Sigmoid Oesophagus: Case Report and Review of Literature

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Abstract

It is the severe form of achalasia that is characterised by massive dilatation and tortuosity of the oesophagus. It is seen in long standing achalasia where the dilated upper segment of the oesophagus sags down below the level of the narrowed lower segment causing mechanical obstruction. Whereas Heller’s cardiomycotomy is considered to be the operation of choice in simple achalasia, it is regarded as inadequate for the treatment of this form of achalasia. While oesophagectomy with substitution is regarded as the treatment of choice in this group of patient, newer options have emerged and preliminary results appear to be promising. We present a case of end-stage achalasia with sigmoid oesophagus that had modified Heller's procedure and reviewed relevant literature.

Introduction

Sigmoid oesophagus (also known as end stage achalasia) derives its name from the tortuous and dilated disposition and axis deviation of the normally fairly straight course of the oesophagus[1]. As the disease advances, the dilated upper segment of the oesophagus elongate and sags down to the lower narrowed segment causing mechanical obstruction. It represents a severeend stage in the progression of achalasia cardia[2]. It possess a challenge in management as often times are only remedied by surgical intervention, have higher incidence of extra-oesophageal complications and has the least satisfactory outcome of all stages of achalasia3,4. The aim of this review is to present a case report and give a concise overview of sigmoid oesophagus and highlight the changing concept and trend in its management.

Case Report

A 55 year old farmer presented to us with 20 years history of total dysphagia associated with regurgitation, retrosternal chest pain and weight loss. Had associated productive cough. Said to have received various treatments for peptic ulcer disease with no significant improvement. At presentation, he was chronically ill looking, weak, dehydrated, and pale but no significant peripheral lymphadenopathy. He had bilateral coarse crepitation’s poster basally on chest auscultation. Abdomen was scaphoid otherwise no significant findings.
He was admitted and resuscitated with intravenous fluid, antibiotics, nasogastric tube decompression of the oesophagus and blood transfusion. His barium swallow showed dilated sigmoid oesophagus loaded with food debris (Figure 1). Oesophagoscopy revealed massively dilated, tortuous and friable oesophagus with retained food debris. It was difficult to navigate through to the stomach. He was worked up for modified Heller’s cardiomyotomy via laparotomy. Patient had deposition of food debris and contrast that was used for barium swallow within the dilated and tortuous oesophagus as such, had on-table irrigation of the oesophagus with the aid of a nasogastric tube placed from the nose to the dilated segment of the oesophagus and a gastrostomy that was performed after a laparotomy via an upper midline incision. The effluent from the oesophagus contained a combination of food debris and barium. Intra-abdominal oesophagus was mobilized and modified Heller’s cardiomyotomy was performed. The gastrostomy and laparotomy were all closed. Post-operatively patient was able to swallow and was discharged.

Figure 1: Sigmoid oesophagus

Epidemiology

Achalasia is a rare disorder and even more so sigmoid oesophagus hence the paucity of large epidemiologic data. The incidence is said to be about 0.5-1.2/100,000 per year and a prevalence of 8-10 per 100,000. It is commoner with ageing but can occur in all age groups and has no sex predilection. Sigmoid oesophagus accounts for 5% of these [5 – 8]. There seem to be a geographic variation from various reported studies with lack of data from sub-Saharan and developing countries[4,9].

Aetiopathogenesis

Sigmoid oesophagus is the end stage in the progression of achalasia cardia, which is a neuromotor disorder of the oesophagus in which there is a hypertensive lower oesophageal sphincter (LES), aperistasis of the body of the oesophagus leading to dysphagia and progressive dilatation of the oesophagus. The aetiology is not fully understood yet however current evidence points to a multifactorial cause. Primary(idiopathic) achalasia has been associated with 4 aetiological factors:
- **Post infective**

  Various viral agents have been implicated in the aetiology. These include the measles virus, varicella zoster virus and herpes simplex virus [10,11]. However other studies failed to establish this relationship and there is still no evidence of direct causal effect[12,13].

- **Autoimmunity**

  There seem to be an immunological aetiology in achalasia. This is based on the findings of inflammatory infiltrate in the myenteric plexus of 90-100% of patient with achalasia [14, 15]. This hypothesis has been further supported by the presence of autoantibody in sera of patients with achalasia and an association with major histocompatibility complex class II antigen [16 - 20].

- **Genetic predisposition**

  There is evidence of genetic basis of achalasia with report of increased occurrence in monozygotic, siblings and familial cases[21 – 23]. Also achalasia has been associated with genetic disorder like Down’s syndrome and Parkinson’s disease [24, 25].

Other associated aetiological factors include polymorphism of certain gene products like nitric oxide synthase gene, VIPR1 gene, interleukin 23 receptor and protein tyrosine phosphatase non-receptor gene [26 –28].

Secondary achalasia is from known causes such as Chagas disease, autoimmune condition like scleroderma, previous oesophageal or vagal surgeries and gastric malignancy.

