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CASE REPORT

Incidental detection of achalasia in a patient with dyspnea

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ABSTRACT

Background: The absence of progressive peristalsis in the esophageal body and aberrant relaxation of the LES are two characteristics of achalasia, which is a main motility disease of the esophageal body and lower esophageal sphincter (LES). The most typical signs of achalasia are dysphagia and recurrent regurgitation.

Case report: We report a case of achalasia in a 54-year-old woman with the main complaint of dyspnea. Physical examination did not reveal any abnormalities. Chest x-ray results showed an enlarged upper mediastinal with air fluid level and superposition with the right border of the heart. The results of a chest computed tomography (CT) scan showed a dilated esophagus containing food material with air fluid levels. The patient then underwent an esophagostomy thoracotomy.

Discussion: Primary achalasia is associated with inflammatory degeneration of the inhibitory ganglion cells of the esophageal myenteric plexus and LES. Dysphagia, regurgitation, problems swallowing food, chest pain, heartburn, and weight loss are the most common symptoms. Barium esophagram is the best diagnostic test for achalasia with "Rat Tail" or "bird's beak" appearance. CT scan examination is not only less invasive and painful, but also has high sensitivity for achalasia. In addition, CT scan are useful for differentiating primary and secondary achalasia and for diagnosing complications of achalasia.

Conclusion: Most cases of achalasia come with dysphagia, but some patients can come with respiratory problems or without complaints. CT scans can help identify achalasia in patients without complaints. Radiological examination plays an important role in detecting achalasia and reducing complications.

Keywords: achalasia, dyspnea, case report

INTRODUCTION

The disease known as esophageal achalasia is characterized by (1) absence of peristalsis in the esophageal corpus, (2) increased LES resting pressure, and (3) inability of the LES to relax during swallowing.[1] The etiology is unknown. One of the most studied primary motility disorders of the esophagus is esophageal achalasia. With an incidence of 1.99 cases per 100,000 people and a prevalence of 27.1 per 100,000 people, this disease is considered rare and the average age is 59 years. However, recent reports suggest that this disorder is more common. This may be due to better diagnostic tools or increased awareness of physicians and patients about this disorder due to the availability of alternative therapeutic methods. Although the exact cause of this disorder is unknown, it is likely multifactorial, with genetics, infection, and autoimmune factors playing a significant role.[2] Failure of the esophagus to empty is caused by abnormal peristalsis and LES spasm. Pathologically, it shows a lack of ganglion cells in the myenteric plexus—also known as Auerbach's plexus—throughout the esophagus.[1]

Clinically, the symptoms are unclear, and usually appear at the age of 30 to 50 years with dysphagia, regurgitation, and aspiration. Dysphagia, which occurs with both solid and liquid foods, is the most common symptom. Other symptoms include regurgitation of food, coughing, aspiration, pneumonia, weight loss, and chest pain that resembles angina, which is worse after eating[3]. In addition, many achalasia patients can tolerate esophageal distension without experiencing complaints. Respiratory distress may be the only symptom of achalasia in some cases. Imaging findings shows (a) uniform esophageal dilation, usually with air-fluid levels; (b) absence of peristalsis, with tertiary waves common in the early stages of the disease; (c)

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tapered “beak” deformity of the LES due to inability to relax; (d) results of esophagitis, including ulceration; and (e) an increase in the number of esophageal carcinomas and epiphrenic diverticula.[1]

CASE REPORT

A 54-year-old woman presented with shortness of breath. Additionally, she had a productive cough with blood and shortness of breath. There was no history indicating regular medication use, radiation exposure, or other medical conditions. The patient had no other previous medical history. Physical examination did not reveal any significant findings. Routine investigations, such as hematological parameters, are within normal limits. The patient will undergo a chest CT scan to determine whether there are lung abnormalities. Chest radiography showed right lateroposterior mediastinal widening with air fluid levels, superposisition with the right heart border (Figure 1).

