Case Report

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Echocardiography as an Initial Diagnostic Tool for Rare Extra Cardiac Diagnosis- Idiopathic Achalasia

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Abstract

Transthoracic echocardiography (TTE) is a noninvasive initial imaging modality in evaluating the anatomic structure and function of the heart. Although the diagnostic coincidence rate of TTE is slightly lower than that of computer tomography (CT) and magnetic resonance tomography (MRT), TTE has certain diagnostic value for extra-cardiac lesions. We report about a very rare and interesting finding observed using TTE study. Our case shows the role of TTE in identifying extra cardiac structure leading to the new important diagnosis and deferent treatment strategy.

61-year-old male patient was admitted to the hospital with main complaints: chest pain, dyspnea, cough, also he had dysphagia and weight loss. First of all, cardiologic examination has been done, TTE revealed left atrial (LA) compression by an inhomogeneous mass and accelerated flow in LA. TTE provided useful information of the location, size, echogenicity of the extra cardiac structure and its relationship with the heart. Esophageal pathology was suspected, the patient had been subjected to X-ray examination with barium and then an endoscopy. The result of which was confirmed diagnosis of idiopathic achalasia.

Our case shows the role of routine TTE in identifying extra cardiac structure - esophageal achalasia, leading to important extra cardiac diagnosis and indication for further surgical intervention.

Incidental echocardiographic extra cardiac findings in the clinical case presented by us led to a new diagnosis, referral of patients to surgeons and, accordingly, changed the treatment strategy.

Thus, it is very important to train, improve knowledge, focus on extra cardiac manifestations on echocardiography and establish appropriate guidelines for conducting a comprehensive study.

Keywords: ***Achalasia; Transthoracic echocardiography; Extra cardiac manifestation.***

Introduction

Transthoracic echocardiography is a noninvasive primarily initial imaging modality in evaluating the anatomic structure and function of the heart. With widespread availability, portability, low cost and safety TTE presents a mainstay of cardiac imaging. In some cases, it may also present as a diagnostic tool to identify different non-cardiac structures within multiple anatomical areas [1,2]. Incidental extra

cardiac findings (INCF) are chiefly investigated and described by cardiac computed tomography and magnetic resonance imaging. The literature in this field about the role of TTE is very sparse. The majority of incidental findings are clinically insignificant; however, some may cause symptoms or require further investigation and management [3,4].

Our case shows the role of routine TTE in identifying extra cardiac structure - esophageal achalasia, leading to important diagnosis and indication for further surgical intervention.

Achalasia is a motility disorder of the esophagus, characterized by impaired peristalsis and inadequate relaxation of the lower esophageal sphincter. Patients most commonly experience dysphagia with solids and liquids, regurgitation, and intermittent chest pain with or without weight loss. Using high-resolution manometry, three subtypes of achalasia have been identified that differ in pressure and contraction patterns. Important diagnostic signs are endoscopic findings of saliva residues with folds at the esophagogastric junction or findings of a dilated esophagus with a bird's beak [13-15].

Achalasia is one of the most studied esophageal motility disorders. In this guide, we discuss the diagnosis, treatment, and general management of adult patients with achalasia. This guidance includes recommendations, key concepts and a summary of the evidence. Each recommendation statement is accompanied by an assessment of the quality of the evidence and the strength of the recommendation based on the GRADE (Grading of Recommendations Assessment, Development and Evaluation) process. Key concepts are statements that do not meet the requirements of the GRADE process due to the structure of the statement or the available evidence. In some cases, key concepts rely on the extrapolation of evidence and/or expert opinion. Although the nature of the symptom may influence clinical judgment, it may be difficult to determine which of these causes is affecting the patient. Therefore, additional diagnostic testing is necessary unless the patient is experiencing heartburn and a PPI is attempted. High-resolution manometry allows you to assess the integrity of the myotomy, as well as determine the presence of spasmodic contractions after treatment; However, it is not possible to accurately determine bolus delay and assess the contribution of GERD, and the procedure can be difficult due to obstruction and abnormal anatomy[16-19].

