

Left Atrial Compression Secondary to Massive Esophageal Dilatation in a Patient With Idiopathic Achalasia



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INTRODUCTION

Idiopathic achalasia (IA) is a primary esophageal motility disorder characterized by aperistalsis and lower esophageal sphincter (LES) dysfunction. It is a rare disease and usually manifests clinically with progressive dysphagia and weight loss. Left atrial (LA) compression by achalasia is an exceptionally rare presentation and may cause hemodynamic compromise.

Here we present a case of LA compression by a massive esophageal dilatation caused by IA, diagnosed on transthoracic echocardiography (TTE), further characterized by performing a carbonated beverage swallow test and complemented by thoracic computed tomography. LA compression is a rare presentation of IA, an uncommon cause of extrinsic cardiac compression that should be kept in mind in a patient with symptoms of progressive dysphagia associated with dyspnea and other signs and symptoms of heart failure.

CASE PRESENTATION

A 22-year-old male patient presented to a primary health care setting with a 2-month history of progressive dysphagia (including liquids), regurgitation, progressive dyspnea, cough, and weight loss (18 kg). There was worsening of dyspnea and cough in the postprandial period, especially when assuming the supine position, with regurgitation of undigested food. He was previously healthy, with no family history of cardiovascular diseases or any previous gastrointestinal disorders, with normal physical activity capacity. Physical examination revealed regular pulse, clear lungs, and normophonetic heart sounds, without appreciable heart murmurs. Of note, the patient was emaciated, was tachypneic at rest, and presented with dyspnea and cough when assuming a supine position for the physical examination. Electrocardiography showed sinus rhythm, with no repolarization abnormalities. The patient was referred for TTE, revealing LA compression

by a mass in the posterior mediastinum, with well-defined borders and heterogeneous hyperechogenic core (Figure 1), in close relation to the pulmonary veins (Figure 2), and with slightly mobile contents (Videos 1 and 2) suggesting a gastrointestinal source. In the long-axis parasternal view, we could see the cylindrical and elongated shape of the structure. Trying to elucidate the gastrointestinal source of the mass, we performed a carbonated beverage swallow test, with the oral administration of 150 mL of a regular soda, to visualize the mobilization of contents caused by the mixture of liquid and gas bubbles. In this case it became very clear, with swirling echodensities well shown inside the mass, and also by an increase in its dimensions with more dilatation, causing progressive accentuation of the hemodynamic compromise, leading to almost complete collapse of the LA cavity (Figure 3, Videos 3–6). The patient evolved with intense dyspnea, hypotension (70/40 mm Hg), and sudoresis, relieved after vomiting and assuming the orthostatic position. Noncontrast thoracic computed tomography confirmed the aforementioned findings, demonstrating a huge dilated esophagus (6.0 cm), extending from the oropharynx to the gastroesophageal junction, with alimentary remnants (Figures 4 and 5), and provided more detailed anatomic information about the spatial relation of the dilated esophagus to adjacent cardiac structures, also ruling out the possibility of pseudoachalasia, as there was no extrinsic compression of the gastroesophageal junction by surrounding structures. The patient was admitted to a general hospital for esophageal decompression and other supportive measures (intravenous fluid replacement, electrolyte correction, and enteral nutrition). Next, he was referred to a tertiary hospital, where high-resolution esophageal manometry (HREM) was performed, showing deficit of relaxation of the LES and aperistalsis, typical pattern of type I achalasia. Figure 6 depicts the HREM results with a parametric display of pressure dynamics inside the esophagus, measured at different levels away from the oropharynx. The absence of peristalsis after swallowing is very clear, without significant pressurization of the esophageal body, and the inadequate relaxation of the LES, demonstrated by a high integrated relaxation pressure (32 mm Hg) at the esophagogastric junction maintained throughout the analysis. The patient underwent LES pneumatic endoscopic dilatation with a 30-mm balloon 1 week after the initial diagnosis with satisfactory manometric and clinical results. After this first pneumatic dilatation session, the patient underwent iodinated contrast esophagography that showed the typical aspect of “bird’s beak,” with a smooth tapering of the distal esophagus with proximal dilatation of the proximal portions (Figure 7). He also underwent thoracic computed tomographic angiography with three-dimensional reconstruction, revealing more details of the dilated esophagus and its spatial relation to mediastinal structures. At this time, computed tomography showed only mild compression of the pulmonary veins and the left atrium, to a much lesser extent compared with admission

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Keywords: Left atrial compression, Echocardiography, Idiopathic achalasia, Carbonated beverage swallow test

Conflicts of interest: The authors reported no actual or potential conflicts of interest relative to this document.

