

A Case of Achalasia Causing Aspiration Pneumonia

Aspirasyon Pnömonisine Neden Olan Akalazya Olgusu

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ABSTRACT Achalasia is a rare disease that can mimic respiratory symptoms. Chest pain occurs in up to half of patients with achalasia and aspiration pneumonia may occur rarely. In the absence of radiological suspicion of achalasia, a patient may initially be managed for respiratory conditions with subsequent development of complications like aspiration pneumonia, leading to a diagnostic delay. Therefore, this case presentation aims at contributing to clinicians' awareness regarding this condition. A 36 year old female patients presented to our chest diseases department with chest pain and high fever. A contrast enhanced computed tomography of lung suggested achalasia and pneumonia. Achalasia was confirmed and treated with upper gastrointestinal endoscopy and barium swallow study. Treatment for aspiration pneumonia resulted in clinical and radiological improvement. A possible diagnosis of achalasia should be borne in mind by pulmonologists in the differential diagnosis of patients presenting with respiratory symptoms.

Key Words: Esophageal achalasia; pneumonia, aspiration; diagnosis, differential

ÖZET Akalazya, solunumsal semptomları taklit edebilen nadir bir hastalıktır. Hastaların yarısına yakınında göğüs ağrısı görülmekte ve aspirasyon pnömonisi nadiren ortaya çıkabilmektedir. Akalazyanın radyolojik şüphesinin yokluğunda, tamıda gecikme görülebilir ve hasta ilk olarak aspirasyon pnömonisi gibi daha sonra oluşan komplikasyonların solunumsal semptomları ile başvurabilir. Bu nedenle, bu olgu sunumu klinisyenlerin bu durum ile ilgili farkındalığını arttırmaya katkıda bulunmayı amaçlamaktadır. Otuz altı yaşında kadın hasta, göğüs ağrısı ve ateş ile göğüs hastalıkları kliniğimize başvurdu. Akciğer kontrastlı tomografisi akalazya ve pnömoni tanısını düşündürdü. Üst gastrointestinal endoskopi ve baryumlu pasaj grafisi ile akalazya tanısı konuldu ve tedavisi yapıldı. Aspirasyon pnömonisine yönelik tedavi verilen hastada klinik radyolojik yanıt alındı. Akalazya tanısı, göğüs hastalıkları uzmanları tarafından, solunum semptomları olan hastaların ayırıcı tanısında akılda tutulmalıdır.

Anahtar Kelimeler: Özofageal akalazya; pnömoni, aspirasyon; tanı, ayırıcı

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As the thoracic cavity contains non-pulmonary organs such as the heart and esophagus in addition to lungs, non-pulmonary conditions should also be taken into account in the differential diagnosis of respiratory symptoms and radiological signs in patients presenting with respiratory symptoms.

Achalasia is a neurodegenerative motility disorder of the esophagus resulting in a derangement of esophageal peristalsis and loss of lower esophageal sphincter function. Historically, annual achalasia incidence rates were believed to be low, approximately 0.5-1.2 per 100 000. Although acha-

lasia is a relatively rare condition, it carries a risk of complications, including aspiration pneumonia and esophageal cancer.¹ Also, achalasia is a cause of non-cardiac chest pain that can also occur in pulmonary conditions.

Gastrointestinal conditions may be associated with radiological changes in the thorax. Similarly, gastrointestinal diseases may lead to unusual radiological findings as well. Conditions that should be considered in patients with such radiological findings include diverticular disease, carcinoma, duplication cysts, esophageal perforation, or postoperative changes in addition to achalasia. Awareness of changes on the chest radiograph produced by gastrointestinal disease allows prompt diagnosis and facilitates the appropriate confirmatory diagnostic studies, such as esophagography or computed tomography.²

Waking from sleep because of coughing and choking is common in patients with regurgitation of undigested food or accumulated saliva. Also, chest pain in patients with achalasia occurs in up to half of all patients and it has been described as angina-like retrosternal pain.³

Achalasia is not only able to mimic certain respiratory conditions both clinically and radiologically, but also may cause aspiration pneumonia.⁴ Aspiration of esophageal contents into the respiratory tract leads to pulmonary infections.³ Therefore, achalasia represents an important differential diagnosis in patients with aspiration pneumonia, particularly among the younger subjects.