Many patients with refractory achalasia or terminal achalasia, characterized by a barium-like esophagus with severe dilation (>6 cm in width) and complex anatomical deformity (concha trap), experience severe symptoms and complications. potentially fatal. Therefore, measures should be taken to prevent aspiration and malnutrition. And the dead. Unfortunately, esophagectomy is associated with a high complication rate and a real risk of death. Additionally, quality of life is reduced after esophagectomy, so this approach should be considered a last resort, and most patients and doctors prefer a more conservative treatment attempt. For patients in whom PD and POEM methods have failed, attempting Heller myotomy may still be reasonable before resorting to esophagectomy, given the number of cases in which severely terminally ill patients may respond to surgery. You should know that the success rate still remains much lower than that of patients with a more favorable anatomy and without prior radical treatment. A comprehensive evaluation including assessment of esophageal anatomy with barium, upper esophageal endoscopy to assess for esophagitis and strictures, and possibly manometry or FLIP to assess LES function may provide evidence that a disease-targeted level of WIS therapy can be effective. Patients with severe anatomy, significant bolus delay, and evidence of complete myotomy may be referred for esophagectomy, while patients with evidence of incomplete myotomy may undergo Heller myotomy. Ultimately, this decision is extremely complex and this approach requires careful assessment and informed discussion, focused on risks and benefits. Patients requiring esophageal resection should be referred to specialized centers with a large number of patients, because the results directly depend on the number of patients and their experience [20-23].

Patients with symptoms suggestive of achalasia should undergo upper gastrointestinal endoscopy to ensure the absence of other pathologies and to rule out possible pseudo achalasia. Heart rate management and timed barium swallows should be used to confirm the diagnosis. The choice between treatment modalities depends on the manometric subtypes of achalasia, patient preference, and institutional experience. PD, HM and POEM are good choices for people with achalasia type I and II. PD should be performed gradually, starting with the smallest balloon (3.0 cm), except in young men (<45 years) who require one initial 3.0 cm to 3.5 cm balloon or surgical myotomy. Patients who do not respond to Parkinson's disease should undergo surgical myotomy. Individual CM or POEM can be used in patients with type III achalasia. If patients are not suitable for radical treatment due to comorbidities, treatment with botulinum toxin and smooth muscle relaxants should be offered. To maximize patient outcomes, all definitive treatments should be offered in centers of excellence with sufficient capacity and experience. Patients should be monitored for recurrence of GERD symptoms and complications after the procedure. CE and endoscopy can be complementary in assessing disease recurrence versus inflammation or stenosis associated with reflux. Repeat PD, CM, or POEM may be performed in individuals with recurrent disease, and antacid therapy should be offered to individuals with symptoms due to GERD. People with a dilated esophagus (more than 8 cm) and an unsatisfactory response to the initial myotomy may require an esophagectomy [24-26].

Cardiac achalasia, a form of dynamic esophageal disorder, is a relatively rare primary esophageal movement disorder characterized by loss of function of ganglion plexus cells in the distal esophagus and lower esophageal sphincter. Loss of distal and lower esophageal sphincter ganglion cell function is a major cause of cardiac achalasia and is more common in older adults. Histological changes in the esophageal mucosa are considered pathogenic; However, research has shown that inflammation and genetic changes at the molecular level can also cause cardiac achalasia, leading to dysphagia, reflux, aspiration, chest pain and weight loss. Current treatment options for achalasia aim to reduce the resting pressure of the lower esophageal sphincter, which helps empty the esophagus and relieve symptoms. Treatment options include botulinum toxin injection, inflatable dilatation, stenting, and surgical myotomy (open or laparoscopic). Surgical procedures are often controversial due to concerns about their safety and effectiveness, especially in older patients. Here, we review clinical, epidemiological, and experimental data to determine the prevalence, pathogenesis, clinical presentation, diagnostic criteria, and treatment options for achalasia to aid clinical management [28-30].

As an important part of the digestive system, the esophagus plays an important role in transporting nutrients. Diseases of the esophagus can be classified into anatomical lesions of the organic cavity (e.g.: digestive or eosinophilic stenosis) or severe dysphagia with progressive injection of the digestive tract (e.g.: severe dysphagia of neurological origin or cardiac achalasia). Esophageal achalasia is a form of dynamic esophageal disorder (DED). It refers to obstruction of the esophageal outflow tract due to impaired relaxation of the lower esophageal sphincter (LES) and

loss of esophageal motility or spasmodic contraction when the esophageal body or esophagogastric junction (EGJ) is not structurally blocked. There are primary and secondary types of achalasia. Cardiac achalasia is characterized by a loss of functional muscle ganglion cells in the distal esophagus and LES. Although histological changes in the esophageal mucosa have long been considered part of the pathogenesis of cardiac achalasia, recent studies have shown that inflammation and genetic changes may also contribute to achalasia at the molecular level. Currently, achalasia is an incurable chronic disease. Different subtypes of achalasia respond differently to medications and surgery, after which some patients develop submucosal fibrosis. This may recur and require additional treatment [32-34].