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2468-6441

<https://doi.org/10.1016/j.case.2018.07.002>

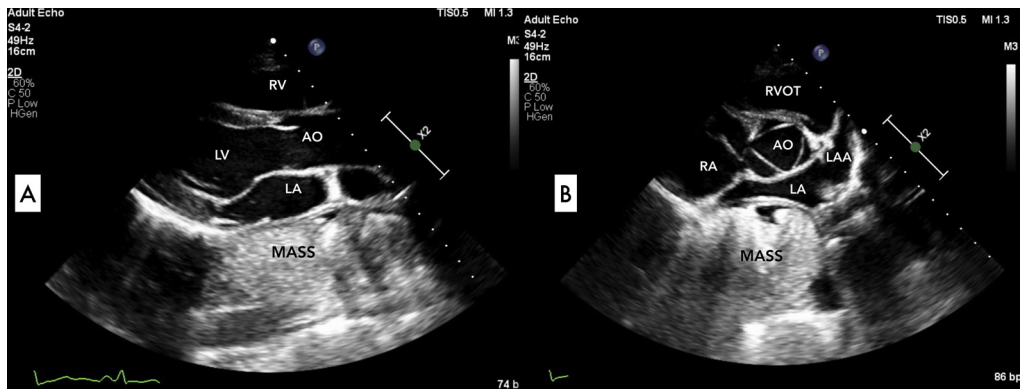


Figure 1 Transthoracic bidimensional echocardiogram. **(A)** Long-axis parasternal view showing extrinsic compression of the left atrium (LA) by a mediastinal posterior mass. **(B)** Short-axis parasternal view clearly depicting LA compression by a mass with well-defined borders and heterogeneous hyperechogenic core, with slightly mobile contents. AO, Aorta; LAA, left atrial appendage; LV, left ventricle; RA, right atrium; RV, right ventricle; RVOT, right ventricular outflow tract.

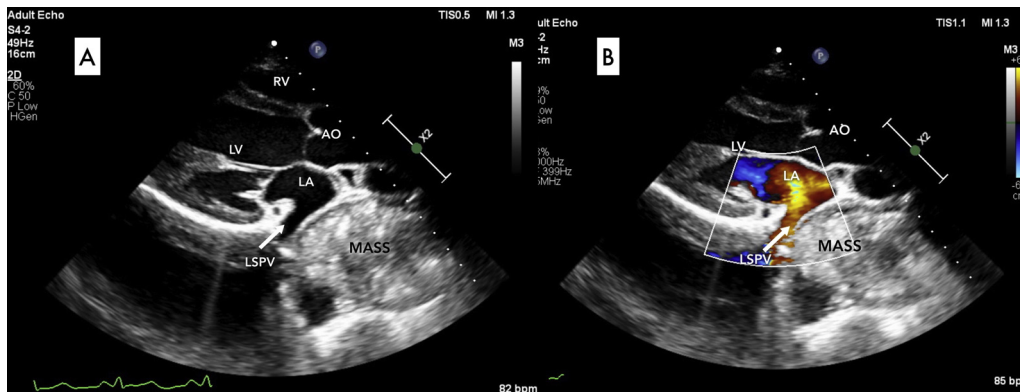


Figure 2 Transthoracic bidimensional echocardiogram. **(A)** Long-axis parasternal view showing extrinsic compression of the left atrium (LA) by a mediastinal posterior mass with intimate relation with the left superior pulmonary vein (LSPV). **(B)** Long-axis parasternal view with color Doppler showing laminar flow inside the LSPV. AO, Aorta; LAA, left atrial appendage; LV, left ventricle; RA, right atrium; RV, right ventricle.

