

A Very Uncommon Cause of Paroxysmal Nocturnal Dyspnea: Achalasia: Case Report

Paroksizmal Nokturnal Dispnenin Son Derece Nadir Bir Nedeni: Akalazyaya

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ABSTRACT Extrinsic compressions of the heart chambers are rare occurrences and may present a variety of clinical presentations depending on the location. A 61-year old man had achalasia clearly compressing left atrium, found by echocardiography and computed tomography. The patient presented with paroxysmal nocturnal dyspnea for 1-month duration. Echocardiography showed an echolucent mass, which was later confirmed to be achalasia by computed tomography, compressing the left atrium. The patient's nocturnal dyspnea was attributed to the compressive effect of achalasia on the left atrium, thereby reducing left atrial volume and increasing pulmonary venous hypertension. It must always be kept in mind that dorsal structures to the left atrium should have paramount attention when evaluating the patients who complain dyspnea.

Key Words: Esophageal achalasia; heart atria; data compression

ÖZET Kalp boşluklarının dış basya maruz kalması son derece nadir rastlanan durumlardandır. Bası yerine bağlı olarak klinik başvuru şekli de son derece farklılık göstermektedir. 61 yaşında erkek hastada ekokardiyografi ve bilgisayarlı tomografi ile tespit edilen sol atriyumuna bası yapan akalazyaya tespit edildi. Hastada son bir aydır olan paroksizmal gece dispnesi mevcuttu. Ekokardiyografide sol atriyumuna bası yapan ekolüsent bir kitle tespit edildi ve bunun daha sonra akalazyaya olduğu tomografi ile teyit edildi. Hastanın gece olan dispnesi sol atriyal volumün azalmasına ve pulmoner venöz hipertansiyon gelişmesine neden olan kitlenin sol atriyum üzerindeki etkisine bağlandı. Nefes darlığı ile başvuran bir hastada ekokardiyografide değerlendirme yapılırken sol atriyumun arkasında yer alan yapılara özellikle dikkat etmek gerekir.

Anahtar Kelimeler: Özofagusun gevşeyememesi; kalp atria; veri sıkıştırması

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Achalasia is a rare primary motility disorder causing dilation of the distal two-thirds of the esophagus. The achalasia is responsible for a number of clinical symptoms as a result of stasis and retention of food and liquids in the esophagus, patients frequently experience weight loss, hiccups, chest pain, dysphagia, regurgitation, and nocturnal cough and pulmonary complications.^{1,2} Additionally dilated esophagus may also lead to compression on the heart chambers because of close proximity. We report a case of experiencing paroxysmal nocturnal dyspnea as result of left atrial compression by achalasia.

CASE REPORT

A 61-year old man presented as an outpatient because of increasing paroxysmal nocturnal dyspnea attacks occurring two or three times at a night during the past one month. He denied chest pain, cough, expectoration, fevers, chills and had an unremarkable history for cardiac diseases. The patient's vital signs were as follows: pulse rate, 69 beats/min; respiration rate, 16 breaths/min; blood pressure, 100/60 mmHg. Cardiac examination and bilateral chest auscultation revealed normal findings. Examination of his nose, throat was also unremarkable for any disease causing airway obstruction. Because physical examination yielded no clear explanation for his dyspnea, a further work-up including transthoracic echocardiography (TTE), chest X- ray was necessary at this point.

Chest X-Ray showed mediastinal widening with no clear etiology. Therefore, echocardiography was performed to determine a definitive etiology for dyspnea. Left ventricular dimensions and functions, right heart dimensions were normal. However, an echolucent mass significantly compressing the left atrium from the posterolateral site was clearly seen (Figure 1). Contrast echocardiography was done for possible vascular formation but microvascular bubble was not seen in that mass. A computed tomography (CT) of the chest with, done as part of later definitive diagnostic procedure for the origin of the mass, demonstrated a heavily dilated esophagus (65 mm) due to achalasia that caused posterior impression of the left atrium (Figure 2). CT also revealed smoothly tapering of distal esophagus, suggestive of primary motility disorder without a neoplastic lesion (Figure 3). A barium esophagogram confirmed the presence of massive esophageal dilation with tapering of the distal esophagus and delayed passage into the stomach (Figure 4). The dilated esophagus also had a mild impression over the trachea and main bronchus at the carinal level.