Background

Achalasia symptomatic consequence the motility disorder is the classic form of solid-liquid dysphagia associated with regurgitation of soft undigested food or saliva. Substernal pain while eating, associated with dysphagia, weight loss, and even heartburn, can accompany symptoms that often lead to achalasia being misdiagnosed as gastroesophageal reflux disease (GERD). Achalasia should be suspected in individuals with solid-liquid dysphagia and in those with ineffective regurgitation after the first use of proton pump inhibitor (PPI) therapy. Endoscopic evidence of retention of saliva, fluids and food into the esophagus without mechanical obstruction due to stenosis. or the mass should arouse suspicion of achalasia. In contrast, other diseases can mimic achalasia both clinically and manometrically. These include pseudoachalasia due to tumors of the gastric cardia or tumors infiltrating the myenteric plexus (adenocarcinoma of the gastroesophageal junction, pancreatic, breast, lung or hepatocellular carcinoma) or secondary achalasia due to external processes such as anterior dense fundoplication or laparoscopic adjustable gastric banding [36-38].

Achalasia is a primary movement disorder of the esophagus, characterized by lack of peristalsis and insufficient relaxation of the lower esophageal sphincter. Given new advances and developments in the treatment of achalasia, there is a growing need for comprehensive evidence-based guidelines to assist clinicians in treating patients with achalasia. Achalasia is a relatively rare primary esophageal motility disorder characterized by loss of function of the ganglion cells of the distal esophageal plexus and the lower esophageal sphincter. Histological changes in the esophageal mucosa are considered pathogenic; However, research has shown that inflammation and genetic changes at the molecular level can also cause cardiac achalasia, leading to dysphagia, reflux, aspiration, chest pain, and weight loss. The aim of this review article is to provide a comprehensive overview of the literature and to present the current state of knowledge on the subject of achalasia [40-42].

Currently, the etiology and pathogenesis of cardiac achalasia remain unclear; However, it is generally accepted that histological changes in the esophageal mucosa caused by loss of esophageal nerve cell function play a fundamental role in the pathophysiology. Autoimmune attack on the esophageal intermuscular nerves by cell-mediated mechanisms and possibly by antibodies can lead to inhibition of esophageal smooth muscle, resulting in loss of nerve function and degeneration of nerve fibers [43-44].

Several pathological mechanisms have been proposed as possible triggers of this immunodeficiency process, including underlying viral infections, idiopathic autoimmune triggers, and genetic predisposition. Herpes zoster virus, herpes simplex virus, measles virus, and human papillomavirus may influence the regulation of functional esophageal movement and control of LES in patients with achalasia, but not in all patients with viral infections. Many patients with achalasia have varicella-zoster virus DNA in their saliva. Limited evidence suggests that eosinophils and mast cells may play a role in the development of achalasia and obstructive esophageal motility disorders. Aggregation of eosinophils and mast cells in the esophagus leads to increased levels of inflammatory cytokines; This leads to fibrous remodeling of the esophageal wall, which ultimately leads to esophageal dysfunction and associated symptoms. Dysphagia (solid or liquid) is a common symptom in patients with achalasia. Initially, this symptom appears sporadically; However, as the disease progresses, the esophagus enlarges significantly, resulting in burning and decompensation of the sigmoid colon with corresponding clinical symptoms. Additionally, patients with achalasia may experience chest pain [5]. There is also a risk of long-term aspiration pneumonia and esophageal squamous cell carcinoma [17]. A study using UK hospital and primary care databases found that patients with achalasia had high morbidity and mortality from oesophageal cancer, aspiration pneumonia and lower respiratory infections [37-39] Additionally, other studies have shown that patients with achalasia suffer from acute respiratory failure and hemodynamic instability. The final stage of achalasia with hypertrophy of the thoracoesophageal region can manifest as an acute illness. Achalasia often presents insidiously, with many subclinical features before a definitive diagnosis is made, which can result in a delay between symptom onset and diagnosis.

Currently, cardiac achalasia is mainly diagnosed by high-resolution manometry (HRM), endoscopy, and barium meal examination. Temporary barium swallow esophagography or functional luminal imaging probe (FLIP) is only used if achalasia cannot be diagnosed [45-46].