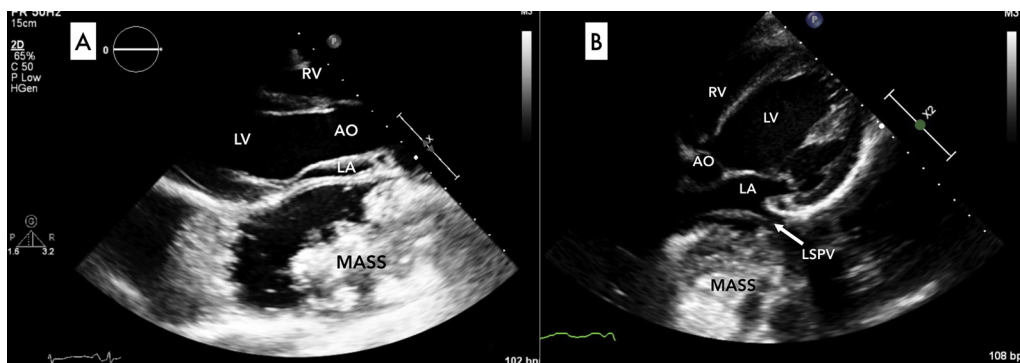


Figure 3 Transthoracic bidimensional echocardiogram after carbonated beverage swallowing test. In these images, we can see not only the mobilization of the heterogeneous content of the mass (confirming its gastrointestinal source) but also the progressive worsening of the hemodynamic compromise due to almost complete collapse of the left atrial (LA) cavity. **(A)** Long-axis parasternal view depicting severe esophageal dilatation after carbonated beverage ingestion (hypoechoic component inside the mass), causing extrinsic compression and almost collapse of the LA. **(B)** Modified apical view showing lateroposterior compression of the LA by the dilated esophagus, also compressing the left superior pulmonary vein (LSPV). AO, Aorta; LV, left ventricle; RA, right atrium; RV, right ventricle.

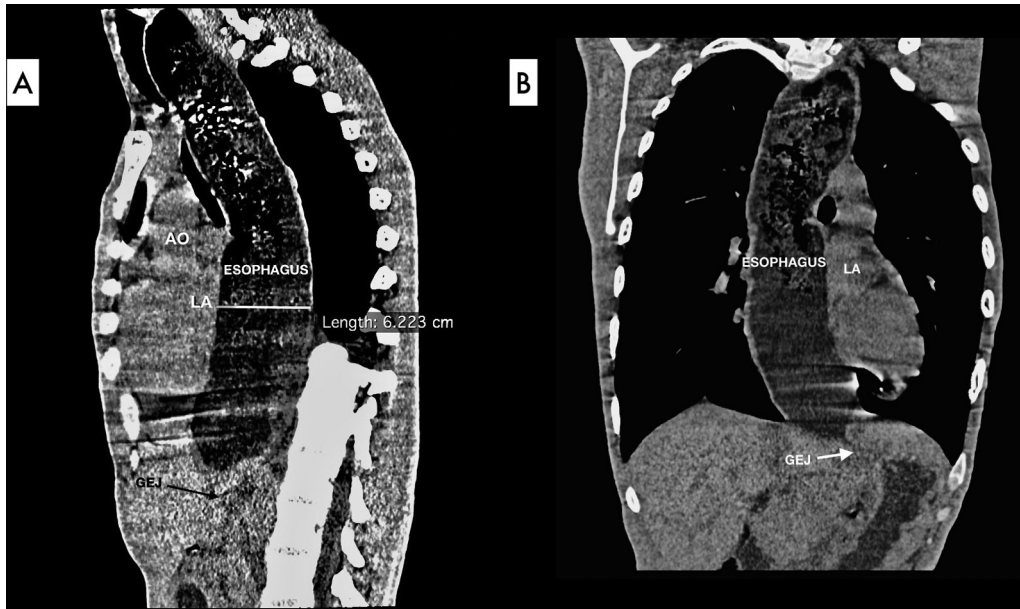


Figure 4 Thoracic computed tomography. Multiplanar reconstructed images. **(A)** Sagittal view. A massively dilated esophagus filled with alimentary remnants extending from the gastroesophageal junction to the oropharynx is seen, causing extrinsic compression to the left atrium (LA). **(B)** Oblique view. We can appreciate not only the dilatation of the esophagus (achalasia), but also the intimate relation and compression of the LA as well as the details of the narrowed gastroesophageal junction (GEJ; arrow). AO, Aorta.

examinations, with clear improvement (Figures 8–10). Serologic tests for *Trypanosoma cruzi* and for other immunologic disorders were performed, the results of which were all negative. This led us to the diagnosis of IA. One month after LES dilatation, the patient had regained 15 kg, with a dramatic improvement in hemodynamic alterations and resolution of symptoms of heart failure. The patient remained asymptomatic, with no further dyspnea or dysphagia and no signs of LA compression on TTE after 6-month follow-up (until the present time; Figures 11 and 12, Videos 7 and 8). The size of the esophagus remained stable at 4.9 cm (Figure 11A).

