CASE REPORT

Unusual case of respiratory embarrassment secondary to tracheal compression by a dilated oesophagus in a patient with recurrent achalasia

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SUMMARY

We present the case of a 79-year-old woman with recurrent achalasia following a laparoscopic Heller's cardiomyotomy. The patient presented to the emergency department, with epigastric pain, severe dyspnoea and profound respiratory acidosis. She required intubation and ventilation followed by gastric decompression with nasogastric tube and the administration of intravenous antibiotics for a lower respiratory tract infection. Once stable, she underwent a CT scan revealing a massively dilated oesophagus causing marked tracheal compression. She received a period of continuous positive airway pressure ventilation while on the intensive care unit, for persistent low saturations, however, this was promptly ceased due to exacerbation of gastric dilation and fears over perforation. The patient responded well to conservative measures and was discharged home 18 days later awaiting follow-up with operating consultant surgeon.

BACKGROUND

Achalasia is a motility disorder of the oesophagus, characterised by high lower oesophageal sphincter resting tone and impaired oesophageal peristalsis. It is a relatively rare condition affecting around 1 in 100 000.¹ The aetiology and pathogenesis are not well understood but are thought to involve neurodegeneration of inhibitory neurons within the myenteric plexus controlling lower oesophageal sphincter contraction. Symptoms include dysphagia, regurgitation and weight loss. Respiratory symptoms are a recognised complication of achalasia, in part due to failure of oesophageal emptying, increasing the risk of aspiration of food content into the lungs. However, tracheal compression by a massively dilated oesophagus is particularly rare, especially in patients who have undertaken surgical management in the form of a Heller's cardiomyotomy. We present such a case and highlight the role of prompt decompression and radiological imaging in managing these patients.

CASE PRESENTATION

A 79-year-old woman was admitted to the emergency department by ambulance, with epigastric pain, abdominal distension, nausea, fever and dyspnoea. The patient reported general malaise, with progressive dysphagia and dyspnoea in the 2 weeks preceding presentation, therefore her family doctor had started oral antibiotics for a presumed lower respiratory tract infection.

Medical history included hypothyroidism and hypertension. Surgical history included hysterectomy and a laparoscopic Heller's cardiomyotomy for manometry-confirmed achalasia performed 9 months earlier. Initial diagnosis of achalasia was made 4 years prior after investigation of symptoms of dysphagia and weight loss with oesophagoduodenoscopy, oesophageal pH manometry studies and barium swallow.

On physical examination, it was noted that the patient had a visibly distended neck; there was inspiratory and expiratory wheeze throughout the lung fields bilaterally; decreased air entry at the right base on chest auscultation and a distended but soft non-tender abdomen with stool in the rectum on digital rectal examination. Initial observations showed a tachycardia of 107 bpm, tachypnoea of 28 bpm, temperature of 37.6°C and oxygen saturations of 94% on room air, Glasgow Coma Scale 15/15. However, during the assessment phase in the emergency department, the patient became drowsy with poor respiratory effort and increasing respiratory acidosis, subsequently, the decision was made by the intensive care physician to intubate, and ventilate and transfer to the intensive care unit (ICU).

Full blood count showed a haemoglobin level of 130 g/L, white cell count raised at $21.7 \times 10^9/L$ and platelet count $352 \times 10^9/L$. Arterial blood gas sampling taken with the patient receiving 5 L/min of oxygen through a Hudson mask revealed a respiratory acidosis with pH of 6.95, pCO_2 20.21 KPa, pO_2 19.23 KPa, bicarbonate 20.8 mmol/L, base excess -4.5 mmol/L and lactate of 1.69 mmol/L.

INVESTIGATIONS

A chest radiograph organised by the family doctor 3 days prior to admission revealed a dilated oesophagus in keeping with achalasia and segmental collapse in the right lower lobe. Similarly, repeat chest and abdominal radiograph (figure 1) performed on presentation demonstrated worsening dilation of the oesophagus and the presence of a distended stomach and small bowel.

A CT scan of the chest, abdomen and pelvis with intravenous contrast (figure 2A–D) was performed once the patient was stabilised. The CT scan revealed gross dilation of the hypopharynx merging into a grossly dilated oesophagus tapering down to a tight gastro-oesophageal junction; a grossly distended stomach; dilated small bowel loops, indicative of ileus, and right lower lobe

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Figure 1 Anteroposterior supine chest X-ray revealing massively dilated oesophagus extending into neck and distended stomach due to ileus.

collapse. On cross-sectional imaging, the dilated oesophagus could be seen compressing the trachea most significantly at the level of the manubrium, with a trachea cross-sectional area of 27.78 mm^2 . Previous CT imaging studies have shown the average cross-sectional area of the female trachea is $250\text{--}350 \text{ mm}^2$.²

A barium swallow performed when the patient's clinical situation improved showed a dilated oesophagus with tapering down to the normally positioned gastro-oesophageal junction in keeping with achalasia (figure 3).

DIFFERENTIAL DIAGNOSIS

Obstruction of the trachea has numerous causes including malignancy, foreign body, anaphylaxis and trauma. The medical history of recent antiachalasia surgery and radiological evidence

made tracheal compression secondary to a dilated oesophagus the most likely diagnosis.

