## Case Report

# Achalasia Esophagus; presenting as acute air way obstruction

Anwaar A. Khan, S. Waqar H. Shah, Altaf Alam, Arshad K. Butt, F. Shafqat, K. Malik, J. Amin
Department of Gastroenterology-Hepatology, Sheikh Zayed Postgraduate Medical Institute, Lahore.

## Abstract

Acute airway obstruction from mega-esophagus is a rare presentation of achalasia. Mega-esophagus is generally considered when the transverse width of the esophagus is more than 7 cm. A 78 year old lady presented with long-standing history of productive cough and nocturnal dyspnoea. She was seen in the emergency department with acute exacerbation of dyspnoea after a bout of vomiting, containing semi-solid food with foetid smell. Her respiratory status deteriorated rapidly with onset of stridor, and cyanosis. Chest x-ray showed widening of mediastinum due to dilated esophagus with air-fluid level. Prompt, repeated, upper respiratory tract suction was carried out. A wide bore nasogastric tube was introduced, esophagus was decompressed with a gush of air and fluid, relieving the respiratory distress.

This case illustrates an unusual presentation of achalasia underscoring the need for urgent, life-saving esophageal decompression. Hypotheses, regarding the mechanism of airway compromise, as well as, treatment options are reviewed.

## Introduction

Achalasia is an esophageal motor disorder with aperistalsis of the esophagus and impaired lower esophageal sphincter (LES) relaxation.[1] Acute airway compromise is a rare presentation of "mega-esophagus" due to achalasia described earlier.[2] This patient presented with acute airway obstruction from a massively dilated esophagus, not previously diagnosed as achalasia. She presented with severe respiratory distress, immediate decompression with a nasogastric tube offered prompt relief of symptoms. This case highlights a rare complication and emphasizes the need for urgent decompression of the esophagus.

## Case Report

A 78 year old, retired school teacher, presented at the emergency department of the Shaikh Zayed teaching hospital, Lahore with 30 minute history of productive cough, severe shortness of breath and chest discomfort. She vomited on her way to the hospital.

She also had eight year history of weight loss, weakness, chronic productive cough, intermittent low grade fever, often relieved with antibiotics. She denied history of dysphagia.

Physical examination showed, an anxious elderly lady with temperature 101$^{o}$F, blood pressure 145/90 mmHg, pulse rate 115/minute regular, respiratory rate 28 /minute, and oxygen saturation 52% on room air, with evidence of peripheral cyanosis. Auscultation of chest revealed few scattered low pitched wheezes and coarse crepitations at right lung base. Abdominal, cardiac and rest of the physical examination was unremarkable. Chest x-ray revealed massively dilated esophagus extending up to neck along the right cardiac border (Figure 1). Laboratory reports showed Total leucocyte count (TLC) of 18.2 x10$^{9}$/l, with 98% polymorphs. Rest of the lab reports, including liver function tests, BUN, creatinine, cardiac enzymes and ECG were normal.

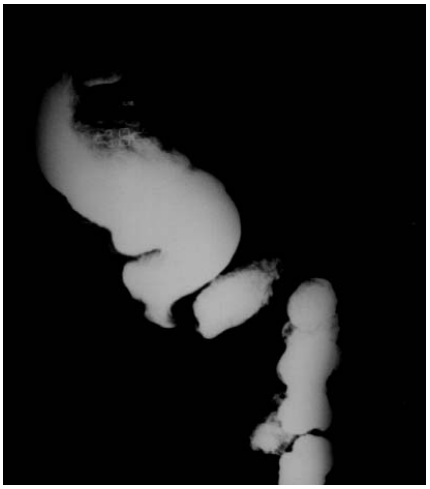

Figure 1. Chest X-ray showing dilated esophagus extending along the right cardiac border in to neck.