DISCUSSION

Extrinsic compression of the left atrium is a very rare presentation of IA and may eventually cause hemodynamic compromise. IA is a primary esophageal motor disorder of unknown etiology characterized by insufficient LES relaxation and loss of esophageal peristalsis, causing esophageal dilatation and usually expressing clinically as dysphagia to solids and liquids, regurgitation, and weight loss.¹ This pattern of esophageal motor disorder is well demonstrated by HREM and classified as type I achalasia.² IA rarely presents with symptoms of compression of adjacent mediastinal structures by the dilated esophagus, such as bronchial³ or cardiac^{4–10} compression. LA extrinsic compression may lead to impaired ventricular filling, elevation of LA and pulmonary venous pressures, clinically manifesting as acute heart failure with signs of pulmonary congestion and low cardiac output.^{10,11} The differential diagnosis comprises a wide class of diseases that may cause compression of mediastinal structures, such as gastrointestinal disorders (hiatal hernia, esophageal hematoma), mediastinal masses (thymoma, lymphoma, soft tissue or germ cell tumors), aorta and intrapericardial

structures (thoracic aortic aneurysms), and pulmonary structures (lung tumors, bronchogenic cysts).¹² The first report of the use of TTE for the assessment of hiatal hernia as a possible etiology of incidentally found retrocardiac masses was made in 1985 by Nishimura *et al.*,¹³ also describing the use of the oral carbonated beverage test for a better evaluation of gastroesophageal pathology, as we did in this case. This technique relies on demonstrating the echocardiographic appearance of air contrast in the esophagus during the ingestion of liquids containing carbon dioxide, revealing the dilatation of its lumen. This then can help differentiate from other mediastinal structures and esophageal tumors. Hiatal hernia is the most prevalent echocardiographic finding of a mass of gastrointestinal origin posterior to the left atrium and is the most common cause of extrinsic LA compression reported in literature.¹⁴ However, esophageal dilatation must be considered as a potential cause, especially in regions with a high incidence of Chagas disease, such as South America and particularly Brazil. Unlike IA, Chagas disease is a secondary cause of esophageal dilatation (caused by *Trypanosoma cruzi* infection) that produces an achalasia-like syndrome, with different pathologic process and HREM pattern (hypotonic LES). After a MEDLINE search on PubMed, we found only nine cases of extrinsic LA compression caused by IA, being a rare presentation of the disease. As with our case, only four of the cited cases were reported to cause a significant hemodynamic repercussion.^{6,7,9,10} Because there is no cure for achalasia, treatment is aimed at relieving symptoms by improvement of LES physiology and reduction of functional obstruction through pharmacologic, endoscopic, and surgical interventions, with the ultimate goal of improving esophageal emptying. In general, the most long-term effective treatments are pneumatic dilatation and surgical myotomy. Pneumatic dilatation is a very effective nonsurgical treatment for achalasia, and almost 50% of