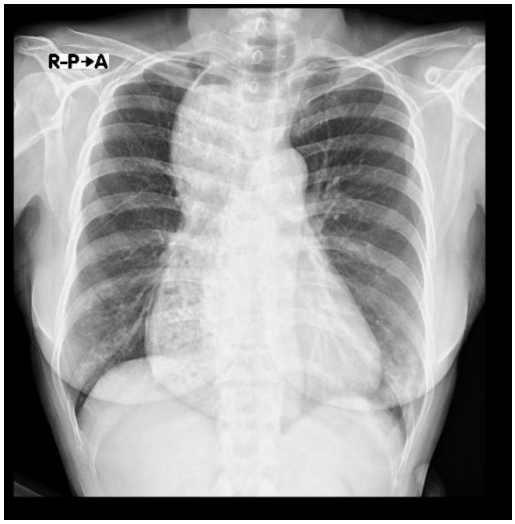

Figure 1. Chest x-ray showed widened mediastinal lateroposterior with air fluid levels, superposition with right heart border

Thoracic computed tomography showed a dilated esophagus containing food material with air fluid levels (Figure 2). After the diagnosis of achalasia was confirmed, the patient was planned to undergo an esophagostomy thoracotomy. After a thorough preoperative examination, an esophagostomy thoracotomy operation was performed. The patient had an uneventful postoperative course. After surgery, thoracic computed tomography was performed. Thoracic computer tomography with contrast showed that the esophagus was still dilated containing food material with an air fluid levels, relatively the same as the previous examination (Figure 3).

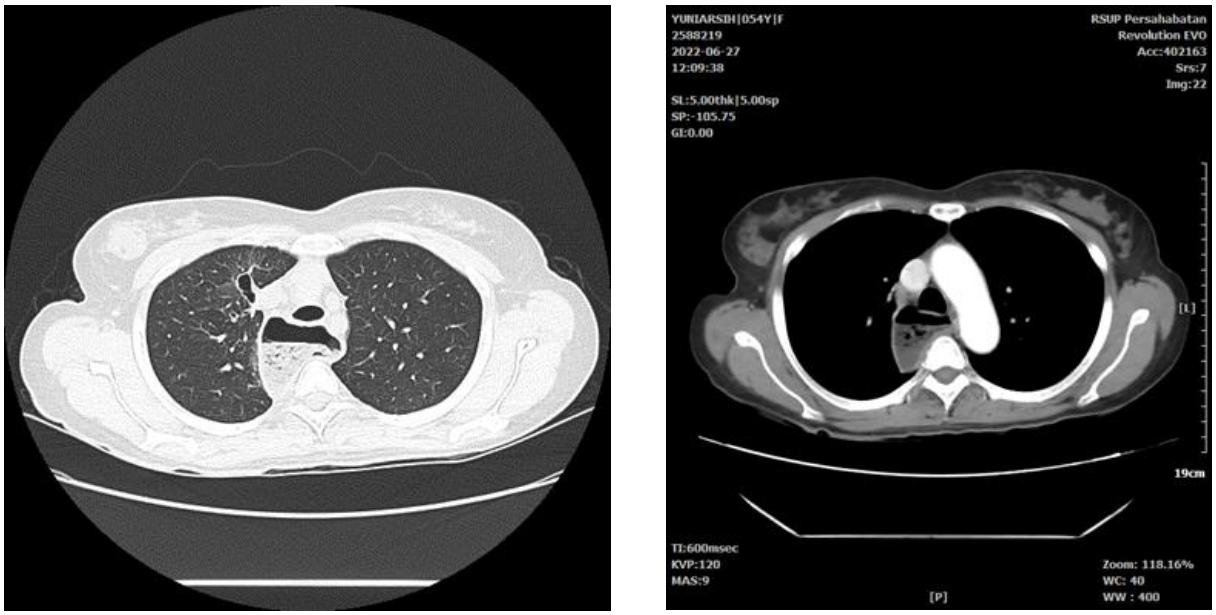

Figure 2. CT scan of the chest revealed dilated esophagus containing food material with air fluid level

DISCUSSION

Primary achalasia is associated with inflammatory degeneration of the inhibitory ganglion cells of the esophageal myenteric plexus and LES, leading to loss of inhibitory neurons. This degeneration causes relaxation of the LES and irregular peristaltic contractions of the esophagus. Achalasia has been described for more than 300 years, but not much is known about its etiology. According to research, there are many factors that can cause achalasia, including infectious agents, autoimmune responses, and genetic factors.[4] Excitatory (such as acetylcholine, substance P) and inhibitory (such as nitricoxide, vasoactive intestinal peptide) neurotransmitters regulate pressure and relaxation of the lower esophageal sphincter (LES) physiologically. Lacking nonadrenergic, noncholinergic, and inhibitory ganglion cells, patients experience

an imbalance in excitatory and inhibitory transmission between their neurons. The result is a hypertensive esophageal sphincter that does not relax.[5]

