

A Rare Cause of Left Atrium Compression: Esophageal Achalasia

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Case report

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Abstract

Background

Extrinsic compression of left atrium (LA) due to esophageal achalasia is uncommon. Patients might present with dysphagia, dyspnea, and even hemodynamic compromise. Prompt detection with thorough differential diagnosis is crucial for subsequent management. We presented a case with LA compression by esophageal achalasia, and literature review regarding the clinical manifestation, diagnosis, and treatment strategy was performed to provide an updating knowledge of the disease.

Case presentation

A 59-year-old relatively healthy man presented with dysphagia accompanied by chest tightness and breathlessness after a large meal. His chest X-ray film disclosed a widened mediastinum. The barium swallow esophagogram revealed contrast pooling at the esophagogastric junction with a bird beak shape. Meanwhile, the transthoracic echocardiogram showed a round-shaped, well bordered, hyperechogenic, and heterogeneous mass (5.1 cm x 3.8 cm in size) compressing the LA irrespective of the systolic or diastolic phase. A chest contrast-enhanced computed tomography scan showed diffuse esophageal dilatation with a smoothly thickening wall aligned compressing the LA. Due to the abovementioned image findings, extrinsic compression of LA by esophageal achalasia was diagnosed.

Conclusion

LA compression due to esophageal achalasia is not common. Remarkably, given a patient presenting dysphagia and concurrent dyspnea after meals, the clinicians should keep this differential diagnosis in mind. Echocardiography and esophagography are useful to ensure the diagnosis promptly.

Introduction

Extrinsic compression of the left atrium (LA) is an unusual condition.¹ The most common etiology involves compression by an abnormal gastrointestinal structure such as hiatal hernia.¹ Other possible causes include pathological changes in the mediastinum, lungs, pericardium, and aorta.¹ LA compression reduces the LA volume, which can result in low cardiac output, high pulmonary venous pressure, pulmonary edema, and hence, unstable hemodynamics.² Depending on the severity of the compression, patients might experience tachycardia, palpitation, dyspnea, chest pain, exercise intolerance, and compromised hemodynamic function.³

This case report demonstrated a patient with LA compression due to esophageal achalasia. The diagnosis was confirmed by transthoracic echocardiography, a barium swallow esophagogram, and chest computed tomography. Besides, we reviewed the clinical manifestations, diagnostic tools, and subsequent management of the reported cases in the literature.

Case Report

A 59-year-old healthy male with intermittent palpitation, heartburn sensation and difficulty in swallowing for 20 years, presented at the emergency department due to acute onset of chest compression and breathlessness after a large meal. Exception for high blood pressure (213/139 mm Hg), his vital signs (heart rate 94 beats per minute, breath rates 28 times per minute) were acceptable and physical examination revealed no abnormal findings. An electrocardiogram showed a non-specific change in ST-T segment, but a chest X-ray showed a widened mediastinum. His laboratory tests showed no abnormality.

With stable vital signs and relief of the symptoms, the patient was referred to the outpatient department for a follow-up examination. Esophagogastroduodenoscopy (EGD) showed a dilated lower esophageal lumen with fluid accumulation and a tight gastroesophageal junction. The barium swallow esophagogram, revealed a bird beak shape at the esophagogastric junction with contrast pooling, which was diagnosed as esophageal achalasia (Fig. 1).

Because of the concurrent dyspnea, a transthoracic echocardiogram (TTE) was used to assess the heart function, which showed a normal-sized left ventricle with an adequate ejection fraction but abnormal in relaxation. A round-shaped, well bordered, hyperechogenic, and heterogeneous mass (5.1 cm x 3.8 cm in size) was found compressing the LA irrespective of the systolic or diastolic phase (Fig. 2). A color Doppler TTE showed the absence of blood flow over the mass. The characteristics, location, and origin of the mass pointed to the presence of extracardiac mass or tumor. Therefore, a chest contrast-enhanced computed tomography scan was arranged, and it showed diffuse esophageal dilatation with a smoothly thickening wall that was aligned with and compressing the LA (Fig. 3). According to the abovementioned image findings, the LA compressed by esophageal achalasia was diagnosed. The patient was then referred to chest surgeon for the evaluation of surgical intervention for achalasia.

