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Case Report **General Surgery**

Achalasia: Case Report and Literature Review

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Abstract

Background*:** Achalasia is a medical ailment characterized by irregular contractions of the esophagus and incomplete relaxation of the lower esophageal sphincter. This condition results in difficulty swallowing food and liquids, and often leads to regurgitation and heartburn. ***Case presentation*:** This case report describes a 36-year-old female patient who presented with difficulty swallowing, heartburn, chest pain, and coughing while eating or drinking. The patient had lost significant weight despite a good appetite, and a physical examination revealed poor nutrition. Esophageal manometry confirmed the diagnosis of Achalasia Type 1, and endoscopy showed severe Candida esophagitis and a very dilated esophagus with diverticula. Due to the patient's condition and endoscopic finding of diverticula, surgery was considered, as she did not qualify for the Peroral Endoscopic Myomectomy (POEM) procedure. The patient was treated with fluconazole to address the esophageal candidiasis, and NG feeding was initiated to improve weight and nutrition. After recovering, the patient underwent a laparoscopic Heller myotomy with Dor fundoplication using a video laryngoscope to facilitate intubation. Anesthesia was maintained with O2/air with Sevoflurane, and the patient was extubated and transferred to the post-anesthesia care unit. ***Conclusion: Achalasia is a frequently researched esophageal motility disorder that is distinguished by inadequate relaxation of the LES and absent or irregular peristalsis in the esophagus. Common symptoms in most patients include difficulty swallowing solids and liquids, regurgitation, and varying degrees of weight loss. This case highlights the importance of prompt diagnosis and treatment of Achalasia Type 1, as well as the use of rapid sequence induction and proper anesthesia techniques during surgery.

Keywords: Achalasia, Laparoscopy, Anesthesia, Laparoscopic surgery.

INTRODUCTION

The term "achalasia" originates from the Greek word "khalasis," which means "not loosening or relaxing." Historically, achalasia has been defined as the lower esophageal sphincter's inability to relax in the absence of peristalsis [1]. It's characterized by a lack of proper muscle contractions in the esophagus and difficulties in relaxing the lower esophageal sphincter during swallowing [2]. The global yearly occurrence of achalasia is around 1 in 100,000 individuals, with an overall prevalence of 9 to 10 in 100,000 individuals [3]. Achalasia does not show a predilection for a particular gender, race, or age. According to recent research carried out in the United States, there has been a notable surge in the number of individuals being admitted to hospitals as well as a corresponding increase in the costs incurred as a result of treating achalasia, over the course of the past sixteen years, particularly in racial minorities and patients under 65 years of age [3].

Although the reason is unknown, the achalasia incidence is higher in spinal cord injury patients, particularly those with damage to the cervical and thoracic vertebrae [4]. The prevalence of achalasia is known to fluctuate across countries outside the United States, ranging from 0.1 to 1 occurrence per 100,000 people on an annual basis. Researches reveal that relapse rates are higher following pneumatic dilatation, which is the initial treatment for achalasia [4].

Achalasia has 3 types based on manometric patterns: type I (classic) with minimal contractility, type II with intermittent pressurization, and type III (spastic)

with premature or spastic contractions in the lower esophagus. Achalasia is believed to result from the degeneration of vagus nerve fibers and myenteric nerve plexus located in the LES [5]. This degeneration causes a loss of inhibitory neurons containing nitric oxide synthase and vasoactive intestinal peptide in the "esophageal myenteric plexus". In situations where the condition is particularly severe, the participation of cholinergic neurons is also observed [6]. However, the precise underlying cause of this degenerative process remains elusive, despite the proposal of various hypotheses including a viral infection, genetic susceptibility, and an autoimmune response [7]. In the United States, primary idiopathic achalasia is the most common type. However, secondary achalasia can occur due to Chagas disease caused by neurodegenerative disorders, viral infections, lymphoma, eosinophilic gastroenteritis, esophageal infiltration by gastric carcinoma, and Trypanosoma cruzi [8].

