The present case concerns a 40-year-old man who presented with post-prandial stridor which resolved spontaneously, later being diagnosed with achalasia. He underwent pneumatic dilatation year later, intended as definitive treatment. Despite intervention, the patient had developed megaoesophagus. One month later he presented with tracheal compression and cardiorespiratory arrest but was successfully resuscitated. He subsequently underwent elective oesophagectomy.

CONCLUSION: Oesophagectomy should be considered for patients with end-stage achalasia and megaesophagus causing respiratory compromise to avoid potential fatal complications such as tracheal compression and subsequent respiratory arrest.

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1. Introduction

Achalasia is an uncommon disease in which degeneration of the lower oesophageal sphincter (LOS) fails to relax. Patients commonly present with gastrointestinal symptoms such as difficulty in swallowing liquids and solids, regurgitation and heartburn. Other common symptoms include chest pain, hiccups and weight loss. Respiratory complaints occur less commonly and may be due to recurrent aspiration. Untreated achalasia may result in the progressive enlargement of the oesophagus into a megaoesophagus (>8 cm). This report presents the case of a man with a megaoesophagus suffering two episodes of respiratory distress within an 11-month period: the second resulted in cardiorespiratory arrest. Due to the high risk of recurrence, the patient underwent elective oesophagectomy. A review of the literature is also presented.

2. Case presentation

A 40 year old man with a history of tuberous sclerosis and childhood epilepsy was referred to the regional unit for oesophagectomy. He had previously received treatment following cardiorespiratory arrest secondary to complete airway obstruction by his megaoesophagus. 11-Months previously he presented to a local hospital with stridor. On that admission, chest radiography and contrast-enhanced computed tomography (CT) of the neck, abdomen and thorax had demonstrated gross oesophageal dilatation, as well as acute oesophageal tapering at the gastro-oesophageal junction: bird's beak sign (Fig. 1). In addition, secretions and food debris layered dependently in his oesophagus. The trachea was compressed to only 4 mm in diameter at the level of the brachiocephalic artery. CT did not show a lower oesophageal sphincter (LOS) mass, pneumomediastinum, pneumothorax or pleural effusion. Respiratory distress resolved spontaneously without the need
for ventilatory support. Subsequent endoscopy confirmed gross oesophageal dilatation with dependent food debris as seen on CT. He was relatively symptom free following this acute episode.

The diagnosis of achalasia with tracheal compression (Chicago classification) was subsequently confirmed by manometry: LES pressure of 57 mmHg (hypertensive) and minimal resting pressure of 44.8 mmHg (Fig. 2). Gastroscopy was attempted but failed due to resistance encountered with solid food despite restriction to a liquid diet five days prior to the procedure. He therefore subsequently underwent lavage and pneumatic dilatation to 30 mm under general anaesthetic with a view to further dilatation later.

Two months following dilatation, he was admitted following a postprandial choking episode that led to loss of consciousness. He initially regained consciousness spontaneously but proceeded to respiratory and ventricular tachycardic cardiac arrest. Following cardiopulmonary resuscitation and endotracheal ventilation, he had a return of spontaneous circulation and was admitted to the surgical intensive therapy unit with no apparent neurological deficits. An updated CT scan revealed further enlargement of the oesophagus and was now completely filled with food debris suggesting a further reduction in oesophago gastric transit (Fig. 3).

The oesophagus was noted to be further dilated, causing compression of the upper trachea to less than 5 mm in parts (Fig. 4). There was also increased atelectasis of the adjacent right lung and slight anterior displacement of the proximal aortic arch. The left lung exhibited features consistent with aspiration (consolidation, atelectasis and small airways exudation). Reduced tracheal calibre was confirmed on bronchscopy which also demonstrated tracheal collapse on inspiration. Rigid oesophagoscopy was performed and only incomplete removal of food debris was achieved due to technical difficulty. Endoscopic lavage and aspiration was subsequently performed to further decompress the oesophagus. A 5-day course of Co-Amoxiclav was commenced to treat his chest infection. He was successfully extubated after 2 days, made a good recovery and was discharged following institution of nasogastric tube (NG) feeding.

Due to the progressive oesophageal dilatation and recurrence of respiratory compromise despite pneumatic dilatation, oesophagectomy was considered more appropriate than BoTox, dilatation or myotomy. His medical history included tuberous sclerosis (causing adenoma sebaceum and CT-evident vertebral sclerotic islands), childhood epilepsy and infantile heart murmur. Of note, his post-arrest echocardiogram had not shown any abnormalities and the cardiologists were satisfied with his cardiac health. He had no relevant family history and worked as a secondary school teacher.

Three-stage oesophagectomy was considered the appropriate surgical approach since mobilisation, dissection and anastomosis to the cervical oesophagus was required. In the first stage, a right posterolateral thoracotomy was performed to mobilise the thoracic oesophagus. Mobilisation of the abdominal segment of the oesophagus was performed via a rooftop incision in the second stage. A
12Fr Foley catheter was then used to construct a feeding jejunostomy. In the third stage the cervical oesophagus was mobilised via a left sternocleidomastoid incision and an 8 cm × 8 cm oesophagus excised (Fig. 5). In view of the size discrepancy, the posterior surface of the distal oesophagus was anastomosed using a circular stapled technique to the anterior stomach to complete the third stage. There were no immediate or early complications.

The patient recovered well in the surgical high dependency unit. Oral diet was slowly introduced after anastomotic integrity was confirmed on Gastrografin swallow. The patient was discharged after 11 days on Lansoprazole 30 MG twice daily, Paracetamol 1G when required and Tramadol 50–100 MG when required. He was readmitted two weeks later complaining of gripping upper abdominal pain occurring 30 min postprandial and lasting 1–2 h. The pain was associated with sweating, bloating and the need to defecate. He was also constipated despite taking lactulose 10 mL twice daily. His blood glucose level was 5.1 mmol/L. Dumping syndrome was diagnosed and dietetics input was sought. He remains underst outpatient review.

