Clinical picture

Megaoesophagus: an unusual cause of stridor

An 81-year old female presented with a long history of dysphagia and dyspnoea exacerbated by eating. On examination, there was marked stridor but no other significant findings. Pulmonary function tests showed FEV1 0.67 (44% predicted) and FVC 1.01 (53% predicted) with expiratory loops suggestive of intrathoracic obstruction.

Chest radiography (Figure 1) revealed an air-filled structure projected over the right of the mediastinum. Computed tomography (Figure 2) showed a tortuous and massively dilated oesophagus. Tracheomalacia combined with extrinsic compression from the dilated oesophagus reduced the tracheal lumen to 2 mm at the level of the manubro sternum. Figure 3 shows 3D surface-shaded reconstructions of the oesophageal lumen. The oesophagus is seen within the bony thorax (A) and alongside the lumen of the large airways (B).

Contrast swallow demonstrated absent oesophageal motility with the barium cascading down the oesophagus by gravity only and pooling at the lower oesophageal sphincter. The diagnosis of likely end-stage achalasia was therefore made.

Endoscopic and radiologic attempts to reach the gastro-oesophageal junction failed. The patient has declined further investigation.

Achalasia is a primary oesophageal motility disorder characterized by absence of oesophageal peristalsis in the distal two-thirds and impaired lower oesophageal sphincter relaxation.1 Manifesting between the ages of 25 and 60, men and women are affected equally. The aetiology is unknown.2

Long-term treatments include pneumatic dilatation and Heller myotomy (incising the circular muscle of the lower oesophageal sphincter). Pharmacological therapies that reduce lower oesophageal sphincter pressure, such as calcium channel blockers and nitrates, may be useful. Botulinum toxin injections delivered endoscopically have shown short-term efficacy.3

Figure 1. PA chest radiograph. A seemingly lobulated air-filled structure can be seen projected over the right side of the heart, the right hilum and centrally within the superior mediastinum.

Figure 2. Computed tomography image. A slice taken at the level of the manubrosternum displayed using ‘lung windows’ to highlight the respiratory and GI tract lumens. The oesophagus is massively dilated and compressing the trachea against the upper mediastinal vessels anteriorly. The tracheal lumen measures 2 mm in AP diameter at this level.
Achalasia, if left untreated, causes stasis and rarely megaesophagus, of which airway obstruction is a rare but potentially fatal complication.

The combination of long-standing dysphagia and dyspnoea should raise achalasia/megaesophagus as a possible differential, particularly if the dysphagia predates the breathlessness.

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References