Dysphagia is the primary symptom in most patients with achalasia, initially with solids and then progressing to liquids. Studies propose that at presentation, 70-97% of patients can have dysphagia for both solids and liquids [9]. Dysphagia and regurgitation are the most common presenting symptoms of achalasia. Chest pain is also prevalent in more than half of the patients, but improving esophageal emptying rarely relieves the pain [10, 11]. Less common symptoms include difficulty belching and hiccups [12].

Diagnostic studies are crucial to confirm the diagnosis of achalasia, as symptoms are not reliable. A barium esophagogram is the best initial test, showing classic findings such as the "bird's beak" appearance and lack of peristalsis [12]. Upper endoscopy is recommended to exclude malignancy. Esophageal manometry is the most sensitive test and remains the gold standard, with high-resolution manometry being the preferred method [13]. Regarding treatment, different management options are available to ease symptoms by decreasing outflow resistance. Nonsurgical options include pharmacotherapy with phosphodiesterase-5 inhibitors, calcium channel blockers, and nitrates [14]. Surgical approaches include the POEM procedure and laparoscopic Heller myotomy [15]. Nitrates, calcium channel blockers, and botulinum toxin injection provide only temporary relief and are predominantly limited to patients waiting for or who refuse more definitive therapy [16]. Pneumatic dilatation is the most cost-effective non-surgical option with long-term symptom relief, but repeat treatments may be necessary. LHM with partial fundoplication is the preferred surgical procedure with a high success rate, while POEM is an effective minimally invasive alternative but carries a high risk of gastroesophageal reflux [4].

CASE PRESENTATION

A 36-year-old female patient with no known past medical history presented with a two-week history of difficulty swallowing and eating and drinking. She reported feeling like food was stuck in her neck and chest, along with heartburn and chest pain. She also experienced coughing while eating or drinking and had lost around 12 kilograms in one month with a good appetite. She had a history of vomiting unchewed food but denied any fever, nausea, or bowel changes. Her past medical history was unremarkable, and the patient had no known allergies, was not on any regular medications, and only took PRN paracetamol and Vitamin C. She was married and had one child. Physical examination revealed poor nutrition, with a weight of 42 kg and a height of 165 cm. Vitals were RR18/min, HR72/min, BP107/69, SpO2 97%, hypovolemia, and expected difficult intubation.

Later esophageal manometry was performed and it confirmed Achalasia Type 1. After the diagnosis, an NG feeding tube was inserted via endoscopy with the tip in the small intestine. During endoscopy, severe Candida esophagitis was also noted, along with a very dilated esophagus with diverticula. Due to this, the patient did not qualify for the POEM procedure (Peroral Endoscopic Myomectomy) and was treated with fluconazole for 21 days to address the esophageal candidiasis. The decision was made to continue with NG feeding to improve weight and nutrition, and surgery was considered for the patient.

The intubation was performed with a rapid sequence induction technique. When the patient reached the OT, she was placed in a supine position with her head elevated 20 degrees, connected to essential monitoring, and preoxygenated for 5 minutes. Upper airway aspiration was performed, and 90 mg propofol was injected, followed by 50 mg succinylcholine. Experienced assistants applied coracoid pressure, and then laryngoscopy done and classified as Lehen-Comark 4 so a gum elastic bougie was inserted, an ETT size 7 was railroaded on the bougie, and the bougie was removed. Fentanyl 50mcg was given, and esophagogastroduodenoscopy was performed uneventfully. The patient recovered and was transferred to the PACU (Post Anesthesia Care Unit).

A few weeks later, laparoscopic Heller myotomy with Dor fundoplication was performed on the patient. The patient was given general anesthesia and the rapid sequence induction technique was used. A video laryngoscope was used, making the intubation easier. After securing the tube, cisatracurium 6 mg and fentanyl 100mcg were given, and anesthesia was maintained with O2/air with Sevoflurane for a 3-hour duration. Before the end of the operation, 500 paracetamol, 30 mg 50 mg Pethidine IM, 4 mg dexamethasone, and 4 mg ondansetron were administered. The patient was extubated at the table and transferred to the PACU.

