Oesophagectomy in the management of end-stage achalasia — Case reports and a review of the literature

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A B S T R A C T

Achalasia is an oesophageal motility disorder characterised by aperistalsis and failure of relaxation of a hypertensive lower oesophageal sphincter. Treatment intent targets the sphincter, and either Heller’s myotomy or pneumatic dilatation successfully relieves dysphagia in the majority of cases. End-stage achalasia, typified by a massively dilated and tortuous oesophagus, may occur in patients previously treated but where further dilatation or myotomy fails to relieve dysphagia or prevent nutritional deterioration, and oesophagectomy may be the only option. We describe two patients with end-stage achalasia and nutritional failure despite exhaustive conventional therapy including pneumatic dilatation and surgical myotomy. Both patients were successfully managed with transhiatal oesophagectomy and cervical gastro-oesophageal anastomosis, with excellent symptomatic control and improved quality of life. These cases are discussed and the literature reviewed.

1. Introduction

Achalasia is a primary motility disorder of the oesophagus characterised by loss of peristalsis in the oesophageal body, impaired relaxation of the lower oesophageal sphincter (LOS) during swallowing, and increased resting pressure of the LOS. It results from loss of ganglion cells in the myenteric plexus, but the precise aetiology remains unknown. Common symptoms include dysphagia, regurgitation and weight loss, and occasionally respiratory symptoms due to aspiration of oesophageal content.

Achalasia is a rare disease, with an estimated annual incidence of 0.5 cases per 100,000 people, and a prevalence of 8 cases per 100,000 people per year. No treatment is available that will restore oesophageal peristalsis and normalise LOS relaxation. Therefore, treatment aims to palliate rather than cure, through reducing the pressure gradient across the LOS to facilitate and improve gravitational oesophageal emptying.

The two most commonly used treatment modalities for achalasia are pneumatic dilatation (PD) and surgical myotomy. Achalasia represents a wide spectrum of disease severity, and while some patients achieve excellent long-term symptom control from a single PD, other patients will go on to require multiple interventions to control symptoms. In the absence of therapy, or if therapy is inadequate, progressive dilatation and increasing tortuosity of the oesophagus may occur, resulting in end-stage disease. End stage disease, characterised by a markedly dilated and tortuous “burned-out” oesophagus and recurrent obstructive symptoms, may require oesophageal resection in order to restore gastrointestinal function, reverse nutritional deficits and reduce the risk of aspiration pneumonia.

In this report, we aimed to examine our own experience in the management of patients with end-stage achalasia and to review the literature available on the use of oesophagectomy in the management of end-stage achalasia.

2. Cases

In this Unit, 70 patients have been managed with achalasia between 1998 and 2007. The following two cases (3.5%) underwent oesophagectomy for end-stage achalasia.

2.1. Case 1

A 57 year old female who had a 39 year history of achalasia was referred to our unit with severe dysphagia, weight loss and recurrent aspiration pneumonia. She had undergone a trans-thoracic Heller’s myotomy at the time of her diagnosis, with good symptom control for 25 years. Following symptom recurrence she was managed with three pneumatic dilatations using the Rigiflex balloon (3–3.5 cm), all of which provided only temporary control of symptoms. At this time, she was referred...
to our specialist oesophageal surgery unit for further management. At the time of referral her BMI was 22, and she required naso-gastric feeding due to hypo-albuminemia (19 g/L) and nutritional failure.

Initial investigations included a timed barium swallow, a CT thorax and an endoscopy. The barium study revealed a markedly dilated oesophagus with food debris, a tight oesophago-gastric junction and markedly delayed emptying of the oesophagus. Endoscopy confirmed a fluid-filled mega-oesophagus. The CT scan demonstrated the mega-oesophagus and signs of aspiration pneumonitis (Fig. 1). At this centre we performed a single pneumatic dilatation with a Rigiﬁex balloon (3.5 cm). This failed, and we elected to do a laparoscopic abdominal Heller’s myotomy with a 6 cm oesophageal myotomy with gastric extension for 3 cm.

Manometry was performed and demonstrated aperistalsis of the oesophageal body. The patient failed to obtain even temporary symptom control post-operatively, and so decision was made to perform oesophageal resection.

A transhiatal oesophagectomy was carried out via abdominal midline and left neck incisions with blunt mobilisation of the mega-oesophagus, gastric pull-up and a cervical anastomosis. Despite moderate peri-oesophageal adhesions, there were no signiﬁcant intra-operative diﬃculties, and a transhiatal approach was safely performed. The approximate intra-operative blood loss was 450 ml.

Routine post-oesophagectomy care was provided, including naso-gastric tube drainage, epidural and patient controlled analgesia (PCA), and enteral feeding via a jejunostomy tube placed intra-operatively. She was discharged home twenty days post-operatively.

