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A Rare Manifestation of Achalasia: Huge Esophagus Causing Tracheal Compression and Progressive Dyspnea

Akalazyanın Nadir Bir Manifestasyonu: Trakeal Kompresyon ve Progressif Dispneye Yol Açan Dev Özofagus

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Abstract

Achalasia is a primary esophageal motility disorder characterized by the absence of primary peristalsis and a failure of the lower esophageal sphincter to relax, resulting in a dilated esophagus. Dysphagia is the classic and most common symptom. Respiratory obstruction due to tracheal compression caused by a massively dilated esophagus is a very rare but fatal complication. Herein, we report a case of a patient with long-standing achalasia who had tracheal compression secondary to a markedly dilated, giant esophagus. These findings are documented with CT scans. His symptoms regressed after a Heller myotomy and fundoplication operation.

Key Words: Achalasia, esophageal expansion, computed tomography, dyspnea, progressive

Özet

Akalazya, özofagusta genişlemeye yol açan, alt özofageal sfinkterin gevşeme boukluğu ve özofagusta primer peristaltizm yokluğuyla karakterize özofageal motilite hastalığıdır.Disfaji en sık rastlanan yakınmadır. Massif olarak genişleyen özofagusun trakeaya basısı sonucu oluşan solunumsal obstrüksiyon, çok nadir ancak ölümcül bir komplikasyondur.Burada ileri derecede dilate, dev özofagusun yol açtığı trakeal kompresyonu olan uzun süredir akalazya tanısı alan olguyu sunuyoruz. Hastanın semptomları laparoskopik Heller myotomi operasyonu sonrasında gerilemiştir.Bulgular Bilgisayarlı Tomografi incelemesiyle dökümante edilmiştir.

Anahtar Kelimeler: Akalazya, özofagial genişleme, bilgisayarli tomografi, dispne, progressif

Introduction

Achalasia is a rare swallowing disorder that affects approximately 1 in every 100,000 people. It is caused by decreased numbers or an absence of ganglion cells in the Auerbach's plexus between the inner and outer muscle layers. This entity is characterized by the absence of primary peristalsis and a failure of the lower esophageal sphincter to relax, resulting in a dilated esophagus. It was first described and treated by physicians in the 17th century [1, 2].

The most common symptom is dysphagia that occurs with both solid and liquid food. Other symptoms include chest pain that mimics angina, which is aggravated after eating, regurgitation of food, cough, aspiration, pneumonia and weight loss. Rarely, tracheal compression caused by a severely dilated esophagus and dyspnea is observed [3-5]. Here, we describe imaging findings of this rare manifestation in a patient with a diagnosis of achalasia.

Case Report

A 50-year-old man was admitted to our hospital for progressive respiratory distress. He had suffered from dysphagia, and the diagnosis of achalasia was made 10 years earlier on the basis of his symptoms. He underwent balloon dilatation at an outside institution. His symptoms subsided for several years, but dysphagia and regurgitation reoccurred. His symptoms now also included dyspnea, which had steadily increased over two years. He had no significant family and/or past medical history.

On physical examination, the patient was noted to have visible distension of his cervical region on deep inspiration. He was also noted to have occasional wheezing and stridor. He was afebrile, and his neck bulged bilaterally at the level of the larynx. No thyroid masses were detectable. In addition to loud stridor, chest auscultation revealed decreased breath sounds bilaterally with an audible inspiratory and expiratory wheeze. The remainder of the physical examination was

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unremarkable. Laboratory results were within normal limits. Chest radiography showed pronounced dilatation of the esophagus in the upper thorax extending up to the neck, with a widened mediastinum (Figure 1). Both lung fields were clear. A barium sulfate contrast examination showed a large elongated esophagus extending to the thoracic inlet (Figure 2).

Computed tomography (CT) examination revealed a gross enlargement of the esophagus, which extended up into the lower neck, compressing the posterior tracheal wall and resulting in tracheal narrowing. The tracheal cross-sectional area at the most significant level of narrowing (tracheal inlet) was approximated by outlining the tracheal lumen on deep inspiratory images. A tracheal cross-sectional area of 0.59 mm^2 was measured at the level of the thoracic inlet (Figure 3a, b, c).