Because the patient had congestive symptoms at nights due to mass effect on the left atrium of the achalasia and at this point, primary goal was to restore adequate blood flow to the left atrium by re-



FIGURE 1: Apical 4-chamber view demonstrating prominent distortion (arrows) posterolateral wall of left atrium by dilated esophagus. The left atrium was small in size. (Es: Esophagus, LA: Left atrium, LV: Left ventricle)

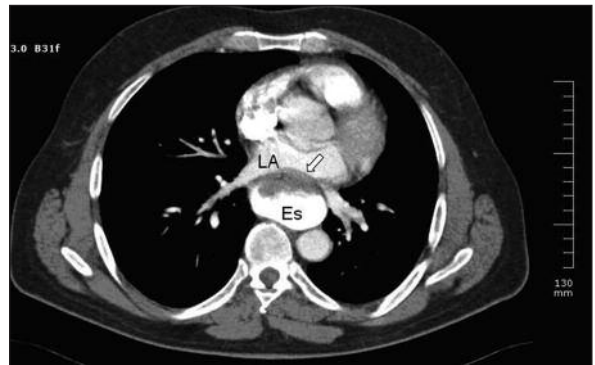


FIGURE 2: Axial contrast enhanced CT image with simultaneously barium swallowing demonstrates achalasia having maximal diameter of 65 mm (arrow). Extrinsic compression of left atrium on the cranial aspect of left atrium by achalasia is clearly seen. (Es: Esophagus, LA: Left atrium)

moving mass effect, the patient was referred to gastroenterology unit for further therapeutic approach.

DISCUSSION

The case presented here identified the structural effect of a massively dilated esophagus on the left atrium. Two-dimensional echocardiography clearly revealed anatomic distortion of the left atrium due to a mass effect of the dilated esophagus.

Echocardiography is commonly used for dyspnea etiology.³⁻⁵ However, findings on TTE analysis are normal and not provide a direct explanation for dyspnea; it must be given special attention to the dorsal region of the left atrium.³ In some cases,

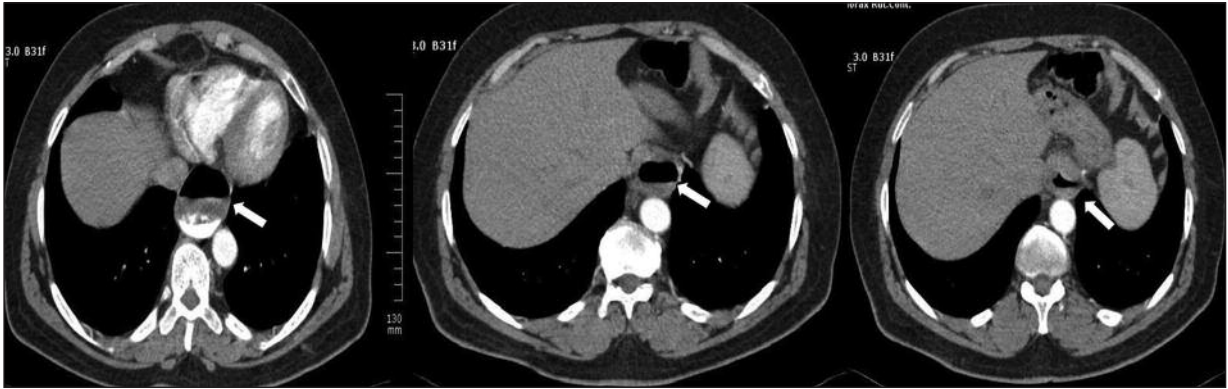


FIGURE 3: Serial axial contrast enhanced CT images demonstrates narrowing of lower esophageal sphincter (arrows). There was also no suspicion of neoplastic infiltration originating from esophagus or stomach.