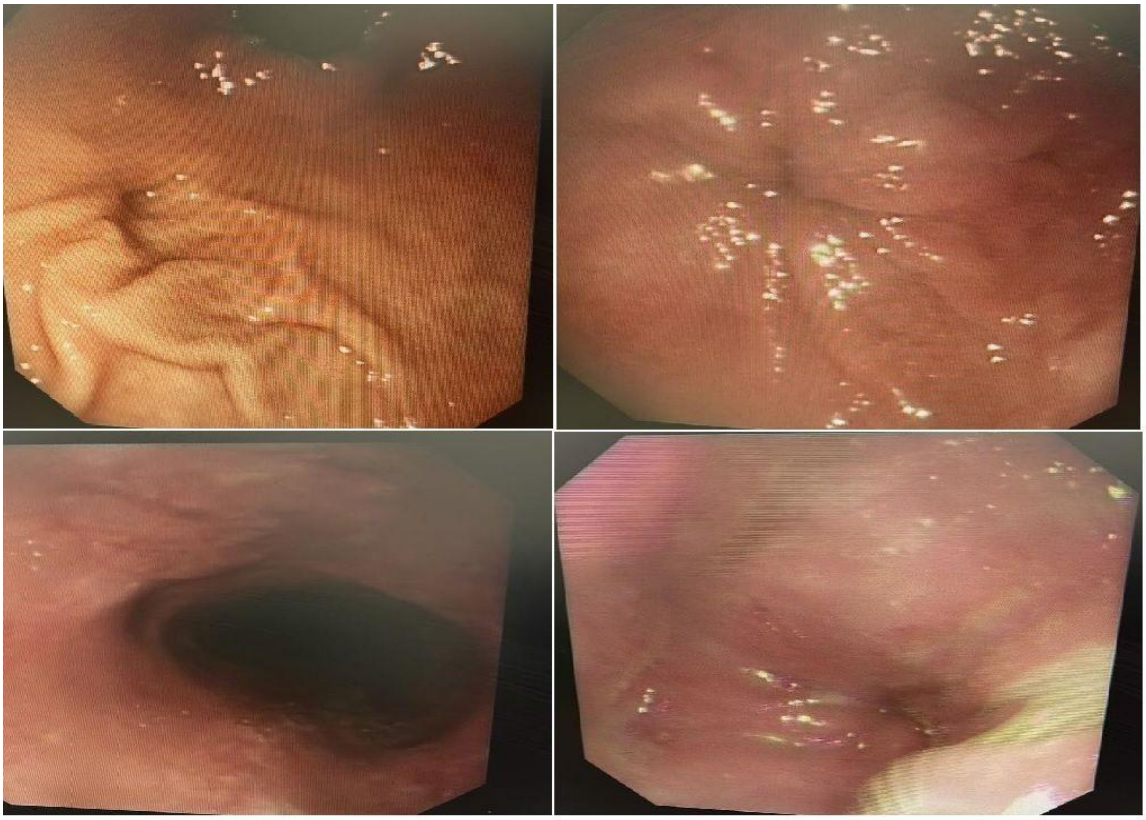

Fig 1: Endoscopic findings in esophageal achalasia. a: Functional stenosis of the esophagogastric junction. b: Wrapping around the esophagogastric junction. c: Abnormal contraction of the esophageal body. d: Mucosal thickening and whitish change

