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Oesophagectomy for End-Stage Achalasia

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Abstract

Presentation

A 53 year old male with known Chicago Classification type II achalasia, and successful pneumatic dilatation five years previously, presented with severe dysphagia and 17.5 kg weight loss over 3 months.

Diagnosis

He underwent OGD and contrast imaging to reveal a mega oesophagus secondary to progressive achalasia.

Treatment

After initial nutritional pre-habilitation with naso-enteric feeding, he underwent a laparoscopic heller's myotomy with clinical and radiological improvement. However quick relapse of symptoms and a failed, atonic, massively dilated oesophagus lead to the decision to proceed to transhiatal oesophagectomy.

Discussion

Achalasia is a spectrum of motility disorder, and where it has progressed to mega-oesophagus, the success of standard therapeutic approaches is limited. End stage achalasia in this context, with nutritional failure or recurrent pneumonia/bronchiectasis, can be safely treated with an oesophageal resection which is curative, removing a "failed" oesophagus in its entirety.

Introduction

Achalasia is a motility disorder of the oesophagus characterized by failure of the lower oesophageal stricture (LOS) to relax on swallowing, and an absence of normal peristalsis. High resolution manometry (HRM) classifies 3 types, with Chicago Classification Type II characterized by pan-oesophageal pressurizations being the most common. There is a spectrum of clinical presentation, from mild intermittent symptoms to severe dysphagia with weight loss.

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Standard treatment options include endoscopic pneumatic dilatation (PD), laparoscopic Heller's myotomy (LHM), per oral endoscopic myotomy (POEM), or botulinum toxin injection of the sphincter [1]. All treatments target the LOS, enabling relaxation on swallowing and minimize risk of aspiration [2]. Oesophagectomy is rarely required and definitely as a last resort [3,4].

At our Centre, from 2010 to 2019, 148 patients of achalasia were managed, 120 by PD, 25 by LHM where PD failed, and in 3 patients (2%) an oesophagectomy was performed. In the same time period 475 oesophageal resections were performed for cancer, and we have recently reported this series [5]. The purpose of this report is to highlight a recent case what required oesophageal resection, and the step-up approach in management employed to the point where this decision was made.

Case Report

A 53 year old male with known type II achalasia, and successful PD five years previously, presented with severe dysphagia and 17.5 kg weight loss over 3 months. His Eckardt score was 10 signifying dysphagia and regurgitation at each meal, occasional retrosternal pain and greater than 10 Kg weight loss. An endoscopy revealed a dilated and atonic oesophagus with food stasis, no evident tumour or associated infection and an eccentrically positioned and closed LOS. Barium swallow showed a mega-oesophagus and the classic birds-beak narrowing at the LOS. CT imaging measured lower oesophagus to be 18cm in maximum transverse diameter (Figure 1, Panel A and B). High Resolution Manometry showed 100% failed swallows.

Having had a previous PD, a long myotomy was fashioned laparoscopically. Clinically he improved immediately, contrast studies showed improved emptying and he began to tolerate a light diet. However, he relapsed 2 months later and was again totally dependent on naso-jejunal tube feeding. He was deemed to have end-stage achalasia, meaning standard treatments were exhausted and no option other than resection was likely to impact on the disease. He underwent an oesophagectomy via a transhiatal approach and made an uncomplicated recovery. Pathology showed complete absence of ganglion cells on immunohistochemistry, and extensive mural thickening with muscular hypertrophy and fibrosis. He regained 8 kg at four months follow up. His weight has since stayed steady and he reports a markedly improved quality of life a year post surgery.

