Radiology Case Reports 19 (2024) 39–43

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

A rare case of mega-esophagus due to achalasia causing tracheal compression ☆

*Leni Santiana, MD, Filomena Fabiola Kusuma, MD**

Department of Radiology, Faculty of Medicine, Padjadjaran University, Bandung, Indonesia

ARTICLE INFO

Article history:
Received 18 July 2023
Revised 19 September 2023
Accepted 25 September 2023
Available online 28 October 2023

Keywords:
Achalasia
Heller myotomy
Respiratory distress
Tracheal compression

ABSTRACT

Achalasia is one of the most common esophageal motility disorders. Typical symptoms include dysphagia, food regurgitation, respiratory symptoms, chest pain, and weight loss. Respiratory obstruction due to tracheal compression by the massively dilated esophagus is a very rare but fatal complication. A 36-year-old male presented with progressive respiratory distress with a history of untreated dysphagia and regurgitation. Further diagnosis revealed dilatation of the esophagus with undigested food. A Heller myotomy with fundoplication was performed and respiratory symptoms were relieved. Tracheal compression and acute airway obstruction caused by esophageal dilatation in achalasia is a rare presentation. Early recognition of this rare manifestation is critical and emergency treatment is necessary for life saving. Radiological examination can help physicians find the dilated esophagus. Respiratory symptoms resulting from tracheal compression by a dilated esophagus rarely occurred. Even though physicians should be alert and early decompression has to be performed immediately.

Introduction

Achalasia is one of the most common esophageal motility disorders, affecting approximately 1 in 100,000 individuals a year [1,2]. It is usually diagnosed between 20 and 50 years of age but can occur at any age with no predilection for either sex [1–3]. The etiology of achalasia is still vague, and its precise pathogenesis remains ambiguous to this day [3–5]. Histologic analysis reveals that achalasia is the result of the degeneration of ganglion cells in the myenteric plexus of the esophageal body and the lower esophageal sphincter [2,6,7]. The disorder is characterized by an impaired lower esophageal sphincter relaxation and the absence of esophageal peristalsis, resulting in esophageal outflow obstruction [1,4,5].

In 1672, Sir Thomas Willis first described the condition as “cardiospasm,” which he treated by dilating the esophagus with a sponge attached to a whale bone [1,3]. It was not until 1922 that AF Hurst discovered that the motility disorder was due to the LES’s inability to relax, and named it “achalasia” (from the Greek a-khalasis, “lack of relaxation”) [1,4,8].

☆ Competing Interests: There are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

* Corresponding author.
E-mail address: filomena.ffk@gmail.com (F.F. Kusuma).

https://doi.org/10.1016/j.radcr.2023.09.081

Patients with suspected achalasia based on clinical presentation should always undergo an upper esophagogastroduodenoscopy to rule out pseudoachalasia from an obstructing mass. The diagnosis of achalasia is confirmed with high-resolution manometry, which is the current gold standard test [6,7].

The clinical presentation and symptoms of achalasia include slowly progressing dysphagia for solids and liquids, frequent food regurgitation, respiratory symptoms (nocturnal cough, recurrent aspiration and pneumonia), chest pain, and weight loss [1,4,5]. Respiratory obstruction due to tracheal compression by the massively dilated esophagus is a very rare but fatal complication [9].

Case report

A 36-year-old male was admitted for progressive respiratory distress. He had a 20-year history of untreated dysphagia and regurgitation. His dysphagia worsened over the course of 2 years, such he was restricted to a liquid diet. His symptoms also included dyspnea, which had steadily increased over 1 year. He had no history of previous respiratory or cardiac disease, nor any significant family and /or past medical history.

On physical examination, we found an increased respiratory rate with decreased oxygen saturation. Other vital signs were normal. Distention in the cervical region occurred on deep inspiration. He was also noted to have stridor, and his neck bulged bilaterally at the level of the larynx. No thyroid masses were detectable. Chest auscultation revealed decreased vesicular breath sounds bilaterally. The remainder of the physical examination was unremarkable. Laboratory results were within normal limits. Chest radiography showed a widened mediastinum with a massively dilated esophagus filled up to the neck with food residue (Fig. 1). Both lung fields were clear. A barium sulfate contrast examination showed a large elongated esophagus with food residue extending to the thoracic inlet, and a narrowing of about 2.4 cm long above the gastroesophageal junction (Fig. 2).