Manometry plays an important role in the differential diagnosis of dynamic esophageal diseases. Human resources management is the gold standard in the diagnosis of achalasia cardiaca. HRM typically involves performing a pressure test using a minimum of pressure sensors distributed throughout the catheter. Each pressure sensor is spaced 1 cm apart to obtain baseline values at rest. The probe enters through the nose and passes through the esophagus into the esophagus, allowing the entire esophagus to be examined. In fact, HRM can not only confirm the diagnosis of achalasia, but also identify specific subtypes demonstrating significantly different therapeutic outcomes. Correct diagnosis of intraoperative HRM can help guide therapeutic approaches and predict therapeutic outcomes. Endoscopy is necessary in patients with digestive disorders, although it is not very sensitive to achalasia. Studies have shown that only a third of patients can be diagnosed with achalasia via endoscopy. Endoscopy is commonly used to evaluate patients with gastrointestinal symptoms and to exclude luminal neoplasms of the esophagus and proximal stomach.

Drugs are usually prescribed to patients who cannot undergo or refuse endoscopic or surgical treatment, as well as to those in whom endoscopic or surgical treatment has failed. Calcium channel blockers, nitrates, and proton pump inhibitors are commonly used to control acid reflux; However, they provide only short-term relief and are less effective [31,42,47].

In the early stages of the disease, dysphagia may be very subtle and may be misinterpreted as dyspepsia, poor gastric emptying, or stress. Heartburn due to stagnant food can increase this confusion. As the disease progresses, difficulty swallowing solid and liquid food usually occurs. Dysphagia affects solid foods more than liquids. To facilitate the flow of food, patients usually change their eating habits: They eat more slowly or resort to certain maneuvers, such as raising their arms or arching their backs. The most common

misdiagnosis of achalasia is GERD, as many patients misinterpret regurgitation symptoms as reflux.8 It is important to ask about dysphagia or shutdown symptoms and to be alert to the possible diagnosis of achalasia in patients whose condition does not improve. Treatment with PPIs after initial suspicion of GERD.

In this section, an algorithm for the individual treatment of patients with achalasia is presented. Symptomatic patients with achalasia who are good candidates for surgery should be provided with information about the risks and benefits of equally effective treatment options such as MP and myotomy. The choice between procedures should depend on patient preference and facility experience. However, to achieve maximum results for patients, both procedures must be performed in centers of excellence with sufficient size and experience. PD should be performed gradually, starting with the smallest balloon (3.0 cm), except in younger men (<45 years), in whom initial placement of a 3.5 cm balloon or surgical myotomy may be beneficial. Patients who do not respond to PD should undergo surgical myotomy. Surgical candidates with poor indications should initially inject botulinum toxin into the LES and be aware that retreatment is often required. Other treatments based on nitrates or calcium channel blockers may be offered if there is no clinical response to botulinum toxin injection. Individuals with a dilated esophagus (>8 cm) and a poor response to the initial myotomy may require an esophagectomy.

Achalasia is a relatively rare disorder of esophageal motility. The main clinical manifestations are dysphagia, reflux, chest pain and weight loss; They can significantly affect the patient's quality of life. Treatment for achalasia cardiaca is primarily aimed at relieving symptoms, as there is no cure for the disease. POEM is expected to be an ideal treatment for cardiac achalasia due to its effectiveness and safety. Individual treatment should be carried out taking into account the clinical characteristics of each patient. Currently, clinical studies on cardiac achalasia suggest the possibility of infectious events associated with certain genetic factors triggering the autoimmune mechanism. However, further research is needed in related areas to determine optimal treatment regimens [11,25,39].

The clinical care of patients with achalasia has changed significantly in the past decade under influence of new developments such as high-resolution manometry, per-oral endoscopic myotomy and studies providing new insights regarding achalasia subtypes, cancer risk and follow-up. Given the substantial growth of knowledge in the past years, there is need for a comprehensive, evidence-based European guideline covering all aspects of the disease. This multidisciplinary guideline aims to provide an evidence-based framework with recommendations on the diagnosis, treatment and followup of adult achalasia patients. Chagas disease and achalasia secondary to other disorders, as can be seen after fundoplication, bariatric surgery, sarcoid infiltration, opiate usage or malignancy, is not covered by this guideline. This guideline is intended for clinicians involved in their management, including gastroenterologists, endoscopists, radiologists, gastrointestinal surgeons, dietitians and primary care practitioners[12,29,36].

The diagnosis of achalasia should be considered when patients present with dysphagia associated with other esophageal symptoms and when upper gastrointestinal endoscopy can exclude other conditions. Barium esophagography may reveal the classic bird's beak sign, esophageal dilation, or free fall. Esophageal manometry is the gold standard for diagnosing achalasia; A diagnostic sign is incomplete relaxation of the VOC, expressed by an increase in integrative relaxation pressure in the absence of normal peristalsis. The use of high-resolution manometry (HRM) has led to the classification of achalasia into three clinically relevant groups based on esophageal contractility patterns.