the identification and diagnosis of extracardiac masses neighboring to the heart can be found in that area which can account for dyspnea.^{3,4} The left atrium is an infero-posteriorly located cardiac chamber with a low intraluminal pressure and has relatively thin wall, making, highly susceptible to compression from the nearby structures.³ A variety of conditions can cause heart chambers compression with varying degree and cause tachycardia, hypotension hypoxia, pulmonary venous congestion and arrhythmia which are known as the findings of hemodynamic compromise.⁵⁻⁸ These structures involve gastrointestinal tract diseases (diaphragmatic and esophageal hernia,⁹⁻¹² esophageal leiomyosarcoma,¹³ achalasia,^{4,5,7,14} chronic gastric volvulus by a para-esophageal hernia,¹⁵ and a diaphragmatic hernia,^{9,11} esophageal hematoma,¹⁶ diffuse large B cell lymphoma presenting originating from esophageus,¹⁷ pancreatic pseudocyst¹⁸); mediastinal masses (thymoma¹⁹/schwannoma,²⁰ sarcoidosis³, mediastinal lymphoma,²¹ angiomyolipoma⁸ metastatic germ cell tumor⁶, thymic cyst,²² teratom cyst²³); pulmonary diseases (lung tumor,²⁴ bronchogenic cyst²⁶) and aorta/pericardial diseases (ascending²⁶/descending aortic aneurysm,²⁷ aortic root dilatation and scoliosis,²⁸ pseudoaneurysm with subepicardial dissection onto the left atrial wall,²⁹ hematoma from rupture of type B aortic dissection,^{30,31} pericardial cyst³² and hematoma^{33,34}). The esophagus and the descending aorta would be expected more to produce left atrial compression because of anatomic proximity.³

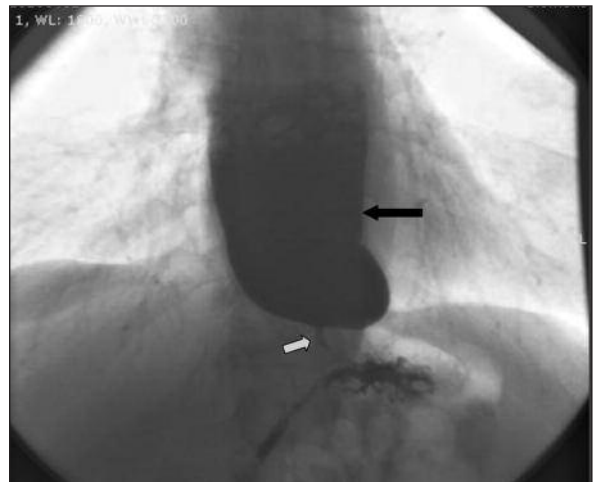


FIGURE 4: Left anterior oblique of barium swallow X-ray clearly demonstrates a dilated esophagus (black arrow) and heavily narrowing of lower esophageal sphincter. There was also minimal passage into the stomach (white arrow).

Left atrium compression may mimic symptoms like congestive heart failure.³ Especially in lying position at nights, venous return from the legs to the heart increases preload. In addition, compression of the left atrium reduces the volume of the left atrium and causes low cardiac output, giving dyspnea, reduced exercise tolerance, or even hemodynamic instability. Along with increased preload to left atrium, left atrial pressure rises with the subsequently elevated pulmonary venous pressure, which may eventually lead to impaired left atrial filling, leading to pulmonary venous hypertension and pulmonary edema.³ This explains the mechanism of paroxysmal nocturnal dyspnea in the

presence of left atrial compression by an extracardiac mass. A combination of these symptoms is often suggestive of heart failure or can mimic a cardiac tamponade.³

Extracardiac impression of the left atrium can present as a variety of clinical conditions such as congestive heart failure, paroxysms of atrial tachycardia and hemodynamic compromise, and should remain in the differential diagnosis for those patients who do not respond to medical therapy of heart failure.^{2,3,5-8,11,12} These patients should be further evaluated using TTE with special attention to the size of the left atrial, distortion of left atrial walls, and the area dorsal to the left atrium.^{3,9,10} Moreover, the lack of evidence of an enlarged left atrium, commonly seen in congestive heart failure on TTE may give a clue for a hidden pathology.

After pursuing a diagnosis of a structure behind the left atrium with TTE, a work-up of additional CT scan can give the greatest diagnostic yield because it provides a variety of information and more insight into its origin.^{2,3,10}

CONCLUSION

The diagnosis of structures dorsal to the left atrium can be visualized on the basis of TTE findings. These structures can increase in size and cause symptoms of inflow disturbance and atrial arrhythmia by compressing the left atrium. Therefore, left atrial compression must be taken into consideration as an infrequent cause of dyspnea and timely detection and recognition is essential for this potentially life-threatening but unusual cause of congestive heart failure.

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