Case report

Achalasia presenting as stridor

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Learning Point for Clinicians

Achalasia is the failure of the relaxation of the lower oesophageal sphincter leading to oesophageal dilatation. Stridor is a rare and unusual symptom of achalasia due to tracheal compression by the dilated oesophagus. Early identification and management with urgent decompression should be undertaken to avoid life-threatening airway obstruction.

Case history

A 93-year-old woman presented to the emergency department with acute-onset dyspnoea and stridor following a bout of coughing, which had started just after finishing her evening meal. Her past medical history included dementia, stroke, asthma, hypertension and peptic ulcer disease.

On examination, the patient had stridor but was able to complete sentences. Observations showed oxygen saturations of 95% on air, pulse rate of 100 beats per minute, blood pressure of 136/96 mmHg, respiratory rate of 20 breaths per minute and temperature of 36.4°C. The trachea was central and the patient had symmetrical expansion of the chest during inspiration. Auscultation revealed transmitted upper airway sounds and marked wheeze. Despite the stridor and dyspnoea, she was comfortable and able to converse well.

She was immediately treated in the emergency department with heliox and parenteral hyoscine butylbromide and metoclopramide. She was kept nil-by-mouth. The ENT team performed a flexible nasoendoscopy which was unremarkable showing no supraglottic obstruction or food bolus.

Routine biochemical and haematological blood tests were within normal parameters. Chest radiograph revealed right basal consolidation. CT scan of the thorax and upper abdomen revealed a markedly dilated oesophagus filled with food debris. The trachea was alarmingly narrowed, with the narrowest tracheal diameter measuring just 2 mm, secondary to oesophageal compression (Figure 1).

Although the stridor persisted, the patient remained clinically well and stable. An oesophagogastroduodenoscopy done subsequently revealed no structural abnormalities down to the duodenum. However, large amount of chronic food debris amassed in the oesophagus was found. This was pushed through into the stomach, bringing the patient instant symptomatic relief.

Further evaluation of the patient’s swallow and oesophageal motility was undertaken with a barium swallow study, which showed a moderate hiatus hernia, a tortuous and dilated oesophagus and impaired motility of the gastro-oesophageal junction.

The patient was diagnosed with achalasia, causing chronic oesophageal food debris and marked dilation of the oesophagus with secondary...
compression of the trachea. There was no further recurrence of stridor and she was discharged on domperidone to assist with gastric motility and outpatient follow up was arranged with the Gastroenterology department.

Discussion

Achalasia is the failure of the relaxation of the lower oesophageal sphincter (cardiac sphincter) and failure of peristalsis of the oesophagus on swallowing. Failure of the sphincter to relax causes a functional oesophageal stricture leading to a dilated oesophagus. Symptoms usually include intermittent dysphagia and retrosternal pain following meals. Over time, the oesophagus may distend grossly. Stridor is a rare and unusual symptom of achalasia, which has been previously documented. Such patients usually present following rapid deterioration secondary to airway compromise. Instant treatment should be instituted to decompress the oesophagus. This can be achieved by passing a nasogastric tube or by rigid oesophagoscopy if available acutely.

Two hypotheses have been proposed for the cause of stridor in achalasia. The first proposes that the oesophageal dilation resulting from worsening achalasia extends superiorly, which causes the oesophagus to kink and wedge behind the cricopharyngeus muscle. This creates a one-way valve for air to enter the increasingly dilated oesophagus but not escape, thus causing pressure anteriorly on the trachea. An alternative hypothesis suggests maintenance of high pressure at the upper oesophageal sphincter (cricopharyngeus) following swallowing and failure of the belch reflex leading to increasing volumes of air being trapped in the oesophagus, resulting in oesophageal distension and tracheal compression against the sternum. In both circumstances, airway narrowing causes stridor with reduced airflow and respiratory distress.

Although achalasia and oesophageal distension presenting as stridor and airway compromise are rare, early identification and management with urgent decompression should be undertaken to avoid life-threatening airway obstruction.

Conflict of interest: None declared.

References