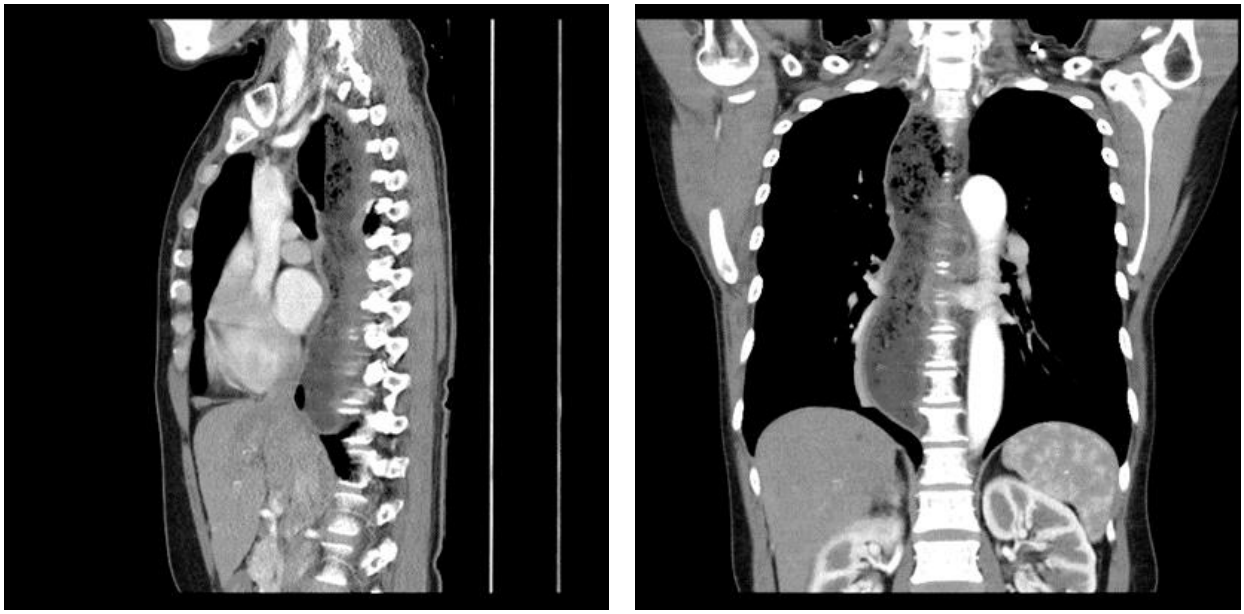

Figure 3. CT scan with contrast of the chest after surgery showed the esophagus was still dilated containing food material with air fluid levels

Approximately 1 in 100,000 incidents of achalasia are reported each year worldwide. Compared with the general population, achalasia patients did not differ in age or gender. Achalasia affects men and women equally, although there are some reports showing a greater impact on women. While other studies suggest that the risk of achalasia increases with age, the bimodal distribution of incidence by age is thought to peak at ages 30 and 60 years.[4] In this case, a 54-year-old female patient was diagnosed with achalasia with complaints of dyspnea and hemoptysis.

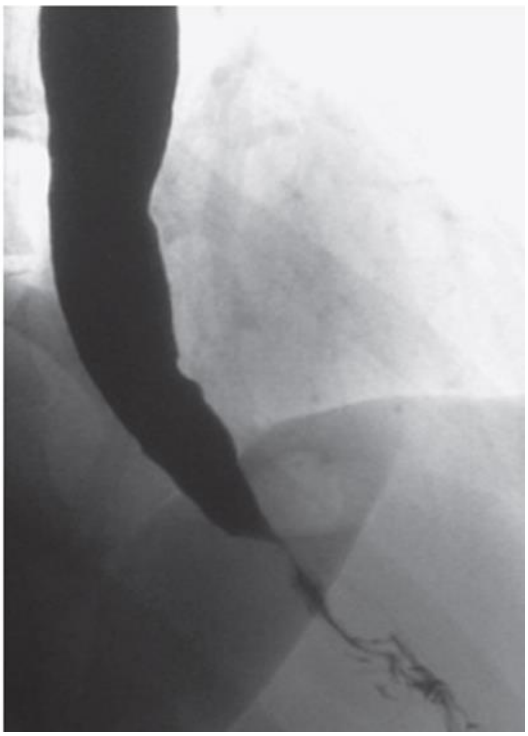

Figure 4. Classical appearance of achalasia, note the tapered appearance of the GOJ.[8]