3. Discussion

There have been over 40 published reports of achalasia causing partial tracheal obstruction and presenting as stridor. However, a de-duplicated search of AMED, EMBASE, MEDLINE, PsycINFO, BNI and CINAHL, in March 2014, combining the search terms “achalasia” and “arrest” returned only 14 unique results. Of these, only two papers were reports of patients suffering cardiac, respiratory or cardiorespiratory arrest. An additional case report was found from
a freehand search engine enquiry. The three eligible case reports are summarised in Table 1.

Brujin and Hicks in 2009 reported the case of a 79 year old woman who suffered respiratory arrest secondary to food bolus obstruction. This was the first presentation of achalasia. She recovered following oesophageal decompression and ventilatory support and subsequently received botulinum toxin (BoTox) injections to her LOS sphincter. It is not clear from the report whether she subsequently underwent either dilatation or myotomy. Alintoprak et al. in 2012 reported the case of a 35 year old woman who had experienced sudden onset dysphagia and respiratory distress whilst dining. She suffered cardiorespiratory arrest requiring seven minutes of resuscitation including external cardiac massage, intravenous adrenaline and atropine. A CT scan showed cardiac and tracheal compression by an enlarged oesophagus. Although this was her first presentation with achalasia, she reported having suffered dysphagia for 18 years. She underwent oesophageal lavage to treat the acute episode and subsequently underwent oesophagectomy and Dor fundoplication for definitive treatment. This patient had no gastrointestinal or respiratory complaints 18 months after this episode. Hifumi et al. in 2013 described the case of a 53 year old woman who had experienced sudden-onset dyspnoea and collapse. She suffered two episodes of cardiopulmonary arrest (pulsatile electrical activity) en route to hospital and on both occasions had return of spontaneous circulation after two minutes of chest compressions. She was not known to have achalasia although she had developed dysphagia 13 years previously but not investigated at the time. Following NG tube oesophageal decompression, she underwent pneumatic dilatation on day 11 and was discharged on day 33. No further follow up details were reported.

The patient described in the present report is unique for several reasons. It is the first report of a man with achalasia experiencing cardiopulmonary arrest although the incidence is equal in men and women. Unlike the patients reported by Bruijn, Hifumi and Alintoprak, this man underwent oesophagectomy as definitive treatment. As demonstrated in these cases, treatments such as BoTox, myotomy and pneumatic dilatation may yield effective results and should be considered. However, since our patient suffered cardiopulmonary arrest despite a trial of definitive treatment with pneumatic dilatation, and since this condition can be fatal, these alternatives presented an unacceptably unknown risk to the patient. Pneumatic dilatation had failed to prevent arrest whilst BoTox wears off and requires reinjection periodically. Also, oesophagectomy was preferred to myotomy, a procedure with lower operative risk, since Orringer et al. have previously shown that patients with megaesophagus respond poorly to myotomy. Oesophagectomy was considered the most reliable risk reduction strategy in this unique situation that is not addressed in current achalasia guidelines.

Although not in cases presenting with cardiorespiratory arrest, oesophagectomy has been used to treat end-stage achalasia successfully. The largest case series of oesophagectomy for achalasia is from the University of Michigan Medical Centre. A database of 93 patients (mean age 51) who underwent oesophagectomy for achalasia over a 20 year period was retrospectively reviewed. Most (64%) had a megaesophagus and a similar number (63%) were post-myotomy. Our patient’s postoperative stay of 11 days compares well with the mean postoperative stay of 12.5 days in the Michigan series. Dumping syndrome is a rare complication, affecting 4% of patients. However, most patients resume normal diets and are highly satisfied with their operative result and postoperative quality of life. Follow up of our patient continues to ensure postoperative quality of life is optimised.

Although cardiac, respiratory or cardiorespiratory arrest as a result of oesophageal compression in achalasia is rare, it can be fatal if not promptly treated. Important points in history include preceding dysphagia to both liquids and solids and sudden onset postprandial respiratory distress. However, as the patients in this report demonstrate, symptoms can be relatively minor. The significance of tenuous sclerosis in the history is not clear. However, there has been one case report suggesting an association. In the acute setting, a detailed history from the patient may not be forthcoming and a collateral history may be necessary. However, early investigations including chest radiography and especially CT may suggest the diagnosis. Early oesophageal decompression may reduce tracheal compression and reduce respiratory distress. This may be performed via NG tube or gastroscopy and lavage may also be required. The choice of definitive treatment will depend on many factors including patient wishes, performance status and surgical expertise. In cases of recurrent respiratory distress due to megaesophagus and where other treatments have failed, oesophagectomy may be the only resort in otherwise fit patients. However, randomised controlled trials to compare the various treatments will be difficult due to the rarity of the problem.

In conclusion, oesophagectomy should be considered for patients with end-stage achalasia and mega-oesophagus causing respiratory compromise to avoid potentially fatal complications such as tracheal compression and subsequent respiratory arrest.

**Conflict of interest**

None.

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None.

**Ethical approval**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

**Author contributions**

All authors were involved in patient care, writing and critical review of the manuscript.

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**Key learning points**

- The differential diagnosis of sudden onset postprandial dysphagia and respiratory distress includes achalasia and Booerhaave’s syndrome.
- Progressive achalasia is potentially fatal and requires thorough investigation and prompt treatment.
- Patients presenting with achalasia suffering recurrent respiratory distress should be considered for oesophagectomy.
- Oesophagectomy may be the best treatment option for megaesophagus.
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