Discussion

LA compression due to esophageal achalasia is uncommon. Apart from achalasia, four categories of abnormal structure could compress LA, which are defined by the origin and location: gastrointestinal structures, mediastinal structures, aorta and intracardial or pericardial structures, and pulmonary structures.¹

LA compression can lead to low cardiac output, dyspnea, chest pain, pulmonary congestion, palpitation, and hemodynamic instability.¹ Some patients with LA compression may develop heart failure.^{4,5} Comparing to congestive heart failure with LA enlargement, heart failure due to LA compression results from reduces LA volume and increases the refilling pressure. Accordingly, patients may also experience repeated syncope and arrhythmia.^{6,7} Upile et al. presented a case of achalasia with paroxysmal atrial fibrillation, whose heart rhythm reverted to sinus rhythm when food debris was removed from the esophagus.⁸ The mechanism for arrhythmia is not fully understood, but Volpi et al. proposed that the plausible explanation is mechanical stretching of the atrial myocardial fibers.⁹

The most common symptoms of esophageal achalasia are dysphagia and acid regurgitation.¹⁰ It is an uncommon esophageal motility disorder, with a prevalence of around 11 cases per 100,000 individuals.¹¹ The pathophysiology of achalasia involves the degeneration of inhibitory neurons in the esophageal myenteric plexus but the exact cause remains unknown.¹² Esophageal achalasia is often diagnosed using endoscopy. The finding of a bird beak shape of contrast pooling on the barium swallow esophagogram is typical.¹³ Clinically, esophageal manometry can be used to confirm the diagnosis.¹⁴ Current treatment mainly targeting the disruption of the lower esophageal sphincter, and no intervention had been verified to reverse the degeneration of esophageal neurons. Other treatment options include pneumatic dilatation, peroral endoscopic myotomy, botulinum toxin injection, and laparoscopic Heller myotomy with partial fundoplication. There is little evidence that calcium blockers or phosphodiesterase are effective, but these medications are commonly used in patients who cannot tolerate invasive interventions.¹⁴

TTE is the most common non-invasive diagnostic tool for extrinsic LA compression, and even a slight compression or deformity in the LA chamber can also be detected. Given a poor echocardiographic window, a transesophageal echocardiogram can be an alternative method to detect the anomaly. With the combination of TTE and carbonated beverage swallow tests, it is possible to differentiate gastrointestinal origins of extrinsic LA compression from the other mediastinal structures.¹⁵ Color Doppler echocardiography can also detect the turbulent flow in the LA and blood flow over the extrinsic mass. Contrast echocardiography can distinguish vascular or non-vascular structures that compress the LA.⁵ Further image tools such as computed tomography and magnetic resonance imaging, are usually performed to provide details of the features and origin of the mass.¹ TTE can also be used to determine the effectiveness of treatment afterwards.¹⁶

This case report involves a patient with LA compression due to esophageal achalasia and gives a brief review of diagnoses and treatments of the disease. Extrinsic compression of the LA due to esophageal achalasia is rare, and the condition causes dysphagia that is concomitant with symptoms of heart failure. A prompt and accurate diagnosis is essential. TTE is a rapid, safe, and non-invasive diagnostic tool for LA compression, and a bird beak shape of contrast pooling on the barium swallow esophagogram is indicative of esophageal achalasia. Although the condition is relatively rare, the diagnosis must be considered in patients presenting with simultaneous dyspnea and dysphagia.

Abbreviations

LA: Left atrium

EGD: Esophagogastroduodenoscopy

TTE: Transthoracic echocardiogram

Declarations

Ethics approval and consent to participate

This study was granted exemption from review by the Ethics Committee of Chi Mei Medical Center.

Consent for publication

This study received subject consent to disclose the related information.

Availability of data and materials

Not applicable

Competing interests

All authors have no conflict of interest

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Authors' contributions

SYC drafted the manuscript. WTC had contribution to the conception. CTL critically revised the manuscript. All authors read and approved the final manuscript.