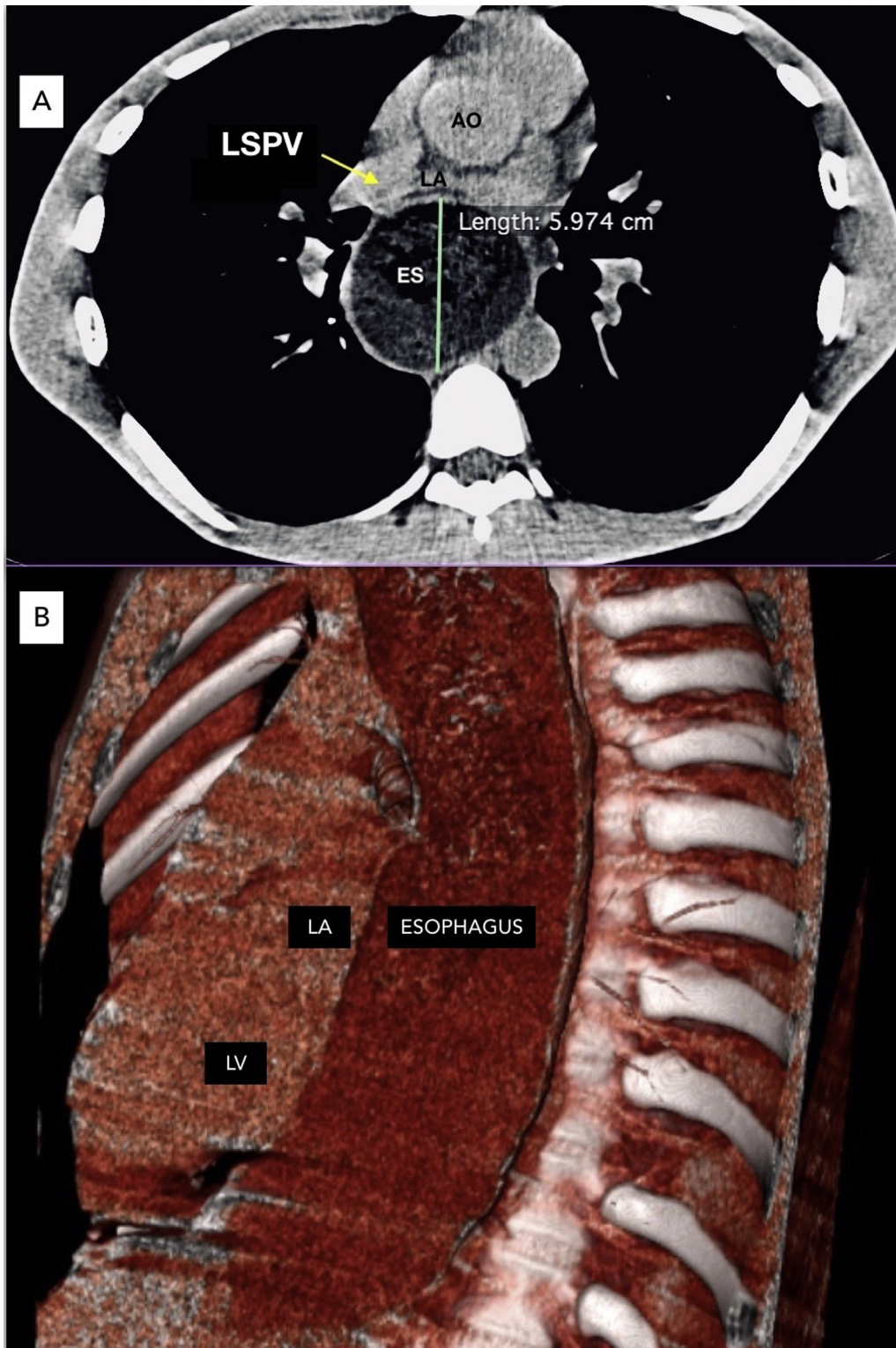


Figure 5 Noncontrasted thoracic computed tomography. **(A)** Axial view. A massively dilated esophagus (6.0cm) filled with alimentary remnants causing extrinsic compression of the left atrium (LA) and pulmonary veins. **(B)** Three-dimensional rendered images from sagittal view. There is extreme dilatation of the esophagus (achalasia), causing extrinsic compression of the LA. AO, Aorta; ES, esophagus; LSPV, left superior pulmonary vein; LV, left ventricle.

