Acute Airway Obstruction in a Patient with Achalasia

Shin’ichi Miyamoto, Yoshitaka Konda, Masashi Matsui, Kazuya Sawada, Kazuki Ikeda,
Norihiko Watanabe, Chiharu Kawanami and Tsutomu Chiba

Abstract

Megaesophagus resulting from achalasia is a rare but serious cause of acute airway obstruction. We treated achalasia in a 52-year-old woman with acute respiratory distress and stridor. Chest X-ray and endoscopy showed a marked dilatation of the cervical esophagus with a large amount of undigested food. Emergency suction of the food through a nasogastric tube led to decompression of the esophagus and the immediate relief of respiratory symptoms. These findings suggest a dysfunction of the upper esophageal sphincter as a possible mechanism. As this exceptional complication of achalasia is fatal, a wider appreciation is required.

Key words: esophageal achalasia, acute airway obstruction, upper esophageal sphincter

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Introduction

Achalasia is a motility disorder caused by denervation of the esophagus leading to failure of peristalsis, raised pressure in the lower esophageal sphincter (LES), and incomplete relaxation of the LES during swallowing (1-3). Dysphagia is the classic and most common symptom. Other forms of presentation include weight loss, heartburn, nocturnal regurgitation, pneumonia, asthma, and chest pain. Respiratory obstruction due to tracheal compression caused by a massively dilated esophagus is a very rare but fatal complication. The pathophysiology of achalasia leading to acute airway obstruction has not been clearly defined, but the inability to swallow and the failure of the upper esophageal sphincter (UES) to relax are thought to contribute to esophageal distension. Swallowed foods can trap air in the esophagus, which can lead to rapidly worsening respiratory obstruction. In this paper we discuss the probable mechanism for this rare complication of esophageal achalasia and its management.

Case Report

A 52-year-old woman was transported from a nearby sushi restaurant to the emergency ward in acute respiratory distress. Her symptoms began with the acute onset of stridor and increased difficulty breathing immediately after eating sushi. She had a 12-year history of untreated esophageal achalasia and had noticed difficulty in belching for a long time. She had no previous history of respiratory or cardiac disease. On physical examination she was afebrile but diaphoretic and her neck bulged at the level of the larynx bilaterally. No thyroid masses were detectable. In addition to loud stridor, chest auscultation revealed decreased breath sounds bilaterally with an audible inspiratory and expiratory wheeze. Laboratory investigations, including blood gas data, revealed no abnormal findings. The electrocardiogram was normal. An asthma attack was suspected initially, and she was given 500 mg of methylprednisolone intravenously and 0.3 mg of epinephrine subcutaneously, but her symptoms did not change. Chest radiography showed pronounced dilatation of the esophagus in the upper thorax extending up to the neck, with a widened mediastinum (Fig. 1). Both lung fields were clear. Emergency esophagogastroduodenoscopy (EGD) showed marked dilatation of the cervical esophagus (Fig. 2), but she could not tolerate further examination because of severe dyspnea. The supraglottic airway and motility of the vocal cords were normal. A large amount of undigested food and saliva were sucked from the esophagus through the nasogastric tube, resulting in decompression of the esophagus and immediate relief of her stridor and dyspnea. She was admitted to our hospital without tracheal intubation. The same symptom occurred twice during her first
Figure 1. Chest X-ray on admission showing striking dilatation of the upper esophagus with a widened mediastinum.

Figure 2. Upper gastrointestinal endoscopy in the emergency room demonstrating the massively dilated lumen of the cervical esophagus.

Figure 3. Gastrograffin swallow showed a tortuous sigmoid-shaped esophagus with a large amount of food residue, which is typical for achalasia.

The cricopharyngeal muscle, of approximately 1 cm in improved. Computed tomography of the chest also showed the extremely dilated esophagus located adjacent to the membranous part of the trachea (Fig. 4). From these findings we postulated that dysfunction of the UES played a crucial role in the development of her respiratory symptoms. She was discharged on the 28th day of hospitalization and is currently being followed up in the outpatient clinic at our hospital. Upper airway obstruction has not recurred in the 10 years since her presentation, although she still has mild dysphagia.

Discussion

Achalasia presenting as an acute upper airway obstruction was first reported by Bello et al (4) in 1950, and nearly 40 cases have been described in the literature since then. The clinical features of these cases have been strikingly similar. Most of the patients have been elderly women, with the onset of symptoms often occurring after a meal. Chest radiographs are easy to do and important for the diagnosis. The dilated esophagus is presented as a widened mediastinum, often with a characteristic mottled appearance or air fluid level on posterior-anterior view, which compresses and displaces the trachea forward on lateral view.

Prompt diagnosis and emergency treatment are necessary for this fatal condition. Air and saliva must be evacuated to decompress the megaesophagus; immediate insertion of a nasogastric tube into the esophagus is the most convenient and effective method (5-8). Transcutaneous needle aspiration of the distended esophagus (presented as a soft mass of the neck) is another possible method of emergency evacuation (9). Pharmacological management of this condition has been universally disappointing. However, because administration of sublingual glyceryl trinitrate is simple and familiar, it should be tried once (10).

The cricopharyngeal muscle, of approximately 1 cm in
length, is a dominant part of the UES, which measures between 2 and 4 cm. Swallowing depends on fine coordination between UES relaxation and pharyngeal contractions, which forces food through the sphincter into the esophagus. A number of abnormalities in UES function have also been described in achalasia, including: (a) elevated UES residual pressure (11, 12), that is, the difference between the pressure recorded at the nadir of UES relaxation and baseline pressure, (b) decreased duration of UES relaxation (13, 14), (c) repetitive UES contractions (15), and (d) a loss of belch reflex with normal deglutitive UES relaxation (16, 17). The present patient had experienced difficulty in belching for a long time and her symptoms were consistent with failure of the belch reflex caused by insufficient UES relaxation, which led to progressive esophageal distension. Interestingly, Yoneyama et al (12) reported that the increased UES residual pressure in achalasia patients decreased significantly after successful LES dilatation. In fact, pneumatic dilatation of the LES was effective in relieving the present patient’s respiratory symptoms. Unfortunately, it is impossible to accurately measure the pressure of UES with our pneumohydraulic pull-through manometer. Another method, such as the multichannel intraluminal impedance technique, may be used to analyze the detailed function of the UES (18, 19). Previous reports suggest that cricopharyngeal myotomy is a logical, well-tolerated procedure that permanently relieves symptoms for this complication (20, 21). If respiratory failure is recurrent, this procedure should be the treatment of choice. Tracheal stenting is an alternative palliative treatment in patients who are unfit for cricopharyngeal myotomy (22).

In conclusion, acute upper airway obstruction resulting from tracheal compression by dilatation of the esophagus may occur in achalasia cases because of UES dysfunction. As any delay in diagnosing this exceptional condition is critical, a wider appreciation of this complication is required. Immediate insertion of a nasogastric tube into the esophagus can lead to prompt decompression of the megaesophagus and marked improvement of respiratory symptoms.

The authors state that they have no Conflict of Interest (COI).

References


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