Computed tomography (CT) examination revealed a massive enlargement of the esophagus with the greatest diameter of about 11.3 cm, compressing the posterior tracheal wall and resulting in tracheal narrowing (Fig. 3). During esophagogastroduodenoscopy, the scope became stuck in the distal esophagus, owing to the narrowed esophageal passage and the presence of food residue. Upper endoscopy was conclusive of distal esophageal achalasia.

In emergency management, a nasogastric tube was inserted to decompress. The patient underwent Heller myotomy with fundoplication as a definitive therapy. Respiratory symptoms were relieved after surgery.

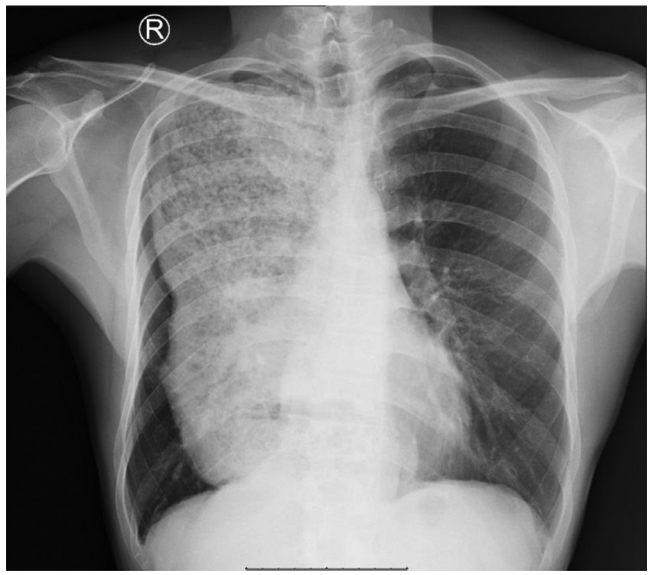

Fig. 1 – Chest radiograph shows a massively dilated esophagus filled with food residue.

Discussion

Achalasia is an idiopathic motility disorder of the esophagus with the cardinal symptom of dysphagia [1,4,5]. In addition, weight loss, vomiting, and respiratory complications occur due to regurgitation of food [5,10]. This may result in aspiration, pneumonia, bronchiectasis, and rarely, lung abscess [2,10]. Tracheal compression and acute airway obstruction caused by esophageal dilatation are rare complications of achalasia [10]. The presenting features in most reported cases have been of a patient with a history of dysphagia, cyanosis, stridor, respiratory distress, and frequently a soft asymmetric non-pulsatile cervical mass [11]. Achalasia presenting as an acute airway obstruction was first reported in 1950 in an elderly woman who presented with megaesophagus and respiratory distress [9–12].

The pathophysiology of achalasia leading to acute airway obstruction has not been clearly defined, though various hypotheses have been proposed. Firstly, it is thought to be caused by the cephalic displacement of the dilated esophageal portion, which then kinks behind the cricopharyngeus muscle. Presumably, this kinking forms a one-way valve that allows air to enter the esophagus but disallows its exit. Secondly, there may be an impairment in the relaxation of the upper esophageal sphincter (UES). This also disallows the escape of air, leading to a ballooning of the esophagus. The distended esophagus may then physically compress the upper airway and result in obstructive respiratory symptoms. The third hypothesis involves the belch reflex, which is the relaxation of the UES as a response to esophageal distention, resulting in decompression. In achalasia, where the belch reflex is impaired, decompression fails and megaesophagus may then occur. Lastly, the diminished cough reflex in prolonged esophageal stasis may result in silent aspiration of liquids [10].