Histopathology and immunostaining of the resected specimen was consistent with achalasia, revealing marked hypertrophy of the inner circular layer of the muscularis propria, ﬁbrosis in the muscle layer of the distal oesophagus, and an absence of ganglion cells in either Auerbach’s or Meissner’s plexus.

She completed the Eckardt achalasia patient satisfaction and asymptomatic outcome score at her 12 month post-operative review. BMI had increased to 27. She had a Total Symptom Score (TSS) of 2, equating to good symptomatic outcome.

2.2. Case 2

A 65 year old female with a fifteen year history of achalasia was referred to our unit with refractory disease. She had undergone a laparoscopic Heller’s myotomy at the time of diagnosis, with symptom recurrence after four years. Following on from this, the patient underwent six pneumatic dilatation’s and re-do myotomy with only temporary symptom control. Re-do myotomy had been performed one year prior to referral, without success, and the patient complained of progressive severe dysphagia, regurgitation, and nutritional failure. BMI at time of referral was 19.5. Diagnostic work-up was performed. OGD showed a grossly dilated and aperistaltic oesophagus. Timed barium swallow demonstrated a grossly dilated oesophagus and tight narrowing at the LOS, with markedly delayed oesophageal emptying, consistent with advanced achalasia (Fig. 2). At this centre we performed a single PD but this failed to provide any symptom relief. Manometry demonstrated aperistalsis of the oesophageal body and incomplete relaxation of the oesophago-gastric junction.

A transhiatal oesophagectomy was performed. The approximate intra-operative blood loss was 510 ml. There were no post-operative complications, and she was discharged home on day 14 post-operatively.

The resected specimen revealed a dilated oesophagus with a diverticulum (Fig. 3). Histopathological examination and immunostaining demonstrated a marked reduction in myenteric and submucosal plexi, consistent with achalasia.

At 9 month follow-up, she was completely asymptomatic, with an Eckardt Total Symptom Score of 0, and reported being satisﬁed with her outcome, and willingness to undergo the procedure again if given her time back. She had re-gained weight with a BMI of 24.5.

Fig. 1. Coronal view from the CT Thorax from patient 1, showing dilated oesophagus with food and fluid stasis.

Fig. 2. Barium meal image from patient 2, demonstrating a grossly dilated lower oesophagus with beaking of the gastro-oesophageal junction, consistent with advanced achalasia.
2.3. Literature search strategy and selection criteria

References for the literature review were identified by searches of Pub Med, MEDLINE and Current Contents using the search terms "achalasia", "end-stage achalasia", and "oesophagectomy". References from identified articles were investigated for relevance. Abstracts and reports from meetings were not included. Only papers published in English between 1970 and May 2009 were included. The papers are shown in Table 1.

3. Discussion

End stage achalasia occurs in less than 5% of all achalasia patients, and may be characterised both clinically and radiologically. Radiological features include a tortuous (sigmoid) and massively dilated oesophagus, usually greater than 6 cm in diameter. Clinically, patients present with severe dysphagia, regurgitation, and nutritional failure. The management of patients with end-stage disease is challenging, and many have already undergone multiple failed therapeutic procedures. Unfortunately, treatment options are limited, and oesophageal resection may be required in selected patients.

All patients who present with symptom recurrence following therapy for achalasia must be carefully evaluated with repeat endoscopy, manometry and contrast study in order to determine the cause of treatment failure, which most commonly results from inadequate or healed myotomy, the development of reflux oesophagitis and stricture, obstruction from the fundoplication wrap, carcinoma, or development of a para-oesophageal hernia.

Patients who have only undergone pneumatic dilatation should be offered surgical myotomy. Patients with failed myotomy should be offered re-do myotomy, or oesophagectomy in those patients with a highly tortuous and dilated sigmoid oesophagus. Patients with failed re-do myotomy should be offered oesophagectomy if symptoms are severe and quality of life is affected.

Devaney et al. reported on the largest series to date, of 93 patients who underwent oesophagectomy for end-stage achalasia. They suggest the use of oesophagectomy only in those patients who have a tortuous, sigmoid shaped mega-oesophagus, stating that the tortuosity will interfere with oesophageal emptying even after adequate myotomy. Guidelines for selection of patients who may benefit from oesophagectomy are described in Table 2. In contrast, if the patient has a massively dilated but relatively straight oesophagus, Heller’s myotomy can provide good symptom relief, and should be performed in the first instance. Both of our patients have obtained excellent symptomatic relief of dysphagia, report excellent symptom control, as well as improved quality of life and satisfaction with their outcome. A review of the literature shows similar results (Table 1), with good symptom control reported in 75–100% of patients. However, oesophagectomy is not without risk, and every patient must be fully informed of all associated risks. Reported mortality rates of 5–10% are described, while morbidity rates of up to 50% have been reported, and anastomotic leak in 10–20% of patients. Patients must also be informed of longer-term complications. Anastomotic stricture has been reported in up to 50% of patients, depending on length of post-operative follow-up. Dumping syndrome, reported in up to 20% of patients, tends to be self-limiting and may be managed medically if necessary, and vagal-sparing oesophagectomy may reduce this risk.