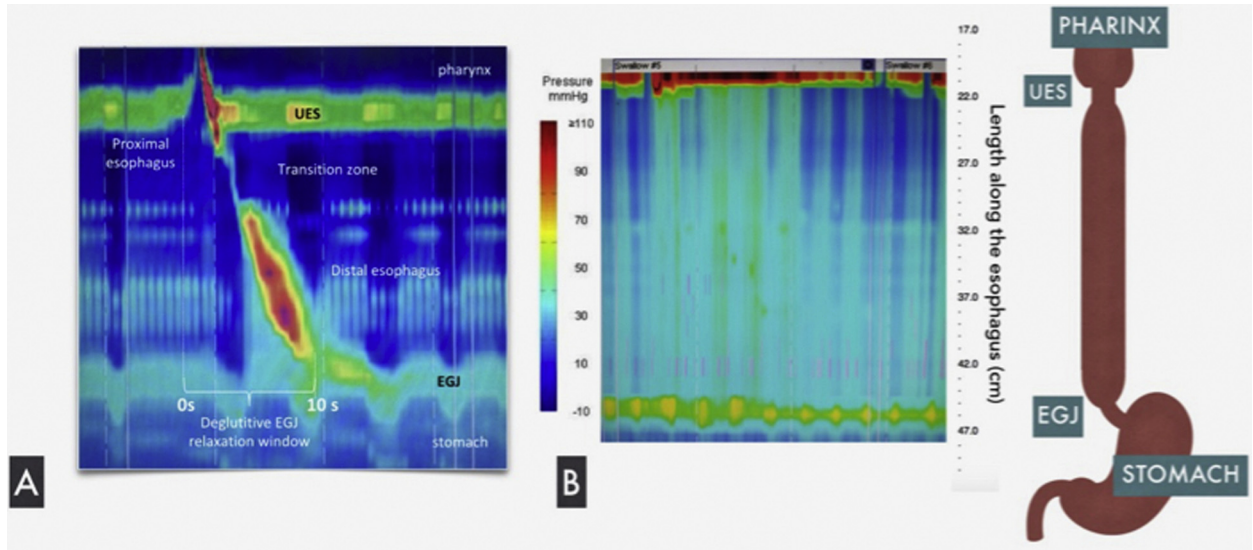


Figure 6 High-resolution esophageal manometry (HREM) results displayed in a color-coded parametric display of pressure dynamics inside the esophagus (Clouse plot), with measurements at different levels away from the oropharynx registered in time after swallowing. **(A)** Example of a normal physiologic pattern of sequential pressurization of the esophageal body after swallowing with normal relaxation of the lower esophageal sphincter (LES) with low pressures at the esophagogastric junction (EGJ). (Courtesy of V.F.) **(B)** HREM of our patient where we can see clearly the absence of peristalsis after swallowing (without significant pressurization along the esophagus body), and the inadequate relaxation of the LES, demonstrated by a high integrated relaxation pressure (32 mm Hg) maintained throughout the analysis at the EGJ, a typical pattern of type I achalasia. UES, Upper esophageal sphincter.

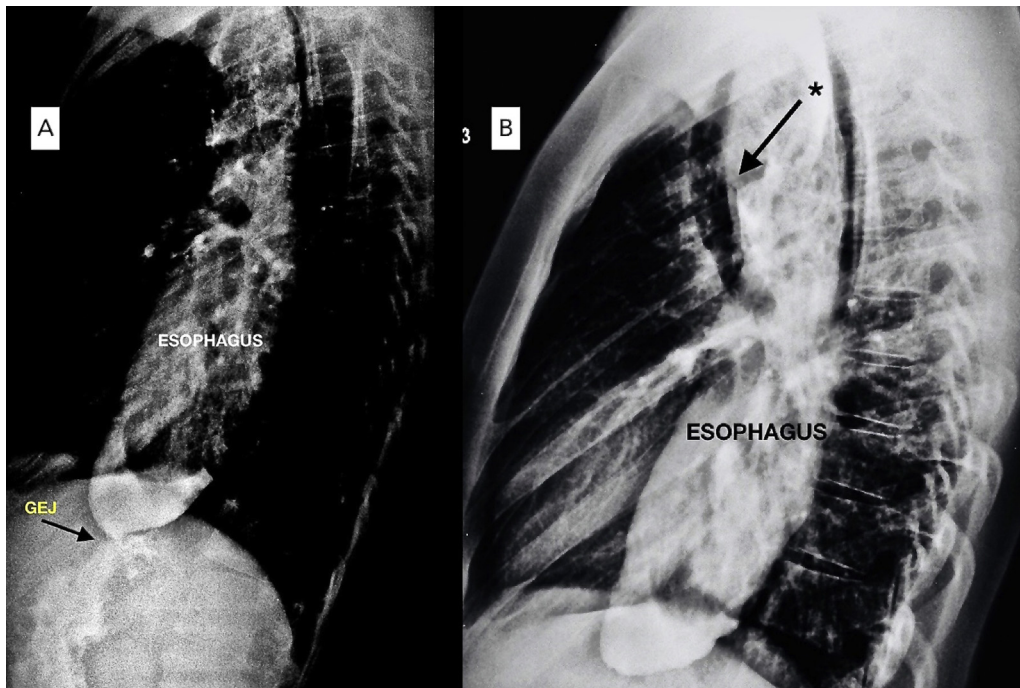


Figure 7 Iodinated contrast esophagography. **(A)** Lateral view showing gastroesophageal junction (GEJ) constriction and dilatation of the esophagus (achalasia). **(B)** Lateral view showing dilatation of the esophagus and anterior displacement of the trachea (black arrow and *).