The radiologic signs and clinical symptoms in these patients may vary and are often unspecific [2]. On chest radiography, the dilated esophagus presents as a widened mediastinum, often with a characteristic mottled appearance or air-fluid level on the posterior-anterior view, which

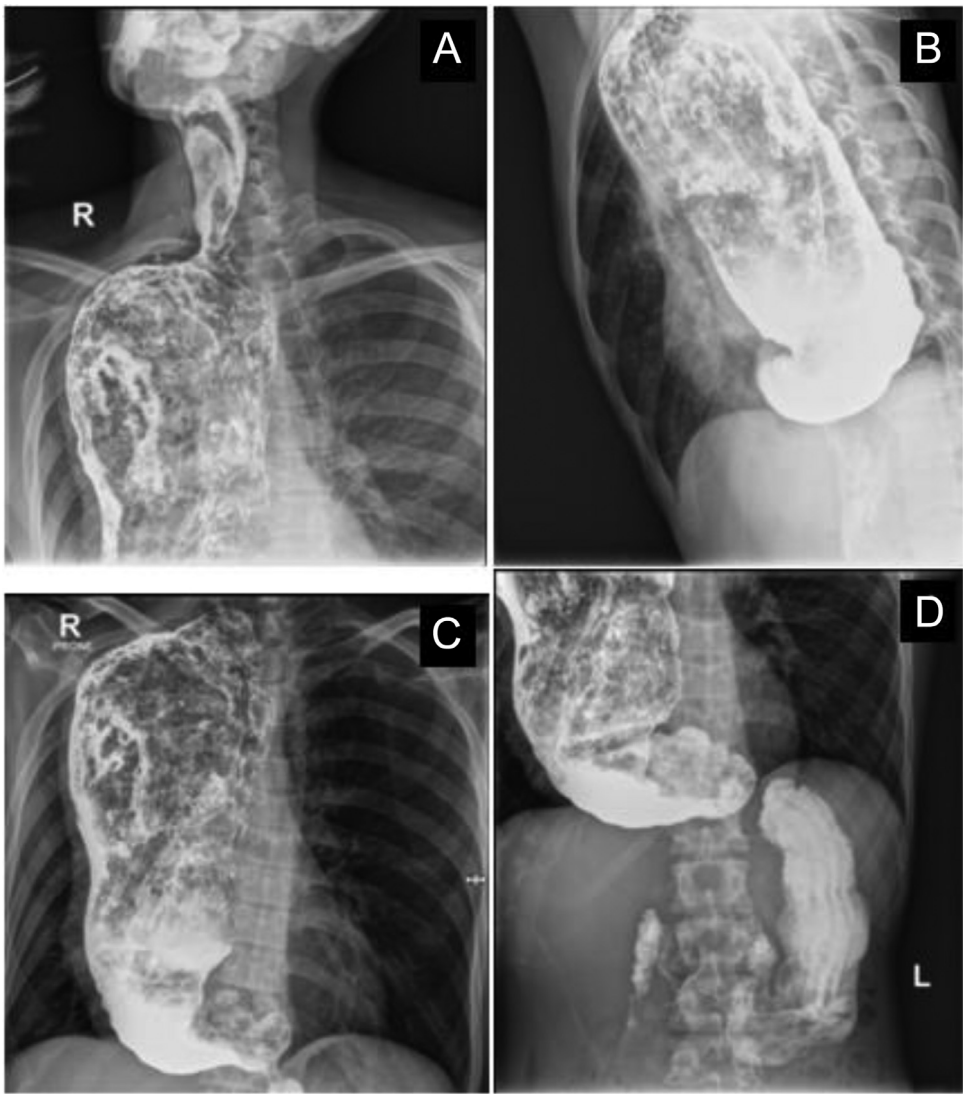

Fig. 2 – Barium sulfate contrast examination shows a large elongated esophagus filled with food residue extending to the thoracic inlet and then barium sulfate contrast enters the stomach slowly.

compresses and displaces the trachea forward on the lateral view [9]. In our study, chest radiography showed a widened mediastinum with a massively dilated esophagus filled with food residue extending up to the neck. Both lung fields were clear. CT scan showed a massive enlargement of the esophagus with the greatest diameter of about 11,3 cm, compressing the posterior tracheal wall and resulting in tracheal narrowing. Mega-esophagus is generally considered when the transverse width of the esophagus is more than 7 cm [10].