Oesophageal resection is often more technically difficult in the patient with achalasia, compared with resection for malignancy, due to a combination of factors. The anatomy is altered in mega-oesophagus, with oesophageal deviation most commonly into the right chest, making pneumothorax and need for tube thoracostomy more common in these patients. Secondly, hypertrophy of the oesophageal musculature can result in a richer blood supply, making haemostasis more difficult, especially during mediastinal dissection. Thirdly, mobilisation of the cervical oesophagus can also be more difficult, especially in those patients who have oesophageal dilatation extending more proximally to the level of the thoracic inlet, and extra care must be paid to avoiding recurrent laryngeal nerve injury. Finally, previous instrumentation and surgery can cause the development of scarring and adhesions with adjacent aorta and left lung, making transhiatal mobilisation more difficult.

We routinely perform transhiatal oesophagectomy with gastric pull-up and cervical anastomosis for benign oesophageal disease. Several surgical approaches have been described for oesophageal resection in achalasia, including transthoracic, transternal and more recently, laparoscopic transhiatal. For reconstruction, most surgeons advocate cervical oesophago-gastric anastomosis, although colonic interposition and jejunal bypass have been described.

In the hands of an experienced surgeon, the transhiatal approach is safe and effective in patients with end-stage achalasia, and reduces the morbidity risk associated with thoracotomy. However, due to additional technical difficulties associated with resection as described above, mediastinal dissection and haemostasis is more challenging, and conversion to a transthoracic procedure may be required. For these reasons, some authors advocate the transthoracic approach. Although still a relatively new procedure, a laparoscopic transhiatal approach can provide much improved visualisation during mediastinal dissection, with improved morbidity and mortality.

In conclusion, these two cases, combined with results from several large studies highlight the usual albeit rare scenario of end-stage achalasia and the therapeutic benefit of oesophagectomy, resulting in long-term resolution of the disabling obstructive symptoms, nutritional failure, and poor quality of life.
Table 1
Table demonstrating results of literature review on the use of oesophagectomy in the management of end-stage achalasia.