CASE REPORT

A 36 year old female patient presented with severe pleuritic chest pain on the right side. She also had productive cough, dyspnea, and high fever for the past two days. There was no history of regular use of medications, radiation exposure, or other medical conditions. Her body temperature was 39 °C, respiratory rate was 16/min, heart rate was 110/min and blood pressure was 110/70 mmHg. Complete blood count was as follows: WBC $9.59 \times 10^9/L$, hemoglobin 10.3 g/dL, platelet $351 \times 10^9/L$, neutrophil count $6.80 \times 10^9/L$, % neutrophil 70.9, CRP was 6.78 mg/dL. Rhonci and right basal crepitations were present on auscultation. X-ray revealed infiltration in the right lower paracardiac zone as shown in Figure 1A.

A computed tomographic scan of the thorax was performed due to pleuritic pain. However, there was dilation along with marked air-fluid level in computed tomography image of the esophagus and consolidation as shown in Figure 2. So, the patient was diagnosed with pneumonia due to the presence of cough, high fever and pulmonary consolidation in the computed tomography. Treatment for aspiration pneumonia was administered. There was not microbiological growth in sputum sample. A detailed history revealed long-lasting complaint of difficulty in swallowing and regurgitation, which had never been brought to medical attention previously. As shown in Figure 3, a barium swallow study showed the bird's beak deformity



FIGURE 1: a) X-ray revealed infiltration in the right lower paracardiac zone, b) After treatment X-ray, there is a radiological improvement

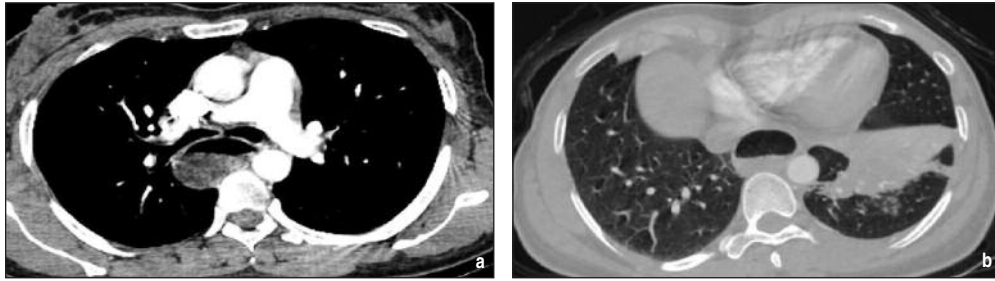


FIGURE 2: a) Dilatation along with marked air-fluid level in computed tomography image of the esophagus, b) consolidation area of right anterior and left lung image.

and a dilated esophagus. A consultation from the department of gastroenterology was requested and a gastrointestinal endoscopy was performed. The lower esophageal sphincter tightly closed around the endoscope and a 35 mm balloon dilatation for achalasia was performed. We achieved a good response and radiological improvement with kinolon treatment given for 10 days as shown in Figure 1B. The patient was referred to the department of gastroenterology for further management and follow up. Informed consent was taken from patient to present this case.

DISCUSSION

In a group of patients with respiratory manifestations, symptoms may actually arise from pathological processes that involve nonpulmonary thoracic organs. For instance, esophageal diseases may readily mimic certain respiratory conditions, subsequently complicating the diagnosis. In this regard, conditions such as Zenker's diverticulum, esophageal fistula, achalasia, esophageal cancer, esophagus cysts, and gastroesophageal reflux disease require further attention in differential diagnosis.⁵ Although pulmonologists rarely see patients with achalasia in their clinical practice, they should be familiar with this condition and perform further workup as needed, since it may delay the diagnosis when confused with conditions such as asthma. In our patient, despite presence of a long history of difficult swallowing, no diagnosis had been established.

The lower esophageal sphincter (LES) may remain contracted due to defective innervation of the smooth muscle of the esophagus and LES resulting in achalasia. The primary etiology of achalasia usually is idiopathic, but secondary causes may include gas-



FIGURE 3: a) Delayed passage of barium in the barium swallow study, b) The bird's beak deformity and a dilated esophagus.

tric carcinoma extending to the esophagus, lymphoma, Chaga's disease, irradiation, and certain medications and toxins. Patients with achalasia may also experience dysphagia, chest pain, and regurgitation, but pulmonary aspiration may occur due to overflow of saliva and ingested food lodged in the esophagus.⁶ Regurgitation becomes a major problem as the disease progresses and, especially, when the esophagus begins to dilate. Aspiration of esophageal contents into the respiratory tract leads to chronic and/or acute pulmonary infections. The majority of reports described cases of patchy bilateral alveolar opacities that resembled aspiration pneumonia.³ Consolidation in our patient was unilateral. The presenting symptom in our patient was chest pain, fever and regurgitation. So, there was aspiration pneumonia symptoms and signs. We achieved a good response with pneumonia treatment. Consequently

the patient was referred to the department of gastroenterology for achalasia etiological diagnosis.

