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Recurrent respiratory distress and cardiopulmonary arrest caused by megaoesophagus secondary to achalasia☆

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ABSTRACT

INTRODUCTION: Respiratory distress and arrest from tracheal compression secondary to megaoesophagus are rare complications of achalasia. We present the case of a man with end-stage achalasia who required oesophagectomy to prevent recurrent life-threatening tracheal compression and respiratory arrest. A literature review is also presented.

PRESENTATION OF CASE: A 40-year old man 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.

DISCUSSION: Over 40 case reports of achalasia presenting with stridor have been published. However, only three cases (all female, age range, 35–79 years old) of cardiac, respiratory or cardiorespiratory arrest have been published. The definitive treatments received by these patients were botulinum toxin injections, open Heller cardiomyotomy with Dor fundoplication and pneumatic dilatation. None of these patients suffered recurrent respiratory distress following definitive treatment. The patient currently reported was unique as he suffered cardiorespiratory arrest following an intended definitive treatment, pneumatic dilatation. As such oesophagectomy was considered the greatest risk-reduction intervention.

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

1. Introduction

Achalasia is an uncommon disease in which degeneration of oesophageal mytenteric plexi results in the lower oesophageal sphincter (LOS) failing 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 dilation, 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

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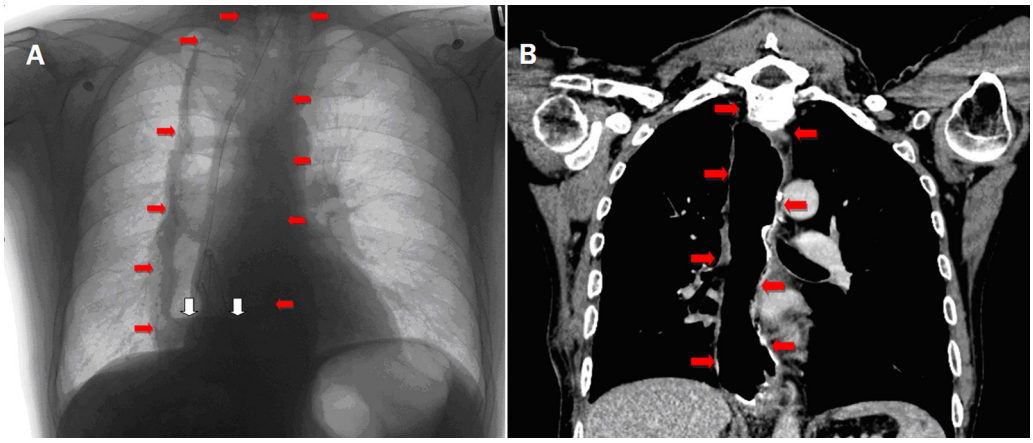

Fig. 1. (A) Chest radiograph showing a dilated oesophagus (red arrows). A nasogastric tube is seen in the lumen of the oesophagus. Oesophageal contents are visible as a fluid level (white arrows) behind the right heart border. (B) Coronal CT scan of thorax showing oesophageal dilatation (red arrows) with tapering towards the gastro-oesophageal junction (bird's beak sign). Food debris is seen at the distal end of the oesophagus. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)

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 oesophagogastric 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 bronchoscopy 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 dilation, 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

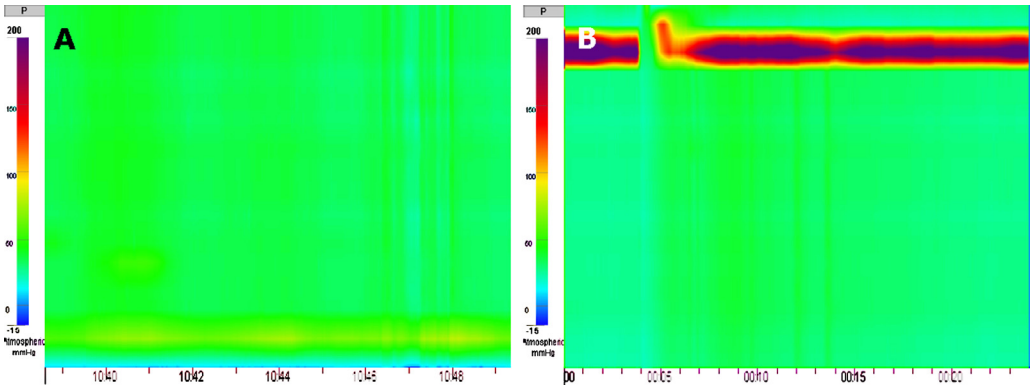

Fig. 2. Clouse plots demonstrating typical patterns in achalasia. In a non-pathological oesophagus, the background colour of the plots is blue, denoting low resting pressure. In this patient with achalasia, the green background in (A) denotes a raised minimal resting pressure of 44.8 mmHg and the green-yellow strip denotes an LES pressure of 57 mmHg. (B) Pressures reached 200 mmHg on swallowing 5 mm of water. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)