Malignant pseudoachalasia is a condition in which a patient is initially diagnosed and sometimes even treated for achalasia, but is later found to have a malignant disease as the underlying cause. This may occur with submucosal adenocarcinoma of the cardia, locally advanced pancreatic cancer, submucosal metastases, or anti-Hu-producing small cell carcinomas (most commonly lung carcinoma). All Patients diagnosed with achalasia should under no circumstances undergo additional tests such as: B. undergo a CT scan or endoscopy. Ultrasound to exclude malignancy. Interpretation of temporary barium esophagus. In a patient with achalasia, radiographs were taken at 0, 1, 2, and 5 minutes in the left posterior oblique position after ingestion of 100 to 200 ml of low-density barium suspension. Measurement of the height and width of a barium column measured from the OGJ to the barium-foam interface. A barium height > 5 cm at 1 minute and > 2 cm at 5 minutes indicates achalasia. The time is lost if malignant disease is not recognized at an early stage. Only two studies have addressed the question of how to identify patients with malignant pseudoachalasia. significant duration of weight loss and aging. A study by Ponds et al. also identified difficulties inserting the endoscope into the stomach, which endoscopists identified as a risk factor. A model was developed in which the presence of fewer than two risk factors did not increase the risk of malignancy, whereas the presence of two or more risk factors increased the risk [14,18,26,35,44].

Idiopathic achalasia is a primary esophageal motor disorder characterized by loss of esophageal motility and inadequate relaxation of the lower esophageal sphincter in response to swallowing. Patients with achalasia often complain of solid and liquid dysphagia, mild regurgitation that often does not respond adequately to proton pump inhibitors, and chest pain. Many, but not all, patients experience weight loss. Although the exact etiology is unknown, it is often thought to be an autoimmune disease, viral immune disease, or neurodegenerative disease. Diagnosis is based on history, barium esophagus, and esophageal motility studies. To exclude malignancy, endoscopic evaluation of the gastroesophageal junction and gastric cardia is necessary. New diagnostic techniques such as high-resolution manometry help predict treatment response in achalasia based on esophageal pressure topography patterns that identify three achalasia phenotypes (I-III), and studies of Results show improved treatment response in Types I and II compared to Types III. Although achalasia cannot be completely cured, excellent results are obtained in more than 90% of patients. Modern medical and surgical treatments (pneumatic dilatation, endoscopic and surgical myotomy and pharmacological agents) aim to reduce the pressure of the LES and facilitate emptying of the esophagus under the influence of gravity and hydrostatic pressure of food debris and liquids. Staged pneumatic dilatation or laparoscopic surgical myotomy with partial fundoplication is recommended as initial treatment, depending on the patient's age, gender, preference, and local experience. The prognosis of patients with achalasia is excellent. Most patients who receive adequate treatment have a normal life expectancy, but the disease recurs and the patient may require periodic treatment.

The increased prevalence of circulating antibodies against the myenteric plexus in some patients with achalasia suggests that autoantibodies play a role in the pathogenesis of this disease. It has been suggested that these circulating antibodies are likely the result of a nonspecific reaction to the disease process rather than its cause. This idea was supported by the discovery of similar antibodies in patients without achalasia. Ultrastructural studies of esophageal tissue in patients with achalasia also revealed inflammatory

infiltrates around myenteric neurons, whereas controls showed a normal myenteric plexus without infiltration. Several case-control studies have reported a significant association with HLA class II antigens in idiopathic achalasia. Patients with achalasia and its associated HLA allele were found to have a higher prevalence of circulating antimyenteric autoantibodies, suggesting an autoimmune etiology. HLA association also suggests immunogenetic susceptibility to idiopathic achalasia; However, this should be treated with caution since not all patients with achalasia have associated HLA antigens. Recent genetic association in patients with achalasia revealed classical HLA haplotypes and amino acid polymorphisms, suggesting immune-mediated processes in idiopathic achalasia [4,9,23,41].

The presence of familial cases may indicate that achalasia is, in some cases, a hereditary disease. Such familial cases have been observed mainly in children, between siblings and, in some cases, in identical twins. There are also reports of a parent-child association in achalasia.61 Although these data suggest an autosomal recessive mode of inheritance for this disorder, the rarity of familial occurrence does not support the hypothesis that genetic inheritance occurs in most cases. case. Case of achalasia. Instead, it is suggested that genetic predisposition in these individuals likely increases their susceptibility to acquiring achalasia following exposure to common environmental factors that may play a role in pathogenesis [28,35,38,43].