<table>
<thead>
<tr>
<th>Reference</th>
<th>Study size</th>
<th>Time from dx to surgery</th>
<th>Access</th>
<th>Reconstruction</th>
<th>Morbidity &amp; mortality</th>
<th>Mean follow-up</th>
<th>Outcome</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Devaney1</td>
<td>93</td>
<td>Not given</td>
<td>Transhiatal: 87 pts, (conversion to transthoracic in 6 pts)</td>
<td>Gastric with cervical anastomosis: 91 pts, Colonic interposition: 2 pts (prior gastric surgery)</td>
<td>Mortality: 2%; Major morbidity: 30%; (Anastomotic Leak: 10%); Anastomotic stricture: 50%</td>
<td>3.2 yrs</td>
<td>95% asymptomatic, 96% would undergo same again</td>
<td>Transhiatal oesophagectomy is a safe and effective therapy in selected patients with end-stage disease</td>
</tr>
<tr>
<td>Miller15</td>
<td>37</td>
<td>13.2 yrs</td>
<td>Transhiatal: 9 pts, Transthoracic: 28 pts</td>
<td>Gastric: 31 pts, Colon interposition: 6 pts</td>
<td>Mortality: 5.4%; Morbidity: 32.4%; (Anastomotic leak: 5.4%)</td>
<td>6.3 yrs</td>
<td>Excellent outcome: 74%, good outcome: 17%. Increased risk of bleeding in transhiatal approach. Surgical conduit did not affect patient outcome 100% on normal diet, 87% felt improved, 7% felt the same, 7% felt worse</td>
<td>Recommended transthoracic oesophagectomy for improved visualisation and haemostasis, rather than transhiatal approach</td>
</tr>
<tr>
<td>Banbury6</td>
<td>32</td>
<td>13 yrs</td>
<td>Transhiatal: 21 pts, Transthoracic: 11 pts</td>
<td>Gastric. Cervical/thoracic anastomosis</td>
<td>Mortality: 0%; Anastomotic leak: 13%</td>
<td>3.5 yrs</td>
<td>96% tolerate normal unrestricted diet</td>
<td>Oesophagectomy with gastric pull-up provides durable and effective relief of dysphagia in the majority of pts</td>
</tr>
<tr>
<td>Orringer16</td>
<td>26</td>
<td>Not reported</td>
<td>Transhiatal (2 pts required conversion to transthoracic)</td>
<td>Gastric, cervical anastomosis</td>
<td>Mortality: 0%; Morbidity: 19%; Anastomotic leak: 4%; Anastomotic stricture: 38%; Dumping syndrome: 19%; Mortality: 0%; Morbidity: 21%</td>
<td>96% felt “cured” with excellent quality of life</td>
<td>Oesophagectomy provides a reliable and effective treatment for end stage achalasia</td>
<td></td>
</tr>
<tr>
<td>Peters20</td>
<td>19</td>
<td>Not reported</td>
<td>Left Thoraco-abdominal</td>
<td>Colon interposition</td>
<td>Mortality: 5%; Morbidity: 50%; (Anastomotic leak: 10%)</td>
<td>1 yr</td>
<td>93% felt “cured” with excellent quality of life</td>
<td>Oesophagectomy is a safe and effective therapy in end-stage achalasia</td>
</tr>
<tr>
<td>Tank21</td>
<td>15</td>
<td>6.5 yrs</td>
<td>Transhiatal</td>
<td>Gastric, cervical anastomosis</td>
<td>Mortality: 9%; Anastomotic leak: 18%</td>
<td>18.5 yrs</td>
<td>Symptomatic improvement in 82%</td>
<td>Oesophagectomy can be performed with acceptable mortality in patients with achalasia</td>
</tr>
<tr>
<td>Palanivelu17</td>
<td>11</td>
<td>13 mo.</td>
<td>Laparoscopic transhiatal</td>
<td>Gastric, cervical anastomosis</td>
<td>Mortality: 9%; Anastomotic leak: 18%</td>
<td>6 yrs</td>
<td>Good patient satisfaction. 75% tolerating normal diet</td>
<td>Laparoscopic oesophagectomy is a safe and effective procedure in specialised centres</td>
</tr>
<tr>
<td>Hsu19</td>
<td>9</td>
<td>17 yrs</td>
<td>Left thoraco-abdominal</td>
<td>Colonic interposition</td>
<td>Mortality: 0%; Major morbidity: 22%</td>
<td>6 yrs</td>
<td>Limited distal oesophagectomy with short-colon interposition is a safe and feasible alternative for patients with end stage achalasia</td>
<td></td>
</tr>
<tr>
<td>Glatz7</td>
<td>8</td>
<td>14 yrs</td>
<td>Right thoraco-abdominal</td>
<td>Gastric. Thoracic anastomosis</td>
<td>0%</td>
<td>6 yrs</td>
<td>100% on a normal diet</td>
<td>Oesophagectomy is a safe and appropriate treatment option from managing end-stage achalasia</td>
</tr>
<tr>
<td>Lewandowski8</td>
<td>7</td>
<td>10.1 yrs</td>
<td>Transhiatal</td>
<td>Jejunal bypass in 6 patients. Transhiatal oesophagectomy with colonic interposition in 1 patient</td>
<td>0%</td>
<td>Not reported</td>
<td>100% had normal swallow and no dysphagia</td>
<td>Jejunal bypass offers a safe and effective method of managing mega-oesophagus</td>
</tr>
<tr>
<td>Schuchert22</td>
<td>6</td>
<td>19 yrs</td>
<td>Laparoscopic transhiatal</td>
<td>Gastric, cervical anastomosis</td>
<td>Mortality: 0%; Morbidity: 50%; (Anastomotic leak 17%)</td>
<td>2 yrs</td>
<td>Not described</td>
<td>Laparoscopic oesophagectomy is a safe and effective procedure in specialised centres</td>
</tr>
</tbody>
</table>
Table 2
Criteria for considering oesophagectomy in patients with end-stage achalasia.

<table>
<thead>
<tr>
<th>End stage achalasia</th>
<th>Failed myotomy/re-do myotomy as evidenced by:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical</td>
<td>Eckardt grading score of 6–12 (“Poor Response”)</td>
</tr>
<tr>
<td></td>
<td>Severe dysphagia or regurgitation</td>
</tr>
<tr>
<td></td>
<td>Nutritional failure</td>
</tr>
<tr>
<td></td>
<td>Poor quality of life</td>
</tr>
<tr>
<td></td>
<td>Massively dilated oesophagus (&gt;6 cm)</td>
</tr>
<tr>
<td>Radiological &amp;</td>
<td>Highly tortuous (sigmoid) oesophagus</td>
</tr>
<tr>
<td>manometric</td>
<td>Oesophageal aperistals</td>
</tr>
</tbody>
</table>

Conflict of interest
None declared.

References