Dysphagia, regurgitation, problems swallowing food, chest pain, heartburn, and weight loss are common symptoms suffered by achalasia sufferers. If a person complains of progressive dysphagia, both solid and liquid, accompanied by regurgitation of food and saliva, the diagnosis of achalasia should be suspected. Physical examination usually does not reveal any typical abnormalities.[5] Cough, asthma, chronic aspiration, hoarseness, and sore throat are respiratory symptoms of achalasia. Since 1950, only 66 cases of adults experiencing airway obstruction due to esophageal dilatation have been reported. According to this case review, 85.1% of patients were female, with a mean age of 71.3 years for women and 44.8 years for men. This shows that older women more often experience airway obstruction. The most common symptoms are acute dyspnea and stridor, which occur in approximately 90% and 70% of cases, respectively. Other symptoms include dysphagia, neck swelling, cyanosis, and wheezing.[6]

The pathogenesis of megaesophagus caused by achalasia has been discussed in several theories. Several factors can cause esophageal dilatation, such as the formation of a one-way valve due to the esophagus twisting behind the cricopharyngeal muscle, causing air to be trapped; insufficient relaxation of the upper esophageal sphincter; and lack of burping reflex which makes it difficult to exhale air.[6] Additionally, lung aspiration can cause respiratory symptoms. There may be aspiration due to saliva and swallowed food getting stuck in the

esophagus. As the disease progresses and especially as the esophagus begins to enlarge, regurgitation becomes a major problem. Chronic and/or acute lung infections can occur due to aspiration of esophageal contents into the respiratory tract. Most reports show bilateral alveolar opacities similar to aspiration pneumonia.[7] In this case, the female patient suffered from dyspnea and hemoptysis and the physical examination showed no abnormalities. The infiltrate in our patient was unilateral. So, we think that shortness of breath and hemoptysis are suspicious for aspiration pneumonia in this patient.

When there is clinical suspicion, a barium esophagram is the best diagnostic test. Dilatation and tortuosity of the proximal esophagus that has a subtle taper at the lower end that resembles the tip of a sharpened wooden pencil is a sign of achalasia. This is usually called the "Rat Tail" or “bird’s beak” appearance (Figure 4).[5] Endoscopy is performed at the junction of the gastroesophagus and the heart of the stomach to identify pseudoachalasia. All patients must undergo endoscopic examination. To support the diagnosis, esophageal pressure manometry is used. It shows pressure at the gastroesophageal junction about twice normal (40 mm Hg) and relaxation after swallowing is incomplete or absent altogether. On chest radiography, patients with achalasia may demonstrate mediastinal enlargement, masses adjacent to the mediastinum, air fluid levels, and absence of gastric air bubbles (Figure 5). However, these patients often appear normal, especially as the disease progresses. In most previous case reports, chest x-rays of patients with pneumonia causing achalasia showed signs of achalasia. However, even if the chest x-ray does not show these signs, achalasia can still be detected through additional CT scans.[9] In uncomplicated achalasia patients, the esophagus is thin-walled and dilated with fluid or food debris (Figure 6). Overall, CT scans are useful for assessing common complications, but are insufficient for directly assessing a patient's achalasia. The esophageal wall should be examined thoroughly for areas of thickening that could indicate malignancy. If there is aspiration, the lungs should be examined. In this case, the chest x-ray showed widening of the right lateroposterior mediastinum with air fluid levels, and the chest CT scan showed dilatation of the esophagus containing food material with air fluid levels.[10] Based on all the findings, the patient was diagnosed with achalasia.

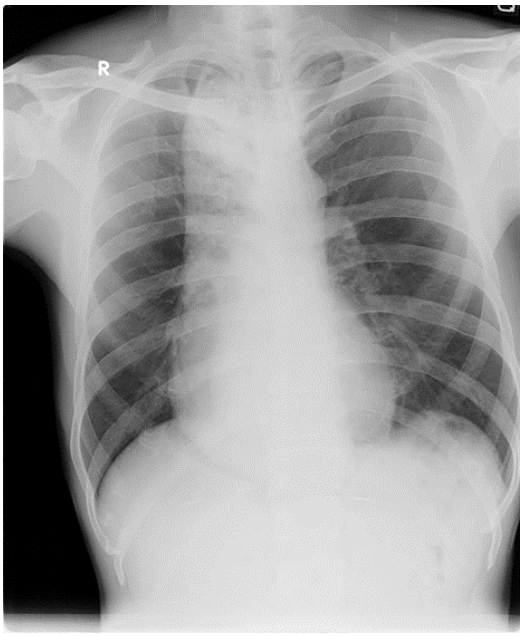

Figure 5. Classic appearance of achalasia on chest radiographs shows mediastinal widening, a trace amount of gastric air-bubble is seen.[11]