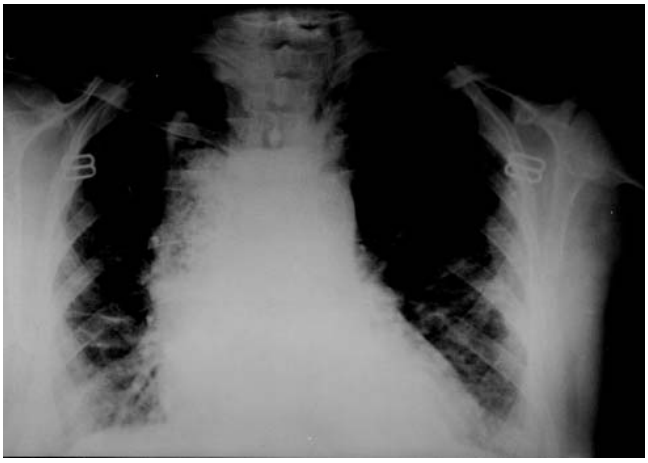

Figure 2. Massively dilated sigmoid esophagus in achalasia.

Tentative diagnosis of mega-esophagus due to achalasia, chronic aspiration, and acute airway obstruction from aspiration was made. A wide bore nasogastric tube was passed which produced an audible "gush of air" with prompt relief of stridor and dyspnoea. Repeated upper respiratory tract suction further relieved respiratory distress. She was started on oxygen 3-4 liter /minute, ceftriaxone 1 gm intravenously 8 hourly with chest physiotherapy. On further inquiry, she denied having dysphagia, however, she had regurgitation of semi-solid food, with 10 kg weight loss in 6 months. A thin barium swallow showed dilated and tortuous "sigmoid esophagus" due to achalasia (Figure 2).

Achalasia was subsequently, confirmed on esophageal manometry. She was successfully dilated with Microvasive, Rigiflex, 35 mm balloon as previously described.[3] All her symptoms markedly improved after balloon dilatation. A follow up visit at one month, 6 months and 1 year intervals did not show recurrence of symptoms and she gained weight.

## Discussion

Achalasia is an idiopathic motility disorder of the esophagus with cardinal symptoms of dysphagia for solids and liquids, a feature invariably present. In addition, weight loss, regurgitation and respiratory complications occur due to regurgitation of food. This may result in aspiration, asthma like presentation, pneumonia and rarely, lung abscess. Acute airway obstruction is a rare presentation of achalasia, first described by Bello and colleagues in 1950[4], where "Mega-esophagus" secondary to achalasia was thought to have caused direct tracheal compression and airway compromise.

Exact cause of distension of the esophagus with resultant respiratory compromise in not known, but various hypotheses have been proposed. Firstly, it is thought that cervical extension of an air-filled, distended esophagus may get kinked behind the circopharyngeus muscle, producing a likely one way valve situation, thus; swallowed air is unable to escape. Secondly, a high residual upper esophageal sphincter pressure, not yielding to release air, leads to balloon-like distension of the esophagus. Physical compression of the upper airway by the dilated esophagus, may also produce obstructive symptoms.[5] Thirdly, the belch-reflex may be abolished due to chronic regurgitation of the esophageal contents, which often occurs in meg-esophagus, thus; aggravating respiratory symptoms. Physiologically, distension of the esophagus produces belch reflex by relaxation of the upper esophageal sphincter with resultant decompression of the esophagus. The belch-reflex may be lost in Achalasia. Lastly, there may be diminished cough reflex in old age and in prolonged esophageal stasis, resulting in silent aspiration of liquids.

Prompt recognition of this rare manifestation of achalasia is critical, inasmuch as, urgent decompression of the esophagus is life-saving and must be carried out by insertion of wide bore nasogastric tube, besides repeated suctioning to clear upper respiratory tract. After stabilization, further definitive therapy for achalasia is rendered.[6] Various treatment options are available in achalasia, i.e. pharmacologic therapy with nitrates, calcium channel antagonists, balloon dilatation of the lower esophageal sphincter[7], injection of botulinum toxin (Botox) at the gastroesophageal junction, Heller's myotomy and esophagectomy.[8] Recently, circopharyngeus myotomy was described in achalasia producing airway obstruction.[9-10] This however, may not be needed if drainage of the esophagus is established by any of the above mentioned procedures. Definitive treatment for achalasia obviates recurrence of above symptoms, as in our patient.

## Conclusion

This case demonstrates a rare but serious complication of achalasia. Acute respiratory compromise resulting from aspiration of esophageal contents during vomiting, resulted due to meg-esophagus. Recognition of this condition and urgent treatment by esophageal decompression was successful in relieving her acute symptoms. Subsequent definitive treatment with balloon dilatation, further relieved her symptoms

## Reffrence


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