The patient underwent a balloon dilatation of the lower esophageal sphincter (LES) twice, after which his respiratory symptoms apparently improved. The patient did not respond to the pneumatic dilation. Therefore, the patient underwent surgery. Heller myotomy-fundoplication was performed, and there were no respiratory symptoms after surgery. A control CT scan of the tracheal area during deep inspirium was taken 1 year after the operation. A tracheal cross-sectional area of 1.81 mm^2 was measured at the level of the thoracic inlet. In our patient, we detected that the tracheal caliber was approximately 67% greater compared to CT examination taken before the surgery (Figure 3c, d).

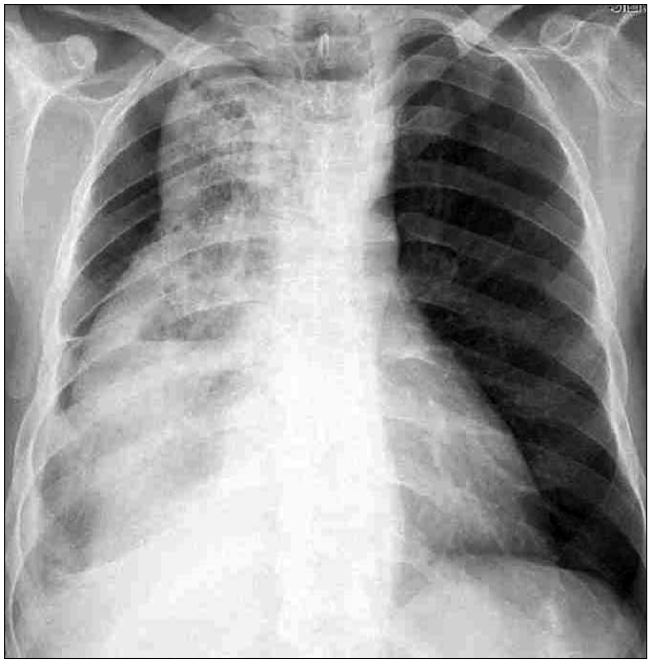

Figure 1. *Chest radiography shows a massively dilated esophagus filled with food residue and multiple air bubbles.*

Discussion

Achalasia is an idiopathic motility disorder of the esophagus. Respiratory findings of achalasia are often secondary to regurgitation and aspiration. After recurrent aspiration in these patients, pneumonia and bronchiectasis may occur. Airway obstruction from tracheal compression is a very rare but important complication of achalasia that can lead to respiratory compromise and even death [3].

The first case of achalasia presenting as an acute upper airway obstruction was reported in 1950 [2, 3]. The clinical symptoms and features of these cases have been similar. Most of the patients reported were older women who had had the diagnosis of achalasia for many years. In general, respiratory symptoms of achalasia occur after a meal. Massive dilatation of the esophagus from achalasia is responsible for airway obstruction and tracheal narrowing [3-6].

The exact cause of airway obstruction resulting from a dilated esophagus with resultant respiratory compromise in not known, but several hypotheses have been proposed in the literature. The first hypothesis is the pinch-cock valve