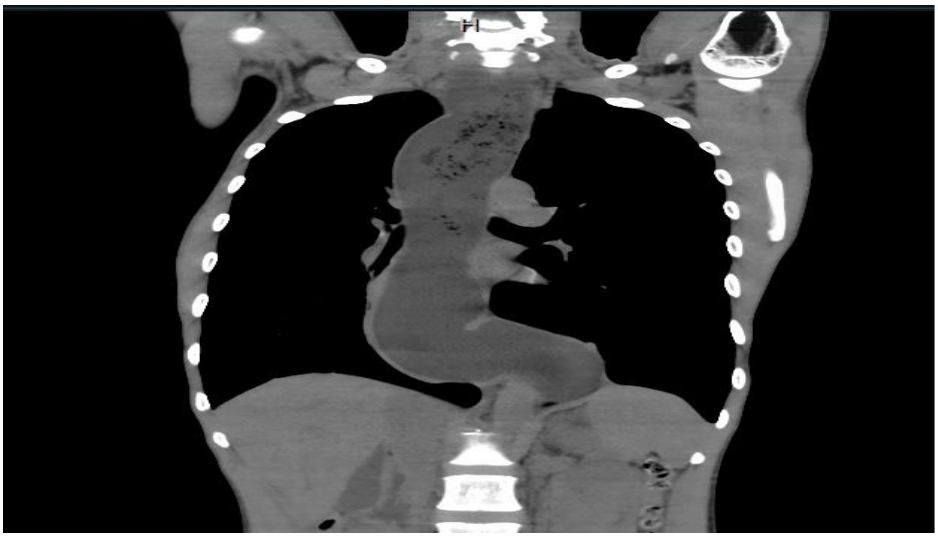

Fig 2: CT chest with contrast showing Achalasia with Dilated Esophagus

DISCUSSION

The case described is of a 36-year-old female patient with difficulty swallowing and eating/drinking, heartburn, weight loss, chest pain, and coughing. The physical examination showed poor nutrition and hypovolemia. The patient was diagnosed with "Achalasia Type 1" via esophageal manometry and severe Candida esophagitis was also noted during endoscopy. NG feeding was initiated, and surgery was considered. Rapid sequence induction was performed during surgery, and the patient was successfully extubated at the table and transferred to the PACU.

Achalasia is a rare disorder that affects the esophagus and impairs its ability to move food toward the stomach. Achalasia can manifest in various ways negatively affecting a patient's quality of life, work performance, and ability to function effectively [17]. The hallmark symptom of achalasia is progressive difficulty swallowing both solids and liquids [18].

Approximately 27% to 42% of achalasia patients may also experience heartburn, leading to a misdiagnosis of gastroesophageal reflux disease and subsequent treatment with proton pump inhibitors [19]. A misdiagnosis can cause a significant delay in diagnosing achalasia until patients experience persistent symptoms, prompting further diagnostic evaluations [19]. Dysphagia and regurgitation are common across all age groups, but younger patients may also experience chest pain and heartburn, while obese patients (BMI ≥30) may report more frequent episodes of choking or vomiting [20]. Women and individuals with type III achalasia may be more prone to chest pain. In addition, the initial presentation of achalasia can result in weight loss, with reports indicating that 35% to 91% of patients experience weight loss, and the degree of weight loss can vary widely, with an average of 20 ± 16 lbs [21]. Our patient also faced difficulty swallowing and eating and drinking. She reported feeling like food was stuck in her neck and chest, along with heartburn and chest pain. She also experienced coughing while eating or drinking and had lost around 12 kilograms in one month with a good appetite.

In situations where achalasia is suspected, the clinical presumption can be corroborated by performing a barium esophagram, which is regarded as the most efficient diagnostic procedure [22]. The features of achalasia include the expansion and convolutions of the upper esophagus, which gradually tapers down towards the lower end and bears a striking resemblance to a sharpened wooden pencil tip, popularly known as the "Rat Tail Appearance [23]." To rule out pseudo achalasia, it is recommended that all patients undergo an endoscopic examination, which assesses the gastric cardia and gastroesophageal junction [23]. Oesophageal pressure manometry is also necessary, as it establishes the diagnosis by showing pressure in the gastroesophageal junction to be about twice the normal level (40 mm of Hg), with incomplete or absent relaxation after swallowing [24]. In the present case, dilation of the esophagus was noted on endoscopy. On Endoscopy, candida esophagitis was also observed [25]. The primary cause of infectious esophagitis is Candida albicans, which is responsible for the majority of cases [26]. This infection is typically associated with a compromised immune system in most patients. However, in approximately 25% of cases, esophageal stasis resulting from underlying conditions such as achalasia and scleroderma can also contribute to fungal colonization of the esophagus [26]. The infection can cause symptoms such as difficulty swallowing, chest pain, and heartburn.