The treatment of achalasia is the result of an evidence-based approach and international interdisciplinary efforts. The guidelines provide advice on key aspects of the diagnosis and treatment of achalasia, as well as commentary based on the best available literature and the opinions of leading European achalasia experts. The main aim of these recommendations is to reduce variations in practice and improve patient outcomes in Europe. Careful and widespread dissemination of these recommendations is therefore necessary to ensure high compliance in clinical practice. It is important to promote counseling and education. Future well-designed clinical trials should address the knowledge gaps and unmet needs that emerged during the development of these guidelines.

Case report

A 61-year-old man with unremarkable past medical history presented to the hospital with a 6-month history of chest pain, dyspnea, cough, dysphagia and weight loss (about 15kg). Chest pain, dyspnea and cough were worsening in the postprandial period, with regurgitation of undigested food.

Physical examination revealed irregular heartbeat, without appreciable heart murmurs, clear lung sounds.

Electrocardiography showed sinus rhythm with supraventricular extra heart beats and nonspecific St Segment and T-wave changes in leads V3-6.

Echocardiography revealed mild hypertrophy of the left ventricle with normal systolic function, without regional wall motion abnormalities, diastolic dysfunction - grade I (abnormal relaxation), right ventricular systolic function was preserved; Estimated pulmonary artery systolic pressure (PASP) was normal of 28mm/Hg. The valves were without significant structural changes; 2D and color Doppler echocardiography revealed left atrial compression by an inhomogeneous mass in close relation to the pulmonary veins and accelerated flow in LA (**Figure 1, Figure 2**).

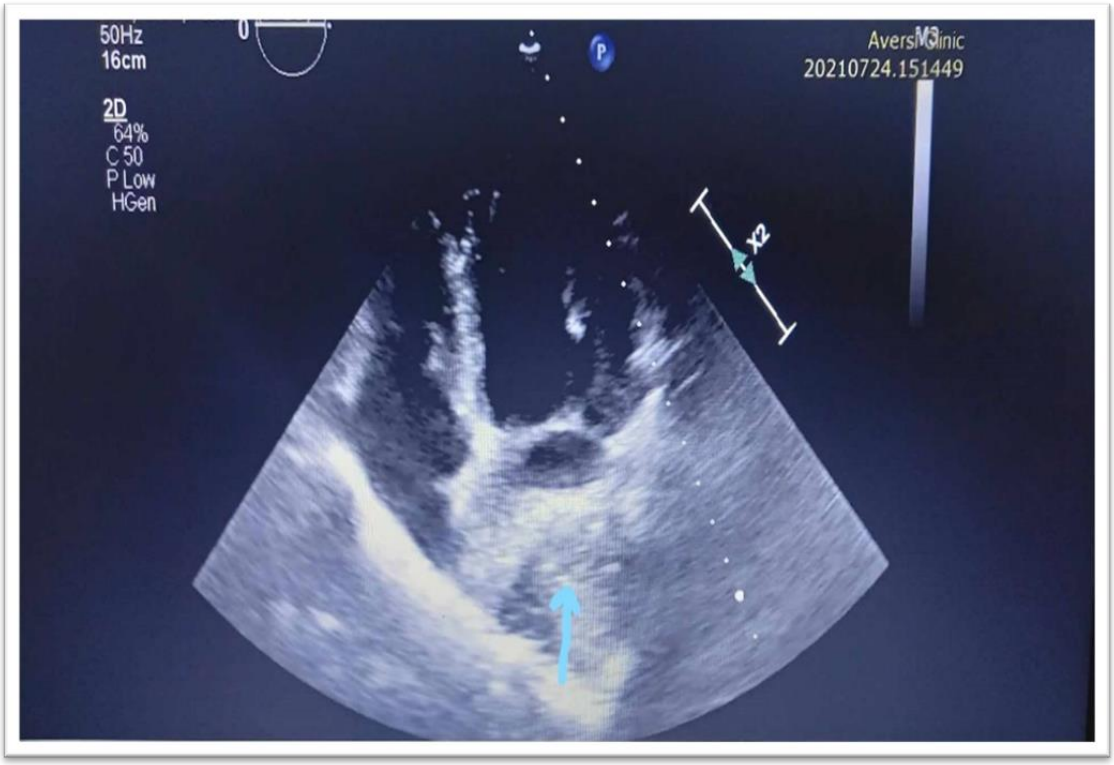

Figure 1: Apical 4-chamber view. Left atrial compression with extra cardiac structure (blue arrow)