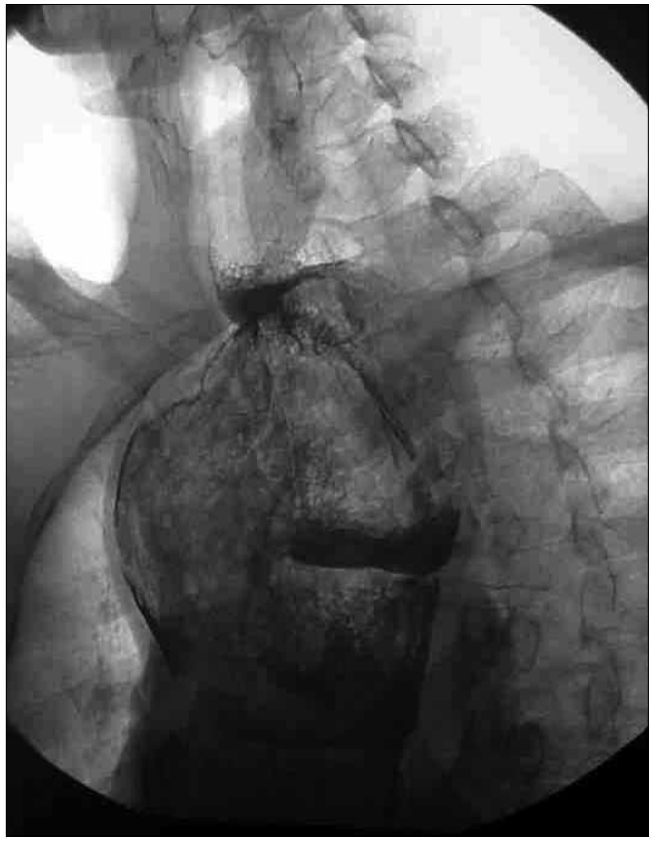

Figure 2. *Left anterior oblique image from a barium sulfate contrast esophagram illustrates a large elongated esophagus extending to the thoracic inlet.*

