authors declare no conflict of interest.

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Author's short biography

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