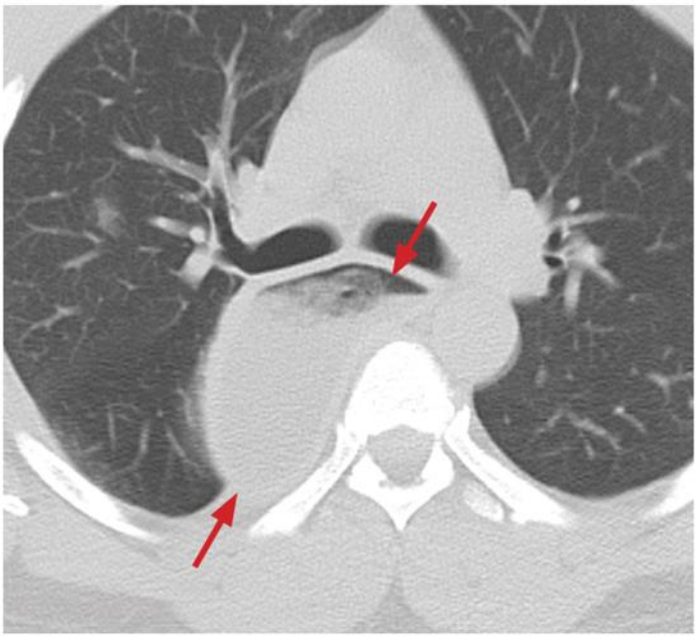

Figure 6. Axial non-contrast CT confirms shows a markedly dilated esophagus, and debris-filled esophagus (red arrow).[12]

Regarding the diagnostic process of achalasia, gold standard diagnosis is mainly based on esophageal manometry, which measures LES muscle pressure.[13] However, only a few facilities can perform all of these tests, making these criteria difficult to apply in the diagnosis of achalasia. Due to lack of facilities at our center, as seen in this case, manometry and esophagography examinations were not performed.

Chest radiography may sometimes be helpful in cases of achalasia where the symptoms and pathognomonic signs are nonspesific.[14] Initially, the patient in this case was suspected to have a lung disorder

because of the unusual symptoms of shortness of breath in achalasia. As shown in this case, a chest x-ray showing a dilated esophagus and absence of gastric air bubbles is suspected as evidence of achalasia. However, a chest x-ray is not enough to make a diagnosis of achalasia. After a chest x-ray, a CT imaging should be performed to confirm the diagnosis and measure the severity of the aspiration. CT also helps differentiate primary and secondary achalasia. In patients with primary achalasia, subtle narrowing of the distal esophagus without thickening of the esophageal wall is more common. Complications of achalasia such as aspiration pneumonia, candidal esophagitis, and secondary esophageal cancer are some that can be diagnosed with CT scans, which are estimated to occur in 2% to 7% of patients. Findings of structural lung pathology on CT scan are found in up to 41% of patients. These findings most often include ground glass opacities.[15]

Chest CT scan and barium swallow test, as screening methods with relatively high sensitivity, are useful diagnostic tests for achalasia. A study by Ishii et al. 2019, shows that the sensitivity of the barium swallow test (88.3-100.0%) and chest CT (83.8-100.0%) for achalasia is quite high, but for endoscopy (50.1-81.4%) it is relatively low. This shows that barium swallow and chest CT have relatively similar sensitivity.[16] Chest CT scan and barium swallow test, as screening methods with high sensitivity, are useful diagnostic examinations for achalasia. Compared with endoscopy and manometry, this examination is not only less invasive and painful, but also has a higher sensitivity to achalasia.

CONCLUSION

We report a case of a 54 years old female with complaints of shortness of breath and hemoptysis. The initial cause was thought to be a lung disorder. After radiological examination, the examination results showed a feature of achalasia. The diagnosis of achalasia was made accidentally. Although esophageal achalasia is a rare disease, a thorough history of dysphagia and regurgitation of solid and liquid foods is essential for detecting the disease. Breathing problems may be similar to achalasia. Moreover, in some cases, they can be found without any symptoms. Getting used to checking the appearance of achalasia on chest X-rays and chest CT can help detect the presence of achalasia and reduce the occurrence of complications. CT scan as screening methods are useful diagnostic examinations for achalasia.

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