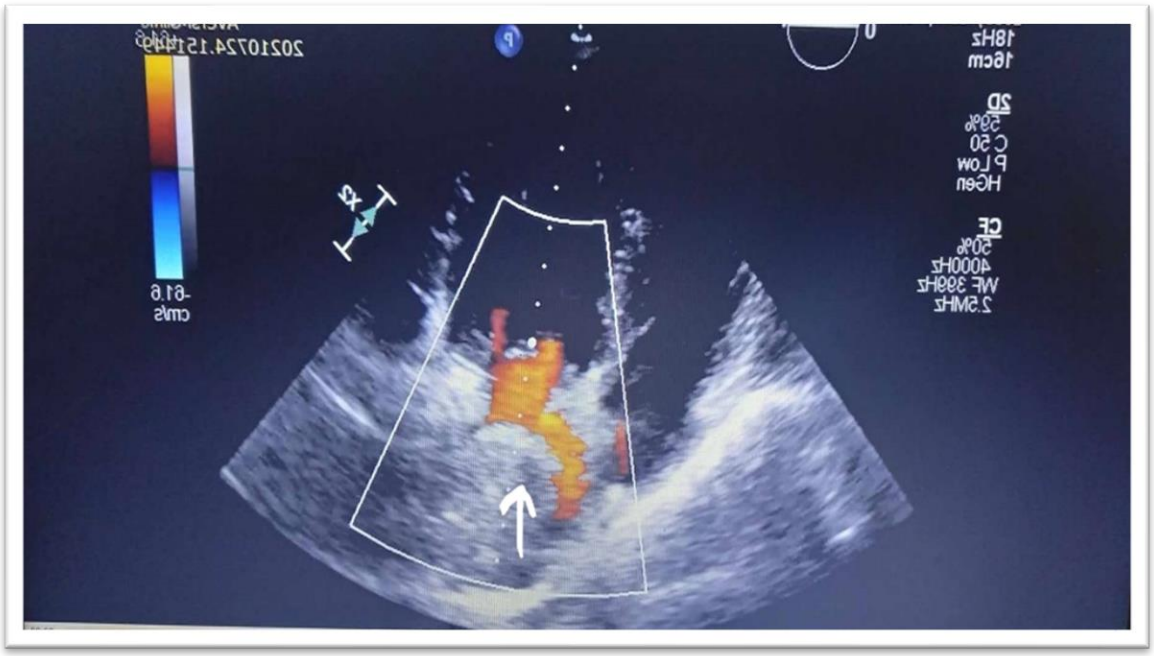

Figure 2: Apical 4-chamber view with color Doppler. Left atrial and pulmonary vein compression with extra cardiac structure (white arrow)

Esophageal pathology was suspected and the patient was referred to the barium swallow study to confirm the diagnosis. The x-ray showed the typical "bird's beak" sign, esophageal dilatation with failure of normal peristalsis, incomplete lower esophageal sphincter relaxation that did not coordinate with esophageal contraction, stasis of barium in the esophagus (**Figure 3**).