Acknowledgment

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Figures

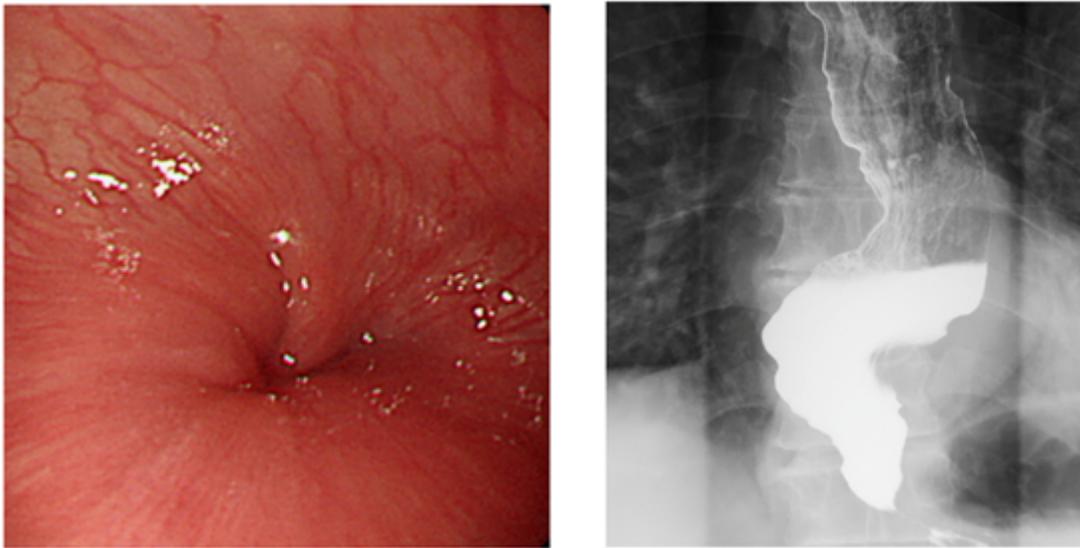


Figure 1

(Left) Esophagogastroduodenoscopy image shows impaired relaxation of the gastroesophageal sphincter without mechanical obstruction. (Right) Barium swallow study shows a bird beak shape of contrast pooling.

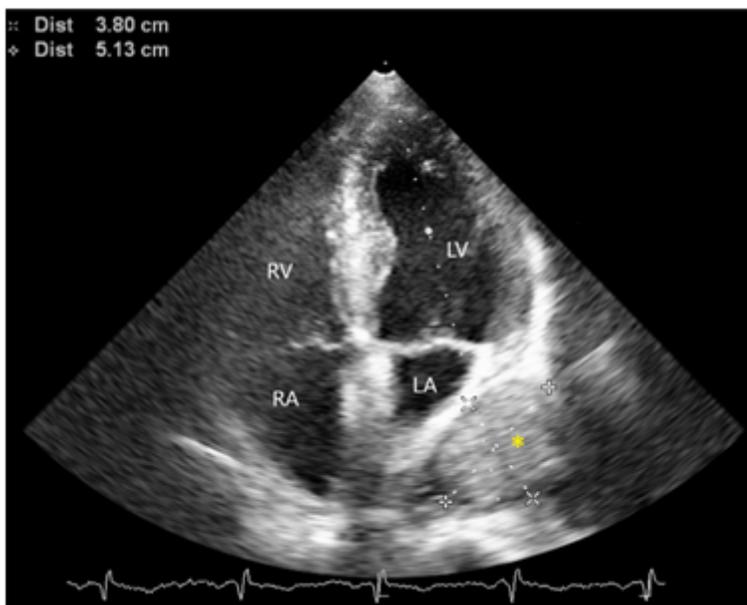


Figure 2

Transthoracic echocardiogram (Apical four-chamber view) image demonstrated a round-shaped, well-bordered, hyperechogenic and heterogenous mass (5.1 cm x 3.8 cm in size) compressing the left atrium in the systolic and diastolic phase. (Asterisk * shows the tumor). RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle.

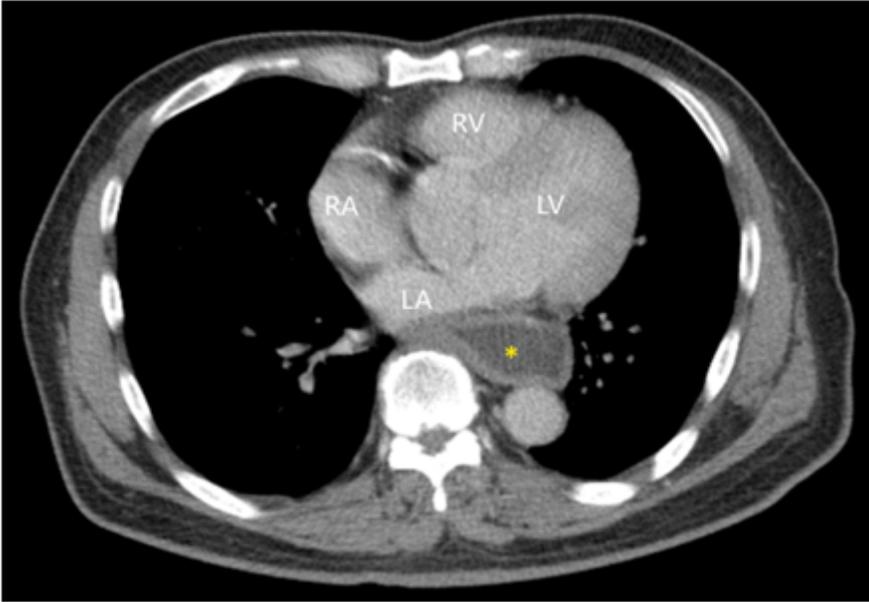


Figure 3

Contrast computed tomography (axial view) image showed that the left atrium was compressed by esophageal achalasia (Asterisk * shows the location of esophageal achalasia). RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle