End-stage achalasia leading to acute upper airway obstruction and respiratory arrest with successful resuscitation, a case report

Jacob Arahill-Whitham, Ben Thomson, Vishak Surendra, Thomas Haig, Subhaschandra Shetty

ABSTRACT

Respiratory arrest secondary to megaoesophagus is a rare complication of achalasia. We treated an 85-year-old female with a history of achalasia who presented with sudden respiratory arrest and cardiopulmonary resuscitation in the community. In the emergency department, she was intubated for respiratory distress secondary to upper airway obstruction and reduced consciousness. Flexible nasal endoscopy revealed a retropharyngeal bulge, and computed tomography (CT) demonstrated megaoesophagus with distal tapering. She was managed with gastric decompression and percutaneous endoscopic gastrostomy (PEG) feeding with an uncomplicated hospital course. This case provides a rare differential for a patient with acute upper airway obstruction and cardiopulmonary arrest and is the first such case described in the literature in Aotearoa New Zealand.

Achalasia is a rare pathology of the oesophagus thought to result from degeneration of ganglion cells in the myenteric plexus. Subsequently, there is an impairment of relaxation of the lower oesophageal sphincter (LES) and lower oesophagus peristalsis.[1] Untreated, gradual dilation of the lower oesophagus can lead to end-stage achalasia/megaoesophagus. Achalasia has a bimodal presentation, typically diagnosed between ages 20 to 40 and 60 to 70.[1] Diagnosis typically involves high-resolution manometry (HRM), endoscopy and barium meal examination.[1] End-stage achalasia occurs with progressive loss of oesophageal ability to contract, decompensation and oesophageal dilation with tortuous angulation.[2] Histologically, the oesophageal paralysis corresponds with absent ganglion cells and severe neural fibrosis.[3]

Case

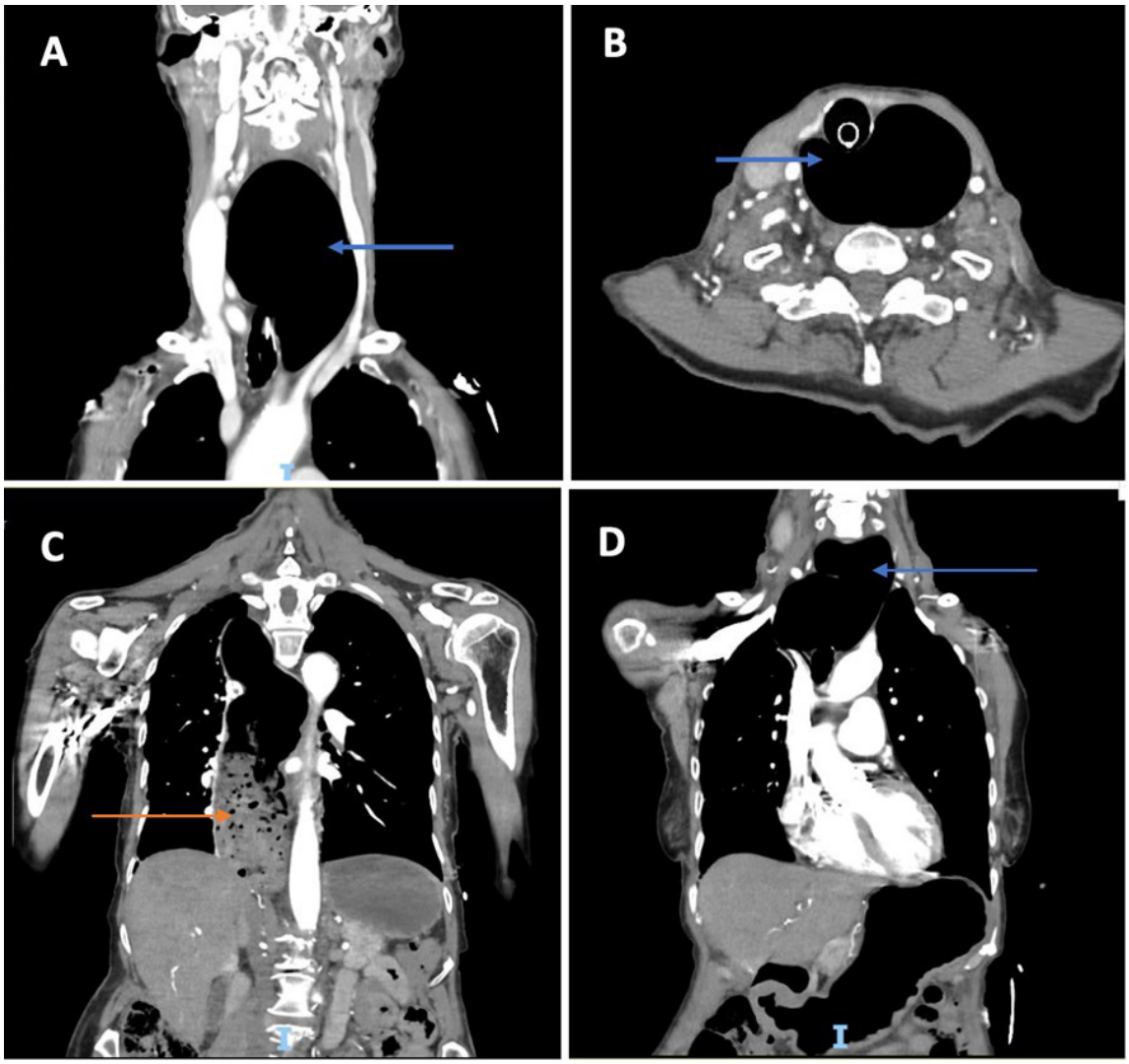
An 85-year-old female presented following sudden collapse and loss of consciousness while eating. Bystanders commenced CPR due to absent respiration before the arrival of emergency responders. Following ambulance transfer to the emergency department, she deteriorated again with increasing respiratory distress and reduced level of consciousness. Fibreoptic airway assessment before intubation revealed a mucosal bulging from the hypopharynx causing mechanical obstruction of the glottic opening. She was emergently intubated in the emergency department.

A venous blood gas revealed a respiratory acidosis consistent with hypoventilation, and a subsequent computed tomography (CT) identified that the abnormality was secondary to a grossly dilated oesophagus obstructing the upper airway (Figure 1A–B), which tapered down to the gastro-oesophageal junction as demonstrated in Figure 1C–D. There was no other sign of underlying pulmonary abnormality.

The patient's prior history was relevant for a clinical diagnosis of achalasia diagnosed in 1981 and undergoing a balloon dilatation that year. She had undergone an unremarkable gastroscopy in 2006 and another gastroscopy in 2013 that revealed a dilated and tortuous oesophagus, but the endoscopist believed the findings were not consistent with achalasia and no dilation was performed. She was never investigated with manometry. Other comorbidities included chronic obstructive airway disease.

Following partial decompression with a nasogastric tube, the patient was admitted to the intensive care unit and mechanically ventilated to correct the acid-base abnormalities. She was later extubated and stepped down to the ward.

Figure 1: Coronal (**A, D**) and axial (**B**) CT slices demonstrating gross distension of the upper oesophagus leading to clinical bulging of the neck, blue arrows. In the image (**C**), the orange arrow indicates food content within the distal oesophagus, which can be seen tapering to the gastro-oesophageal junction.



Unfortunately, the nasogastric tube became dislodged. She developed progressive upper airway obstruction and respiratory compromise, requiring re-intubation and re-insertion of a nasogastric tube with endoscopic guidance in the operating theatre. Following successful extubation, it was agreed between the patient, family and clinicians that she was not a suitable candidate for surgical correction of megaoesophagus given her age and comorbidities. The patient did not wish to be transferred to a tertiary centre where less invasive endoscopic procedures such as botox injection and balloon dilatation could occur. A percutaneous gastrostomy tube was placed for feeding. She was discharged home with multidisciplinary follow-up from dieticians and speech-language therapists and has subsequently established a near-normal oral dietary pattern.

Discussion

Bello et al.[7] first reported acute upper airway obstruction caused by achalasia in 1950. Since then, a relatively small number of presentations of acute airway obstruction secondary to achalasia and megaoesophagus have been reported, including four instances of sudden death.[8–10] There have been four published cases of cardiopulmonary arrest secondary to megaoesophagus with full recovery.[10–14]

This case presents a rare differential diagnosis for acute upper airway obstruction and respiratory arrest. The cause of airway obstruction could be temporised with the minimally invasive intervention of nasogastric decompression, and the patient recovered fully following resuscitation.

Achalasia can be radiologically graded based on the oesophageal diameter (I–IV).[4,5] Endoscopic botox injections and pneumatic balloon dilatation of the lower oesophageal sphincter can be offered to assist with voiding of the oesophagus, but do not address the impaired oesophageal peristalsis.[6] A number of surgical techniques exist to manage end-stage achalasia, such as Heller myotomy with Dor fundoplication, per-oral endoscopic myomectomy and oesophagectomy; however, these are associated with high risk of morbidity in high-risk patients.[6]

On endoscopic assessment, the patient had a widely patent lower oesophageal sphincter despite the grossly dilated oesophagus. The above endoscopic interventions were of unclear benefit for the patient and would have required transfer to a tertiary centre. Due to the non-functional oesophagus, a decision was made to manage the diagnosis conservatively due to frailty, comorbidities and the patient's wishes not to have further endoscopic or surgical procedures following discussion with the general surgical team, patient and their family.

Conclusion

This case highlights a rare complication of end-stage achalasia causing airway obstruction and respiratory arrest with successful resuscitation, and highlights some of the difficulties in the management of such conditions, particularly in rural settings.

COMPETING INTERESTS
The authors declare no conflict of interest in preparing this article.

AUTHOR INFORMATION
Jacob Arahill-Whitham, MBChB: Dunedin Hospital, Southern District Health Board, Otago University.
Ben Thomson, MBChB: Christchurch Hospital, Canterbury District Health Board.
Vishak Surendra, MS, DNB, FRCS (ORL-HNS): Whangārei Hospital, Northland District Health Board.
Thomas Haig, MBChB: Whangārei Hospital, Northland District Health Board.
Subhaschandra Shetty, MB BS, MS ENT, FRACS (ROL-HNS): Whangārei Hospital, Northland District Health Board.

CORRESPONDING AUTHOR
Jacob Arahill-Whitham: Dunedin Hospital, Southern District Health Board, Otago University.
E: jacob.arahill-whitham@southernDHB.govt.nz

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