Chest pain is common in patients with achalasia, particularly in younger subjects. Its exact cause is unknown and treatment may not result in complete resolution, although improvement with aging has been reported.⁷ In our patient, an endoscopy was performed by a gastroenterology specialist and a partial improvement was observed following balloon dilatation.

On the other hand, patients with achalasia may also present with complications other than chest pain. Patients with achalasia have an increased risk of aspiration pneumonia. A diagnosis of pneumonia should be considered, particularly in younger patients with pneumonia who are suspected to have aspiration.⁸ Similarly, our patient was young and a concomitant diagnosis of aspiration pneumonia and achalasia was made by contrast enhanced computed tomography. A high fever exceeding 38 °C had suggested aspiration pneumonia, and clinical and radiological response were achieved with appropriate treatment. Also it should be remembered that aspiration of the gastric content may also cause respiratory failure in patients with achalasia and etiological factors other than pneumonia should also be evaluated in patients presenting with high fever and cough.^{3,9} Accordingly our patient had both aspiration pneumonia and achalasia as an etiological factor.

Aspiration involves a spectrum of clinical situations, from laryngeal penetration to frank pulmonary aspiration. In patients with aspiration, airways and lungs become soiled with nongaseous materials including consistencies that are solid or

liquid, caustic or bland, infected or sterile. Pulmonary aspiration can involve segmental or lobar areas of the lung, can be associated with either focal or diffuse inflammatory reactions, and can evolve to include systemic effects such as bacteremia, sepsis, end organ consequences of hypoxia, and death. Our patient had focal segmental involvement. While aspiration pneumonitis implies the presence of an inflammatory response to aspirated material not associated with infection, aspiration pneumonia, on the other hand, implies the presence of infection with pneumonitis.⁶ Pulmonary infiltrations, high fever, and elevated CRP in our patient were consistent with a diagnosis of aspiration pneumonia.

Achalasia may also present as mediastinal enlargement and x-ray findings may mimic cardiomegaly, particularly in patients with a long-standing history of the disease.¹⁰ Our patient also had mediastinal enlargement, although minimal, probably associated with the dimensions of the achalasia and with the absence of a long-standing disease history. A suspicion of achalasia in plain x rays should prompt further imaging with computed tomography, as was the case with our patient. Differential diagnosis and balloon dilatation were performed endoscopically. Currently the patient is being treated and followed up by the gastroenterology unit.

Achalasia may be mimic respiratory conditions. A diagnosis of achalasia should be borne in mind in patients with respiratory symptoms as it may readily be overlooked in x rays and lead to severe consequences including aspiration pneumonia. Therefore it should be particularly considered by pulmonologists in the differential diagnosis of younger patients, who are less likely to have aspiration.

REFERENCES

- O'Neill OM, Johnston BT, Coleman HG. Achalasia: a review of clinical diagnosis, epidemiology, treatment and outcomes. *World J Gastroenterol* 2013; 19(35):5806-12.
- Cole TJ, Turner MA. Manifestations of gastrointestinal disease on chest radiographs. *Radiographics* 1993;13(5):1013-34.
- Akritidis N, Gousis C, Dimos G, Paparounas K. Fever, cough, and bilateral lung infiltrates. Achalasia associated with aspiration pneumonia. *Chest* 2003;123(2):608-12.
- Dudley MT, Soubani AO. A patient with severe emphysema and aspiration pneumonia. *QJM* 2011; 104(10):899.
- Didilescu C, Dinu M. Respiratory manifestations in esophageal diseases. *Pneumologia* 2007;56(1):38-40.
- Robinson KM, Zorowitz RD. Mechanism of aspiration disorders. In: Elias JA, Fishman JA, Grippi MA, Robert M. Senior RM, eds. *Fishman's Pulmonary Diseases and Disorders*. Vol 1. 3rd ed. New York: McGraw-Hill, 1997: p.1211-24.
- Eckardt VF, Stauf B, Bernhard G. Chest pain in achalasia: patient characteristics and clinical course. *Gastroenterology* 1999;116(6): 1300-4.
- Hirata M, Kohno K, Murakami S, Yoneda S, Kajita Y, Fujita Y, et al. [A case of recurrent aspiration pneumonia by achalasia]. *Nihon Kokyuki Gakkai Zasshi* 2002;40(2): 149-53.
- Kendall AP, Lin E. Respiratory failure as presentation of achalasia of the oesophagus. *Anaesthesia* 1991;46(12):1039-40.
- Endlicher E, Fürst A, Strotzer M, Messmann H, Kerner T, Schölicher J, et al. ["Cardiomegaly and pneumonia"--a misdiagnosis in long-standing achalasia]. *Dtsch Med Wochenschr* 1999;124(13):386-90.