Anesthesia plays a crucial role in the management of achalasia [27]. Diagnostic or therapeutic procedures for achalasia often require anesthesia to help relax the LES muscles, reduce the risk of complications, and provide greater comfort to the patient. Conscious sedation is a common anesthesia method for patients with achalasia. It involves administering medications through an IV to help the patient relax and feel less anxious during the procedure. However, conscious sedation is not the preferred method for patients with achalasia because it can increase the risk of aspiration [28]. Alternatively, general anesthesia can be used in achalasia patients. This type of anesthesia involves the use of medications to cause the patient to lose consciousness during the procedure, and it is typically used to protect the patient against aspiration.

The objective of achalasia therapy is to alleviate symptoms by eliminating outflow resistance, which is caused by the hypertensive and nonrelaxing lower esophageal sphincter (LES). Lowering LES pressure can be achieved using calcium channel blockers and nitrates [23]. However, only about 10% of patients benefit from this treatment and it is typically used in elderly patients who are not suitable for pneumatic dilatation or surgery. Endoscopic treatment involves injecting botulinum toxin into the LES to block acetylcholine release and restore the balance between excitatory and inhibitory neurotransmitters. However, this treatment has limited value, and only about 30% of patients experience relief of dysphagia one year after treatment. Most patients require repeated botulinum toxin injections [21]. Pneumatic dilatation, performed by a qualified gastroenterologist, is the recommended treatment for sporadic cases where surgery is not appropriate [29]. A laparoscopic Heller myotomy is considered the primary treatment for achalasia by many experts. However, a partial fundoplication and Heller myotomy performed from the chest (thoracoscopic) have a high incidence of gastroesophageal reflux [29].

Rapid sequence induction is a technique used to secure the patient's airway quickly and safely during anesthesia induction. It involves administering a sedative and a muscle relaxant, followed by intubation, to prevent the patient from aspirating stomach contents into the lungs [30]. The technique is commonly used in patients who are at a high risk of aspiration, such as those with achalasia, and those undergoing surgery that may cause gastric contents to enter the respiratory tract [30]. In the current case, the patient underwent rapid sequence induction and was intubated using a gum elastic bougie and Lehen-Comark 4 laryngoscopy. After recovering, the patient underwent laparoscopic Heller myotomy with Dor fundoplication using a video laryngoscope to facilitate intubation. Anesthesia was maintained with O2/air with Sevoflurane, and the patient was extubated and transferred to the post-anesthesia care unit. Later, the patient made a full recovery.

It should be noted that, currently available treatments for achalasia do not aim to halt disease progression but rather to alleviate symptoms such as

regurgitation, chest pain, and dysphagia as well as prevent complications such as megaesophagus, weight loss, and gastroesophageal reflux disease [4]. Patients should be informed that achalasia is a chronic condition, and treatment outcomes may be focused on symptom relief rather than a complete cure. Following myotomy, patients must make necessary lifestyle changes, including eating small food portions in an upright position, which helps to facilitate gravity-assisted food transit and avoid lying flat but instead maintain a 30 to 45-degree angle to minimize the risk of aspiration.

CONCLUSION

Achalasia is a frequently researched esophageal motility disorder that is distinguished by inadequate relaxation of the LES and aperistalsis in the esophagus. Common symptoms in most patients include difficulty swallowing solids and liquids, regurgitation, and varying degrees of weight loss. This case highlights the importance of prompt diagnosis and treatment of Achalasia Type 1, as well as the use of rapid sequence induction and proper anesthesia techniques during surgery. The use of rapid sequence induction technique in laparoscopic Heller myotomy with Dor fundoplication is helpful in managing the patient's symptoms.

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