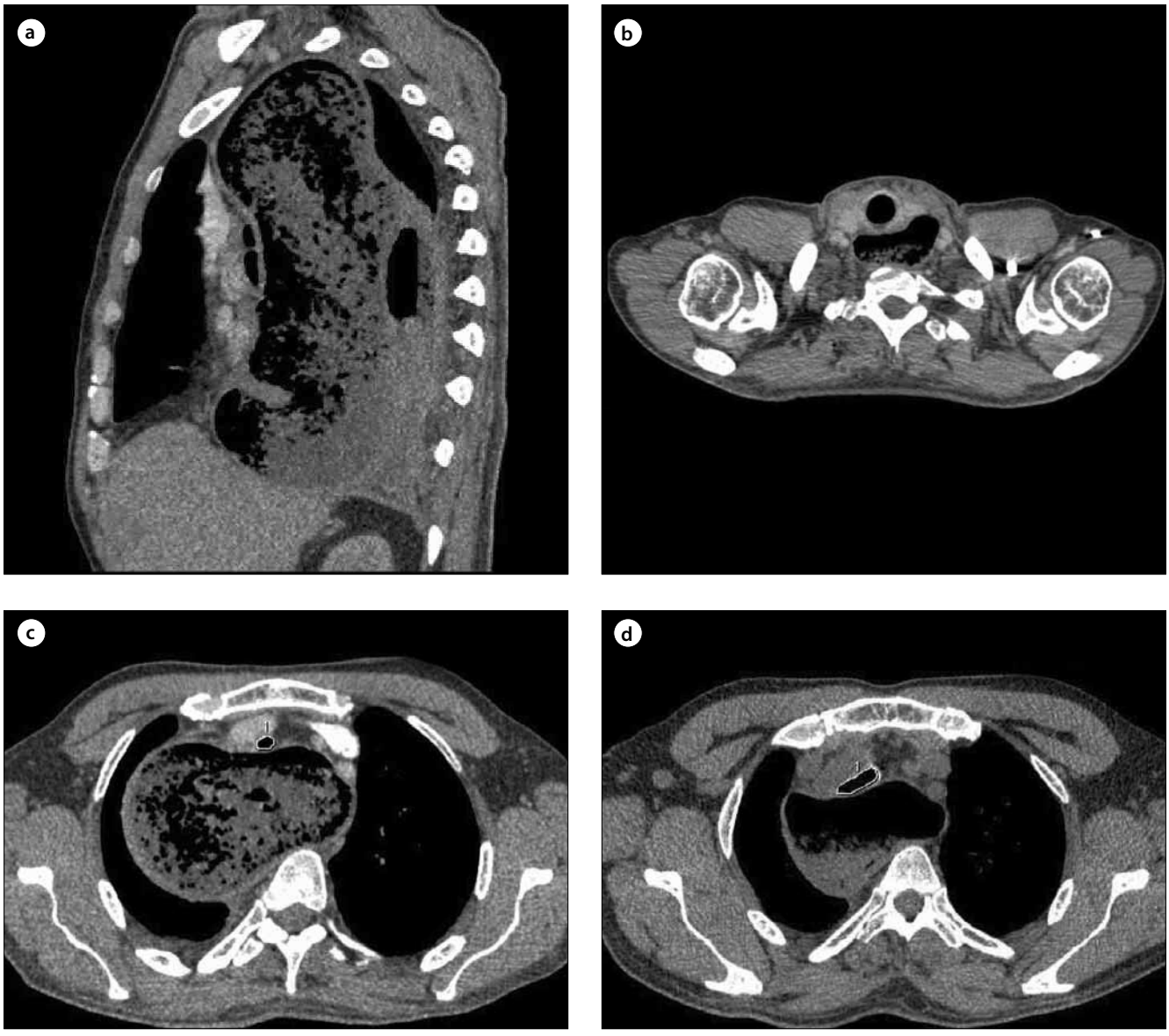

Figure 3. a-d. *Sagittally reformatted (a) and axial (b) CT images just below the level of the cricopharyngeus muscle shows dilation of the upper esophagus without evidence of tracheal compression. Axial CT images at the level of the thoracic inlet obtained before operation (c) and after operation (d) demonstrate that the tracheal area is 0.59 mm^2 before and is 1.81 mm^2 after operation. These two examinations were taken approximately at the same level and during deep inspiration. Note that the tracheal lumen becomes approximately 67% greater compared to before the surgery.*

theory, or cervical extension of an air-filled, massively dilated esophagus. The dilated esophagus becomes kinked behind the cricopharyngeus muscle, likely producing a one-way valve scenario; thus, swallowed air is unable to escape [7]. The second hypothesis is that the upper esophageal sphincter (UES) does not properly relax during swallowing. A high residual UES pressure does not allow air to escape and leads to distension of the esophagus. Physical compression of the upper airway by the dilated esophagus may also produce obstructive respiratory symptoms [8, 9]. The third hypothesis is the defective belch reflex. Spontaneous belch reflex occurs when the LES relaxes to allow the food bolus to pass into the stomach. In patients with achalasia, the UES also relaxes, allowing air to exit. Failure of this reflex, presumably secondary to achalasia, leads to progressive esophageal dilatation and airway obstruction [10]. The absence of a belch reflex in patients with a history of achalasia presenting with airway obstruction has been reported previously [10, 11]. It is gene-

rally observed along with an acute airway obstruction. Rapid diagnosis and emergency treatment is necessary for this potentially fatal condition. Early recognition is essential, as immediate decompression of the esophagus with nasogastric tube placement and drainage may prevent respiratory failure [12-14]. After stabilization, further definitive therapy for achalasia is rendered. Various treatment options are available for achalasia, including pharmacologic therapy with nitrates, calcium channel antagonists, balloon dilatation of the lower esophageal sphincter [15], injection of botulinum toxin (Botox) at the gastroesophageal junction, Heller's myotomy and esophagectomy, and cricopharyngeus myotomy [11, 16].

Decompression of the esophagus with nasogastric tube placement was not the preferred treatment because our patient had mild symptoms. He underwent pneumatic dilation twice but did not respond to this treatment. Therefore, Heller myotomy and fundoplication were performed. One year after surgery, the patient is now well, and respiratory symptoms have disappeared dramatically.

Massive dilation of the esophagus from achalasia leading to tracheal compression and respiratory distress, as in our case, has been reported in the clinical literature [17, 18]. Doshi et al. have reported the role of CT scans in the evaluation and treatment of patients with achalasia, demonstrating respiratory tract symptoms in the imaging literature [19]. In our patient, the CT study demonstrated increased compression of the trachea on inspiration. However, the post-operative control CT study demonstrated that tracheal compression significantly decreased. Pre-operative and post-operative tracheal cross-sectional areas were measured in the same plane and during deep inspiration. We detected that the tracheal caliber was approximately 67% greater compared to its value before the surgery.

In conclusion, most patients with achalasia can present with dysphagia, regurgitation, chest pain, and weight loss. However, upper airway obstruction resulting from tracheal compression by dilatation of the esophagus rarely occurs in achalasia. Tracheal compression manifested by stridor or respiratory distress should be quickly identified and treated appropriately. CT imaging may play an important role in these patients to quickly and easily evaluate the severity of tracheal compression, and it may also help to evaluate the benefits of surgical treatment.

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Informed Consent: Written informed consent was obtained from patients who participated in this case.

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