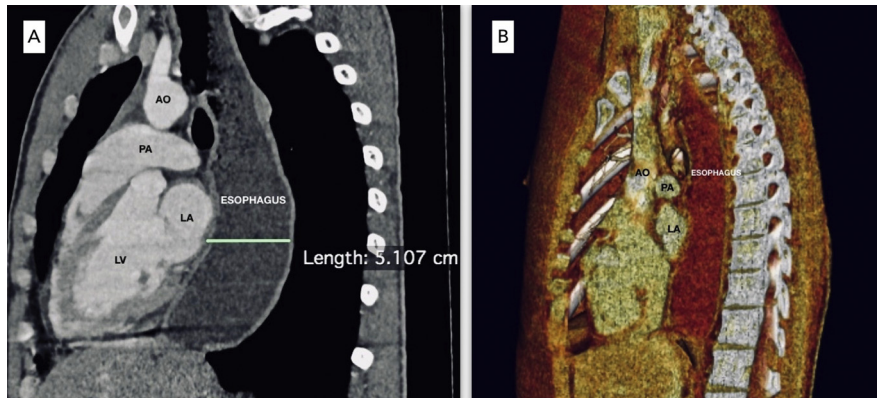


Figure 8 Thoracic computed tomography (CT) angiography after the first session of pneumatic dilatation of the esophagus. **(A)** Sagittal view demonstrating esophagus dilatation, filled with alimentary remnants, less pronounced than the initial exam at presentation (5.1cm) causing only slight extrinsic compression to the left atrium (LA). **(B)** Surface-rendered CT, from a lateral perspective, showing dilatation of the esophagus and slight extrinsic compression to the LA. AO, Aorta; LV, left ventricle; PA, pulmonary artery.

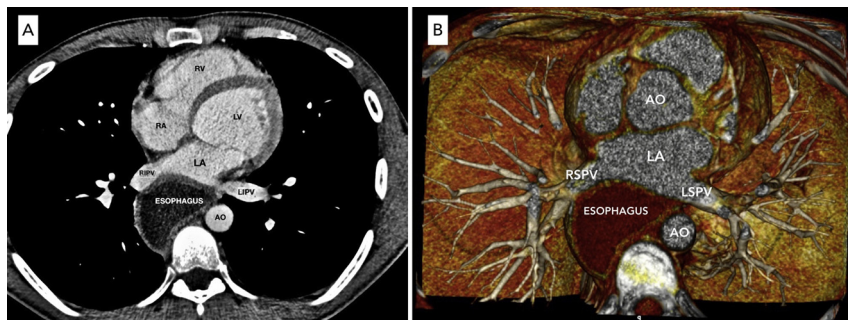


Figure 9 Thoracic computed tomography (CT) angiography after the first session of pneumatic dilatation of the esophagus. **(A)** Axial view demonstrating esophagus dilatation, filled with alimentary remnants, causing only slight extrinsic compression to the left atrium (LA) and left inferior pulmonary vein (LIPV). **(B)** Surface-rendered CT, axial view, showing dilatation of the esophagus and slight extrinsic compression to the LA and left superior pulmonary vein (LSPV). AO, Aorta; LV, left ventricle; RA, right atrium; RIPV, right inferior pulmonary vein; RV, right ventricle; RSPV, right superior pulmonary vein.

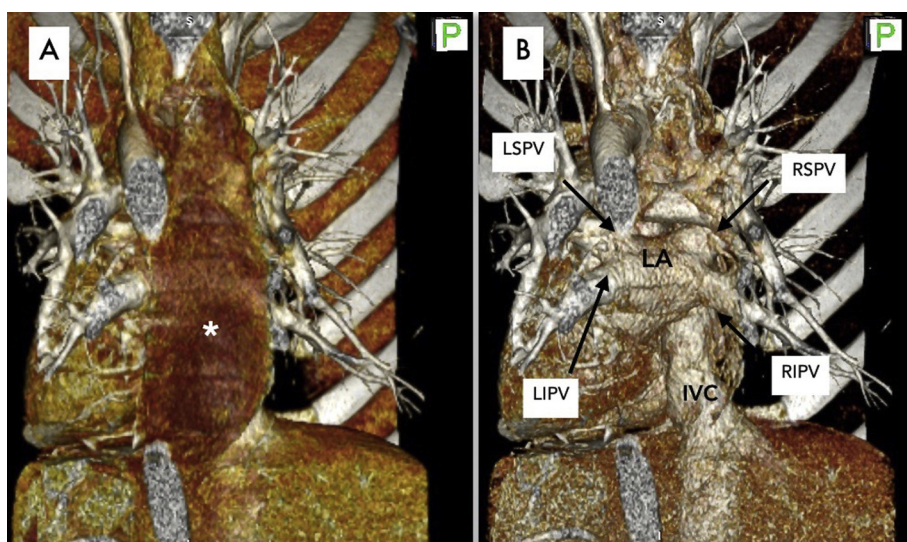


Figure 10 Thoracic computed tomography angiography after the first session of pneumatic dilatation of the esophagus. Surface-rendered images from a posterior oblique view **(A)** showing the intimate relation of the dilated esophagus (*) to the posterior aspect of the left atrium (LA) and pulmonary veins. **(B)** Suppressing the esophagus, we can see the anatomy of the posterior wall of the LA and the anatomic position and orientation of the pulmonary veins. IVC, Inferior vena cava; LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein.