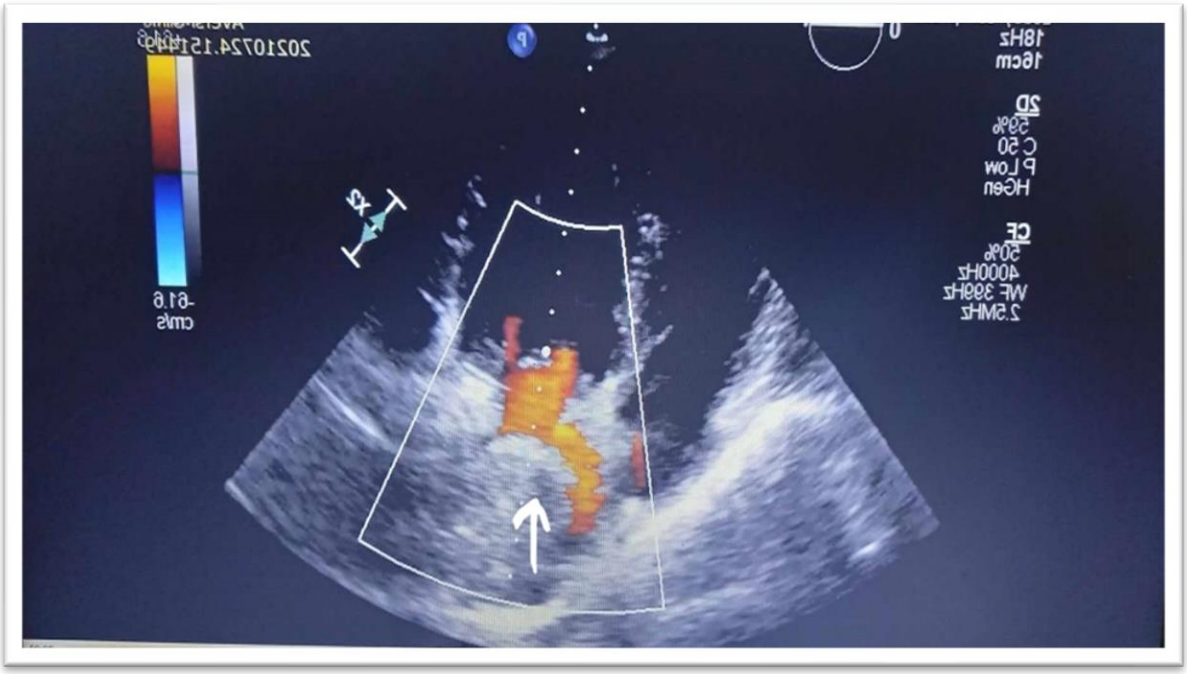

Figure 3: The x-ray shows the typical "bird's beak" sign, esophageal dilatation.

Endoscopy showed dilation of the esophagus with food remnants, whitish coating of the mucosa caused by adhesion of the remained food inside of the esophagus and thickening of the mucosa. Endoscopy excluded esophageal and gastric cancer.

Secondary causes of achalasia, such as: esophageal malignancy and stricture, gastric carcinoma, Chagas disease, systemic sclerosis was excluded.

Coronary angiography was unremarkable and has been performed 1 years ago due to unexplained acute chest pain.

As soon as diagnosis was established, the patient was referred to the laparoscopic Heller myotomy with fundoplication, after which his complaints disappeared and patient fully recovered.

Discussion

Idiopathic achalasia (IA) is a primary esophageal motility disorder characterized by aperistalsis and lower esophageal sphincter dysfunction, with an annual incidence of approximately 1.6 cases per 100,000 individuals and prevalence of 10 cases per 100,000 individuals [5].

IA results from inflammation and degeneration of inhibitory neurons in the esophageal wall that primarily release vasoactive intestinal peptide and nitric oxide. A localized decrease of these substances causes failure of lower esophageal sphincter relaxation and disruption of esophageal peristalsis [6]. The cause of the inflammatory degeneration of neurons in primary achalasia is not known [7]. IA is most frequently seen in middle and late adulthood (age 30 to 70 years) with no gender and racial predisposition.

Left atrial compression by achalasia is a rare presentation and may cause hemodynamic compromises [8].

Patients with achalasia may suffer symptoms for a long time before being diagnosed. The most common symptoms are: dysphagia, regurgitation, chest pain, difficulty swallowing both liquids and solids and thus weight loss. Some patients may have a cough due to aspiration of food particles into their airways [9].

Laboratory studies don't play important role to establish diagnosis of IA.

Several diagnostic tests are routinely utilized for the diagnosis of achalasia: Barium swallow, esophageal manometry, esophagogastroduodenoscopy, prolonged esophageal pH monitoring, endoscopic ultrasonography.

Because conservative treatment of achalasia is not curative, therapeutic success is determined by the improvement in symptoms as reported by patients. All available treatments are hence palliative [10].

Management of achalasia involves improving the esophageal outflow in order to provide symptomatic relief to patients [11].

The treatment modalities employed for this purpose included pharmacological therapy (e.g., Calcium channel blockers, long-acting nitrates), endoscopic interventions (e.g., botulinum toxin injection to lower esophageal sphincter, pneumatic dilation, paroral endoscopic myotomy) and surgical interventions (surgical myotomy, esophagectomy) [12].

Our case shows the role of routine TTE in identifying extra cardiac structure – esophageal achalasia, leading to important diagnosis and indication for further surgical intervention.

Incidental echocardiographic extra cardiac findings in the clinical case presented by us led to a new diagnosis, referral of patients to surgeons and, accordingly, changed the treatment strategy.

Conclusion

Thus, it is very important to train, improve knowledge, focus on extra cardiac manifestations on echocardiography and establish appropriate guidelines for conducting a comprehensive study.

Conflicts of interest

No potential conflict of interest relevant to this article was reported.

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Conflict of Interest

Authors declare no conflict of interest.

Ethics approval

N/A

Data availability

Available upon request

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