TREATMENT

Deterioration in the patient's conscious levels, decreased respiratory effort and worsening respiratory acidosis, prompted swift intubation and ventilation with rapid sequence induction in the resuscitation area of the emergency department. Initial attempts at intubation were unsuccessful due to the abnormal airway anatomy. A bougie was used to assist the placement of an endotracheal tube and, subsequently, a 16 French Ryle's tube was inserted under direct vision and a large volume of gastric contents suctioned off. The patient was transferred to the ICU, sedated and started on intravenous tazocin and metronidazole for 7 days. Several days after admission onto ICU, the patient was extubated, after which her saturations dropped. A trial of continuous positive airway pressure (CPAP) was started but quickly ceased due to a deleterious effect on the patient's respiratory function, presumably due to increased oesophageal dilation. Total parenteral nutrition was started on day 2 and continued for a further 10 days with gradual introduction of a soft diet under dietician supervision.

OUTCOME AND FOLLOW-UP

The patient was extubated on day 3, spent 9 days in ICU and a further 8 days on the general surgery ward. She made a full and uneventful recovery with intravenous antibiotics, gastric decompression, total parenteral nutrition and respiratory support. She was followed up in outpatient clinic 2 weeks later when she reported occasional ongoing abdominal bloating, but was much improved and tolerating a solid diet. She remains under regular six monthly review with the option of further intervention in the form of pneumatic dilation if needed.

Figure 2 (A) Coronal CT image lung window revealing massively dilated oesophagus. (B) Sagittal CT image lung window revealing dilated oesophagus with compression of trachea (red arrow) at the level of the manubrium. (C) Axial CT image lung window revealing trachea in red and oesophagus in green; level of greatest tracheal narrowing at the level of the manubrium. (D) Axial CT image lung window formatted at the level of the manubrium, trachea in red revealing cross-sectional area of 27.78 mm^2 .

Figure 3 Barium swallow of lower oesophagus revealing 'bird-beak' sign.

DISCUSSION

Respiratory symptoms are relatively common in patients with achalasia, with as many as 40% of patients reporting at least one respiratory symptom daily.³ Aspiration pneumonia, pneumonitis and lung abscesses can occur, and the mechanism responsible is retained food products within the oesophagus aspirating into the lungs, particularly at night when patients are supine.⁴ Despite respiratory symptoms being relatively common, tracheal compression with resulting respiratory compromise is very rare, particularly in patients who have previously undergone cardiomyotomy. The first reported case of achalasia and tracheal compression was in the 1950s,⁵ since then, there have been sporadic cases reported in the literature. Similarities between cases include preponderance to female sex, old age and symptoms arising postprandially. Deaths have been reported with varying mechanisms suggested including cardiac arrhythmia, respiratory compromise and aspiration pneumonia.^{6 7} There have been no documented cases of tracheal compression in patients with recurrent achalasia following laparoscopic Heller's cardiomyotomy surgery.

Rapid diagnosis is essential in the management of these patients and usually takes the form of a chest radiograph followed by a CT scan if the patient is stable. Early recognition allows for early management and the key treatment strategies are respiratory support, intubation in this case, and decompression of the gastrointestinal system by placement of a nasogastric or Ryle's tube. In newly diagnosed achalasia or treatment-naïve achalasia, definitive treatment may be needed to reduce oesophageal dilation, of which there are many options. Medical therapies include calcium channel blockers and nitrates aimed at reducing chronic contraction at the lower oesophageal sphincter, however, studies so far have shown limited efficacy.⁸ Injection of botulinum toxin A has been shown to have better, if transient, benefits, particularly in the patient subset considered unfit for surgery. Pneumatic dilation confers the best symptom relief among the non-surgical options, with good-to-excellent symptom relief in 50–93% of patients.⁹ Surgical management mainly involves laparoscopic Heller's myotomy with or without Dor/Toupet fundoplication. Success rates are good in regard to symptom relief but, as with all the treatments, available

recurrence rate are high and efficacy wanes with time.¹⁰ Fundoplication following myotomy is recommended as gastro-oesophageal reflux disease is common. The patient in this case underwent myotomy only; whether this resulted in early disease recurrence is difficult to determine but such practice goes against recent guidelines from the Society of American Gastrointestinal and Endoscopic Surgeons recommending that patients undergoing myotomy should have simultaneous fundoplication.¹¹

In conclusion, upper airway obstruction due to a dilated oesophagus is a rare but potentially lethal complication of achalasia. Prompt radiological imaging, high index of clinical suspicion and early gastric decompression are key to successfully managing this condition. There is little information available as to what factors predispose a patient with achalasia to airway obstruction and, clearly, more research needs to be carried into this area as well elucidating the pathological process in a bid to innovate new improved treatment strategies.

Learning points

- ▶ Little is understood regarding the aetiology and pathogenesis of achalasia.
- ▶ Achalasia can lead to life-threatening upper airway obstruction.
- ▶ Simple measures such as basic respiratory support and gastric decompression can be life-saving.
- ▶ Prompt CT scanning is an important diagnostic tool in managing this condition.
- ▶ Continuous positive airway pressure must be used cautiously given the risk of gastric swallowing and worsening oesophageal dilation.

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