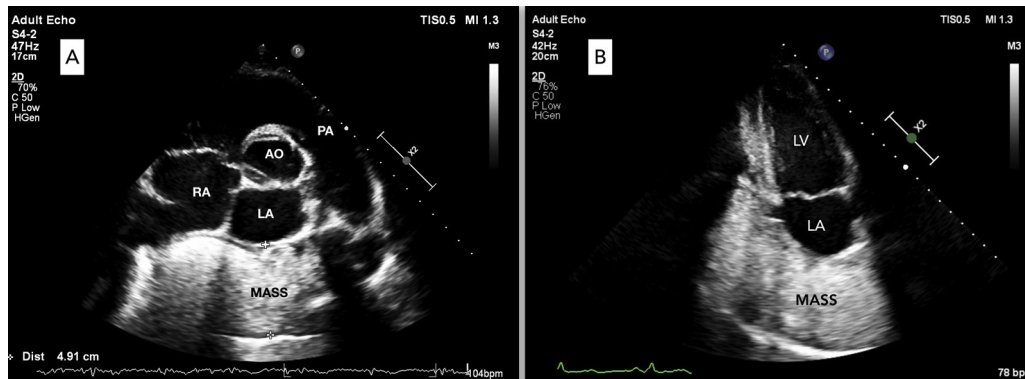


Figure 11 Transthoracic two-dimensional echocardiogram after two sessions of esophageal pneumatic dilatation. **(A)** Basal parasternal short axis. The left atrium (LA) has recovered its round geometry and there has been a clear reduction in the esophagus caliber, measured as 4.9cm. **(B)** Two-chamber apical view. We can see the dilated esophagus elongated adjacent to the LA but without signs of significant extrinsic compression. AO, Aorta; LV, left ventricle; PA, pulmonary artery; RA, right atrium.

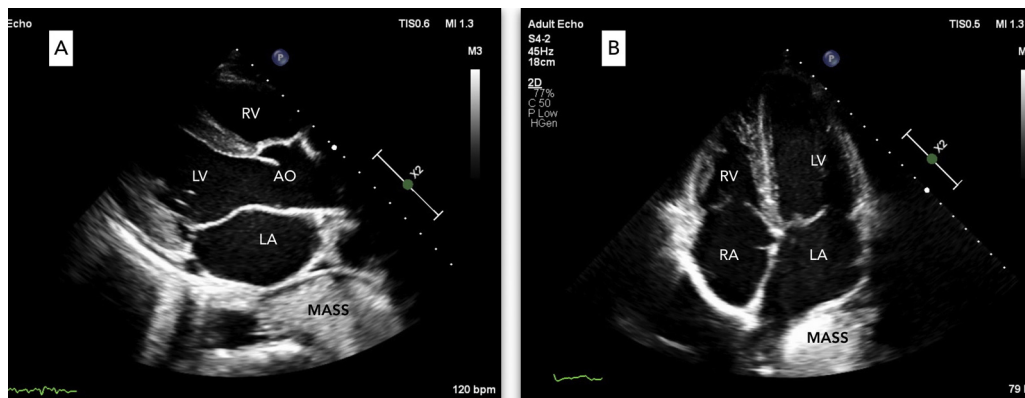


Figure 12 Transthoracic two-dimensional echocardiogram after two sessions of esophageal pneumatic dilatation. **(A)** Long-axis parasternal view. The left atrium (LA) has recovered its round geometry, despite there still being a dilated esophagus. **(B)** Four-chamber apical view. We can no longer identify extrinsic compression of the LA by the dilated esophagus. AO, Aorta; LV, left ventricle; RA, right atrium; RV, right ventricle.

patients can obtain clinical improvement and remission for 5 to 10 years,¹⁵ as we observed in our patient in his first 6 months of follow-up.

CONCLUSION

LA extrinsic compression by a massively dilated esophagus is a very rare clinical entity and may be associated with IA. It is important to consider this etiologic possibility in young patients with histories of progressive dysphagia and associated dyspnea. This case report reinforces the value of TTE using the carbonated beverage swallow test as an important tool to disclose gastrointestinal source when investigating retrocardiac masses.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2018.07.002>.

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