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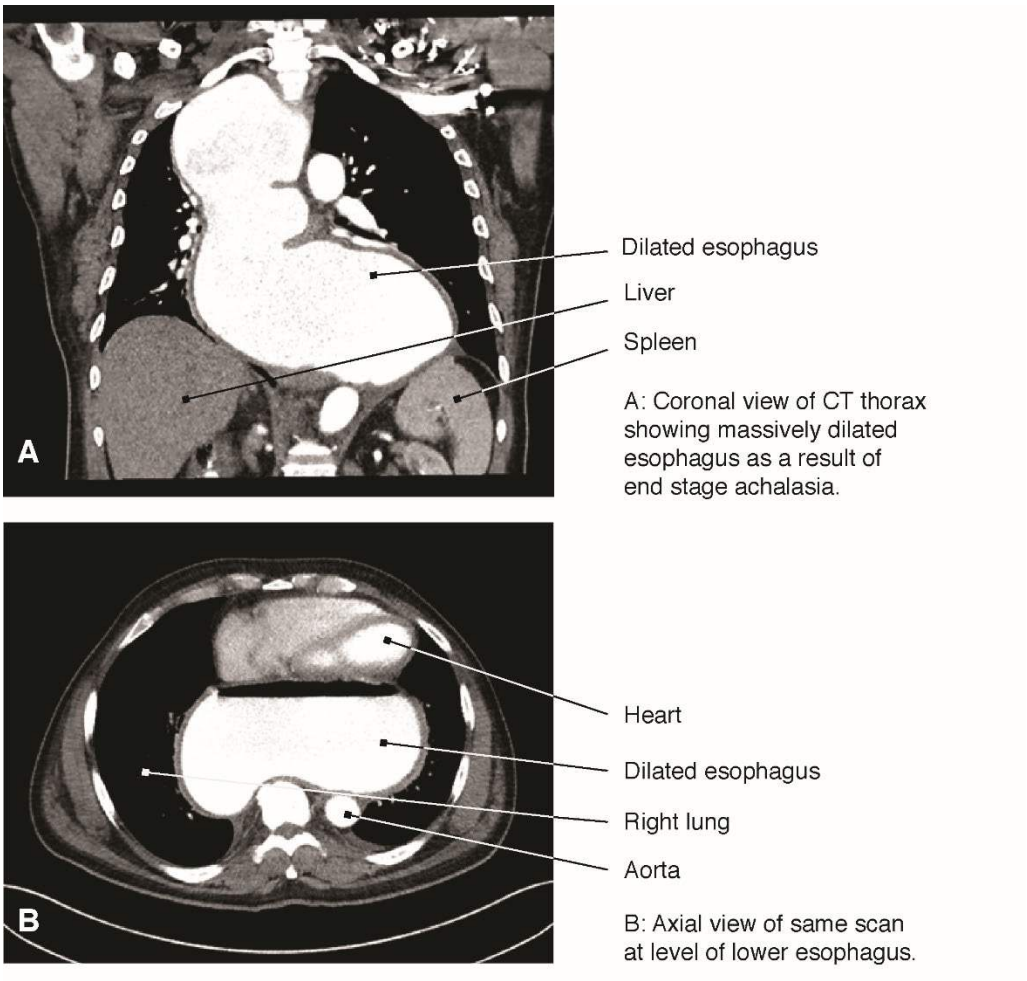

Figure 1: Mega-oesophagus in end stage achalasia

Discussion

This case, we feel nicely illustrates the typical end stage achalasia case, with therapies exhausted, but the remarkable anatomic feature of an 18 cm diameter oesophagus, with a transverse lie above the LOS, clearly highlighting how other modalities are likely to fail in this context. PD is reported to usually fail in mega-oesophagus (diameter >6cm) [3]. A myotomy was therefore preferred, as it may be successful even in the presence of a mega-oesophagus, and Type II achalasia is also associated with the best response, however the effect was short-lived in this case even after a technically satisfactory procedure with an immediate clinical improvement [2,6]. A transhiatal surgical approach is a relatively safer, avoiding a thoracotomy which may increase pulmonary morbidity [7]. Minimally invasive approaches to resection

could also be considered, notwithstanding he made an excellent recovery and has good recovery of quality of life.

In conclusion, Achalasia presents as a spectrum of motility disorder. End stage achalasia, where it has progressed to mega-oesophagus, the success of standard therapeutic approaches is limited; with nutritional failure or recurrent pneumonia/bronchiectasis, it can be safely treated with an oesophageal resection which is curative, removing a "failed" oesophagus in its entirety. The procedure should only be done in a high-volume specialist center that can provide excellent operative outcomes.

Declarations of Conflicts of Interest:

None declared.

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