Prompt recognition of this rare manifestation of achalasia is critical, and emergency treatment is necessary for this condition. Air and saliva must be evacuated to decompress the esophagus [9,10]. A nasogastric tube may be inserted into the proximal esophagus to reduce the volume of retained food material and thereby reduce airway compression [10–13]. Nasogastric tube insertion is simple and is a relatively low-risk life-saving intervention. Another viable intervention is esophageal decompression with rigid esophagoscopy, but this may not always be readily available in such a situation [11,12].

After stabilization, further therapy is rendered. As the underlying etiology of achalasia remains unclear, current treatments are hence palliative [3]. Management of achalasia involves improving the esophageal outflow in order to provide symptomatic relief to patients. The treatment modalities included pharmacologic therapy (calcium channel blocker, anticholinergic, and nitrates), endoscopic intervention (endoscopic botulinum toxin injection, per oral endoscopic myotomy, pneumatic dilation), and surgical interventions (laparoscopic Heller myotomy with partial fundoplication) [3,6,7]. Laparoscopic Heller myotomy is performed when drug treatment or endoscopic intervention fails to relieve symptoms [7]. The safest and most effective treatment for achalasia is still surgery, which involves cutting the muscle fibers of the abnormally functioning lower esophageal sphincter (a Heller cardiomyotomy). To reduce the risk of gastroesophageal

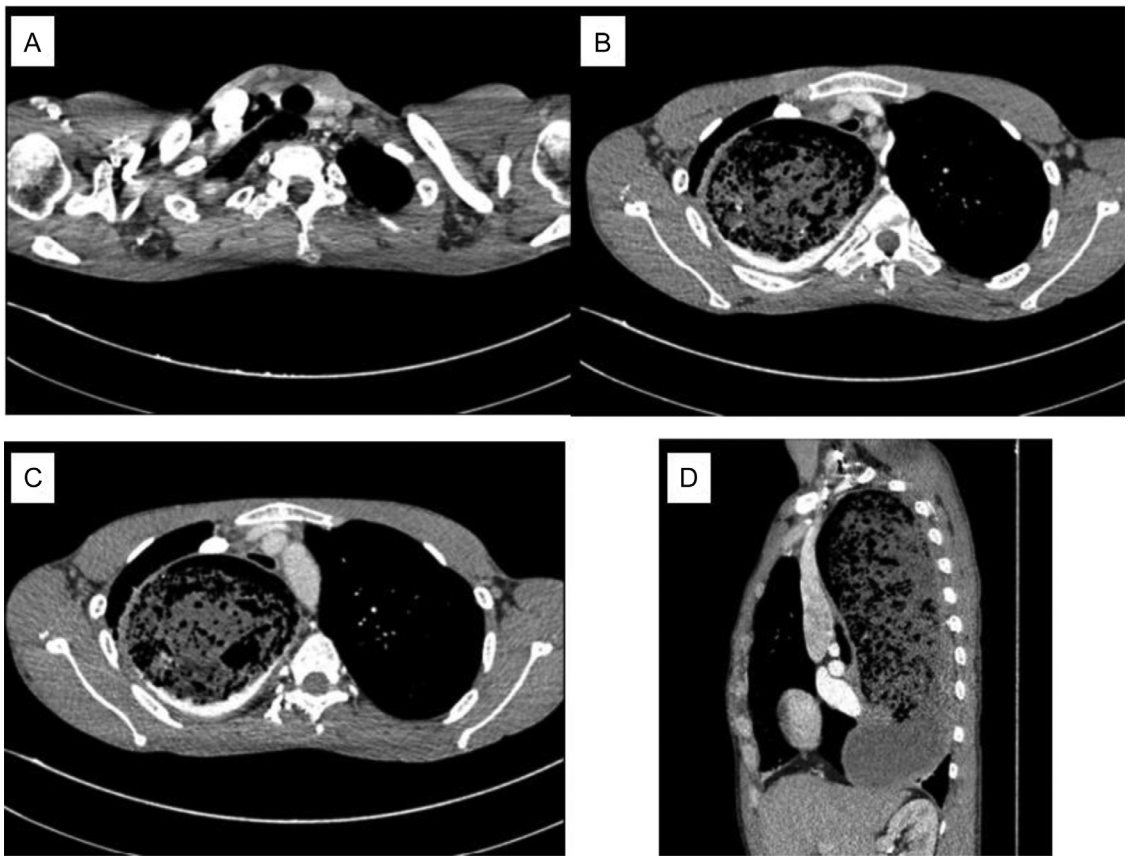

Fig. 3 – CT scan with contrast mediastinal window. (A) Axial CT image just below the level of the cricopharyngeus muscle shows dilation of the upper esophagus without evidence of tracheal compression. (B) and (C) Axial CT images show massive dilation of the esophagus with tracheal compression at the level of the thoracic inlet. (D) The sagittal CT image shows massive dilation of the whole esophagus. The examination was taken during deep inspiration.

