

Primary Achalasia with Pneumoesophagus and Bronchopleural Fistula Causing Right Lung Collapse and Bronchiectasis

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CLINICAL SUMMARY

A 35-year-old male non-smoker, presented with a history of recurrent respiratory tract infections, associated with intermittent haemoptysis and haematemesis since childhood. His vital parameters were normal with pulse oximetry saturation of 97% on room air. On physical examination, clubbing was observed. Chest auscultation revealed tubular type of bronchial breath sounds over the right hemithorax.

INVESTIGATIONS

Haemogram and serum chemistry values were in the normal range. Plain chest radiograph (Figure 1) showed a peripheral right lung collapse with cystic



Figure 1. Chest radiograph (postero-anterior view) showing peripheral right lung collapse with cystic opacities and a lucent area medial to the collapsed lung.

lucent areas and a lucent area medial to the collapsed lung. Oral contrast enhanced computed tomography (CT) of thorax showed a dilated thoracic segment of the oesophagus that was air-filled (pneumoesophagus/mega-aeroesophagus). Additionally, there was a right lung collapse with cystic bronchiectasis. Barium oesophagogram (Figure 2) showed dilated oesophagus with pooling



Figure 2. Barium oesophagogram showing dilated thoracic segment of oesophagus that was air-filled (pneumoesophagus/mega-aeroesophagus).

of contrast. Minimum phase intensity reconstruction axial and coronal images of the CT (Figure 3) revealed bronchoesophageal fistulae (arrows). Fibreoptic bronchoscopy showed dilated main and sub segmental bronchi on the right side. Oesophagoscopy showed a dilated oesophagus in its entire length with an oesophago-bronchial fistula at 34cm from the lower incisors. Oesophageal manometry showed absence of peristalsis in distal oesophagus.

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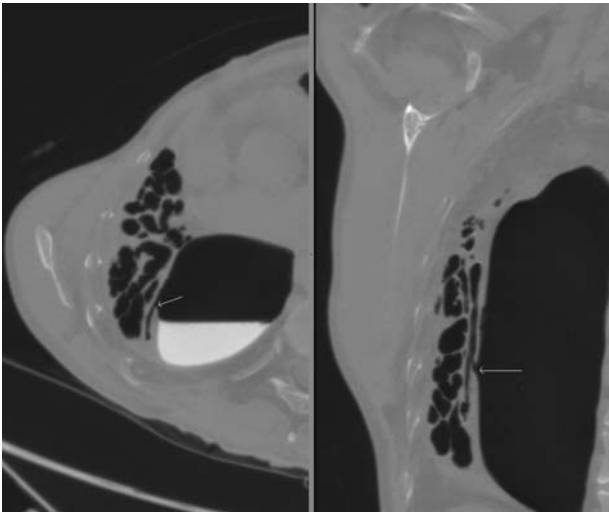


Figure 3. Minimum phase intensity reconstruction axial and coronal images of the CT showing bronchoesophageal fistulae (arrows).

DIAGNOSIS

Primary achalasia with pneumoesophagus/mega-aeroesophagus and bronchoesophageal fistula causing a peripheral right lung collapse and bronchiectasis.

DISCUSSION

Primary achalasia is a motor disorder of the oesophagus of unknown aetiology with loss of inhibitory neurons in the oesophageal myenteric plexus. It is characterised by absent primary peristalsis and incomplete relaxation of the lower oesophageal sphincter during the act of deglutition.¹ Achalasia when long-standing results in abnormally dilated oesophagus known as megaesophagus or pneumoesophagus/mega-aeroesophagus if air-filled.² The exact aetiology of loss of oesophageal myenteric plexus inhibitory neurons still remains unknown.³ The disease can manifest at any age. Clinical manifestations include dysphagia, chest pain, heartburn, regurgitation, weight loss, haematemesis and aspiration. Chronic aspiration from dilated oesophagus in achalasia or an abnormal persistent fistulous communication between oesophagus and respiratory tract may result in infections causing recurrent trauma to the airways and consequent bronchiectasis.⁴

The diagnosis of achalasia cardia is mainly by clinical history, radiography and oesophageal manometry. Chest radiograph findings in achalasia include mediastinal widening due to dilated oesophagus, air-filled or with air fluid level and absent gastric air bubble.⁵ The barium oesophagogram shows narrowing at the gastro-

oesophageal junction, with smooth tapering of the lower oesophagus resembling a Bird's beak appearance.⁶ Long-standing cases show a dilated, tortuous oesophagus known as sigmoid oesophagus with pooling of contrast in the oesophagus.⁷ Oesophageal manometry shows lack of peristalsis in the body of oesophagus. Lower oesophageal sphincter shows a high resting pressure and fails to relax, or shows partial relaxation during the act of deglutition.^{8,9}

The treatment of achalasia cardia is mainly palliative and includes pharmacological treatment with drugs that include organic nitrates or calcium antagonists. These have a transient effect with high failure rates. Endoscopic treatment includes injection of botulinum toxin into the lower oesophageal sphincter, injection of sclerosant and balloon dilatation. Surgical treatment has high success rate and involves longitudinal division of musculature of distal oesophagus (Heller's myotomy).¹⁰

In our case, primary achalasia cardia was complicated by congenital tracheoesophageal fistulae. Right lung collapse with cystic bronchiectasis probably developed as a result of aspiration and repeated respiratory tract infections due to the long-standing achalasia cardia and tracheoesophageal fistulae. Congenital tracheoesophageal fistula complicated by achalasia in an adult is rare but has been reported previously.¹¹ Lobar collapse with bronchiectasis in achalasia with megaesophagus has also been reported previously.⁴ The radiographic picture in our case was unusual; the collapsed right lung caused a peripheral opacity with a medial lucency due to pneumoesophagus/mega-aeroesophagus.

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