reflux disease (GERD) following a cardiomyotomy, which had previously been performed on its own, the procedure was later completed with partial fundoplication (anterior – Dor, or posterior – Toupet). This modified laparoscopic Heller cardiomyotomy completed with semifundoplication has proved to be the most effective procedure, with minimal morbidity, both in the short and long term [14].

Conclusion

Achalasia is one of the most studied esophageal motility disorders. Most patients with achalasia present with slowly progressing dysphagia, regurgitation, chest pain, and weight loss. Respiratory symptoms can result from upper airway obstruction resulting from tracheal compression by dilated esophagus. Though this rarely occurs in achalasia, any delay in diagnosing this exceptional condition is critical. Decompression using a nasogastric tube can be performed as initial therapy. Definitive therapeutic options include a variety of pharmacologic, endoscopic, and surgical techniques, depending on appropriate patient-based considerations.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

REFERENCES

[1] Riccio F, Costantini M, Salvador R. Esophageal achalasia: diagnostic evaluation. World J Surg 2022;46(7):1516–21.
[2] Kaths JM, Foltys DB, Scheuermann U, Strempel M, Niebisch S, Ebert M, et al. Achalasia with megaesophagus and tracheal compression in a young patient: a case report. Int J Surg Case Rep 2015;14:16–18.
[3] Arora Z, Thota PN, Sanaka MR. Achalasia: current therapeutic options. Ther Adv Vaccines 2017;8(6–7):101–8.
[4] Mari A, Baker FA, Pellicano R, Khoury T. Diagnosis and management of achalasia: updates of the last two years. J Clin Med 2021;10(16):3607.
[5] Zhang B, Wang Y, Liao Y, Zhang J, Wu Y, Xiao T, et al. Advances in the diagnosis and treatment of achalasia of the cardia: a review. J Transl Intern Med 2021;9(1):24–31.

[6] Patel DA, Lappas BM, Vaezi MF. An overview of achalasia and its subtypes. Gastroenterol Hepatol 2017;13(7):411–21.
[7] Li MY, Wang QH, Chen RP, Su XF, Wang DY. Pathogenesis, clinical manifestations, diagnosis, and treatment progress of achalasia of cardia. World J Clin Cases 2023;11(8):1741–52.
[8] Swanström LL. Achalasia: treatment, current status and future advances. Korean J Intern Med 2019;34(6):1173–80.
[9] Miyamoto S, Konda Y, Matsui M, Sawada K, Ikeda K, Watanabe N, et al. Acute airway obstruction in a patient with achalasia. Intern Med 2011;50(20):2333–6.
[10] Khan AA, Shah SWH, Alam A, Butt AK, Shafqat F, Malik K, et al. Achalasia esophagus; presenting as acute airway obstruction. J Pak Med Assoc 2007;57(8):423–5.
[11] Arcos E, Medina C, Mearin F, Larish J, Guarner L, Malagelada JR. Achalasia presenting as acute airway obstruction. Dig Dis Sci 2000;45(10):2079–84.
[12] Basavarajaiah S, Barnes DJ, Boyle N. Simple treatment of stridor caused by achalasia of the cardia. JRSM Short Rep 2011;2(4):29.
[13] Adamson R, Lee YI, Berger KI, Sutin K, Nolan A. Acute respiratory failure secondary to achalasia. Ann Am Thorac Soc 2013;10(3):268–71.
[14] Andrási L, Paszt A, Simonka Z, Ábrahám S, Erdős M, Rosztóczy A, et al. Surgical treatment of esophageal achalasia in the era of minimally invasive surgery. J Soc Laparoendosc Surg 2021;25(1) e2020.00099.