Irrespective of the aetiological contributing factors, the initial stage begins with degeneration of nitrinergic VIPnergic inhibitory nerve in the myenteric plexus and the unopposed action of the excitatory neurotransmitters such as acetylcholine. This leads to high amplitude non-peristaltic contractions. This initial phase is known as vigorous achalasia. With progressive destruction of the cholinergic nerves and ganglion cells, there is dilation and low amplitude simultaneous contraction in the body of the oesophagus. This phase is known as the classical achalasia[29,30]. With progression of the disease and in the absence of any remedy, there is massive dilatation of the oesophagus and with axis deviation and tortuosity giving rise to a sigmoid or “tortuous” or “end stage” oesophagus. These structural changes are irreversible hence the aim of treatment is often palliative.

- **Clinical Features**

  Sigmoid oesophagus result from neglected, undiagnosed or poorly treated achalasia cardia. Dysphagia is by far the most common symptom, occurring in as much as 80 -100% of patients. This dysphagia is usually long standing, progressive with periods of improved swallowing and has been classically described as paradoxical, first occurring to liquid before solid. Regurgitation is also a prominent feature of sigmoid oesophagus with it the risk of aspiration pneumonitis. Ho et al. reported about 80% of patients would present with this symptom, which is worsened by the recumbent position. There may be a long preceding history of nonspecific symptoms of chest pain and heartburn for which the patient may have been treated for gastro-esophageal reflex disease (GERD) in the past. Nutritional failure occurs in as much as 60-67% [31 –33] and there may be a history of slow eating, avoidance of social function that involves meals or self-taught adaptations like postprandial ambulation to encourage eating, either from aspiration of regurgitated feeds or from tracheal compression that could be life threatening as has been widely reported [34 –37]. They may present with other respiratory complications like lung abscess.

- **Investigations**

  Investigation of sigmoid oesophagus mainly depends on 3 standard modalities [38]:

  - **Oesophagogram:** This can show both the static and dynamic features of the oesophagus. It shows absent peristalsis and marked dilatation of the oesophageal body and smooth narrowing in the LES classically described as “bird beak” appearance. There is also hold up of barium within the oesophagus and delayed entry onto the stomach. These features are only 75% specific for achalasia cardia. Radiographically, sigmoid oesophagus is defined as dilatation of the distal oesophagus of more than 10cm and which takes a sigmoid course or one with axis deviation[39]. However, Orringer et al. defined it as greater than 8cm or more while others used 6 - 7cm as the cut off[40 – 43]. Other features that may be evident on the scout radiograph includes widening of the mediastinum, presence of air fluid levels within the mid oesophagus, absent gastric fundal gas and lung field abnormalities from recurrent aspirations.
Oesophagoa gastroscopy: The spectrum of endoscopic findings in sigmoid oesophagus ranges from normal looking mucosa with a dilated lumen and a tortuous course, to friable mucosa with debris from stasis of ingested materials, which can be sucked at this stage. The LES appears puckered and may not open with air insufflations however with a gentle pressure; the resistance of the sphincter is easily overcome[32, 33, 44]. Endoscopy also plays an important role in excluding pseudo achalasia and other causes of dysphagia. In some cases with extreme tortuosity, it may be difficult to navigate the whole length of the oesophagus with attendant risk of injury and perforation.

Manometric studies: Oesophageal manometry is regarded as the gold standard for diagnosis of achalasia cardia. However the dilatation and tortuosity of sigmoid oesophagus is readily demonstrated on an oesophagram or oesophagostoscopy. Manometry typically reveals incomplete or absent LES relaxation in response to a swallow, a high resting LES pressure > 45 mmHg and aperistalsis in the distal 2/3 of the oesophagus[45 – 47].

Chest CT Scan: This is done mainly to evaluate patients that present with respiratory symptoms that do not respond to routine treatment of lower respiratory tract infection. CT chest may show tracheobronchial tree compression by dilated oesophagus and lung changes like consolidation, fibrotic patch, ground glass and nodular opacities[59]. CT scan classification of sigmoid achalasia

1. Sigmoid type 1 (S1) - the oesophagus is significantly dilated and tortuous, but only a single lumen is seen on any computed tomography (CT) slice.
2. Sigmoid type 2 (S2) - the oesophagus is very dilated and tortuous and some CT slices show a double lumen.

Other investigation that may be needed however not routinely includes endoluminal ultrasound scan and magnetic resonance imaging (MRI)

Treatment

Medical treatment

It is well established that pharmacological treatment has no role in the treatment of sigmoid oesophagus because reduction in the LES pressure alone would not improve oesophageal emptying due to the tortuous, aperistaltic and dilated oesophagus and symptom relieve is only produced by endoscopic or surgical intervention. This was also observed in endoscopic botulinum toxin injection and endoscopic dilatation[40, 48, 49].

Pneumatic dilatation

Pneumatic dilatation has been reported to be very difficult using the standard method in patients with sigmoid oesophagus because of the extremely tortuous and dilated oesophagus. Akin et al. successfully dilated a sigmoid oesophagus by attaching a balloon to the endoscope with a plaster and both were passed into the stomach successfully. Pneumatic dilatation was performed with endoscopic and fluoroscopic guidance, after the balloon reached the mid-LES level, which was observed endoscopically in the retroflex position. Other methods that have been reported include attaching the pneumatic dilator with internal stiffener to the guide wire[50], use of an over tube and attaching the pneumatic dilator to an endoscope using a string.

Surgical treatment

Surgical management offers the only option in the treatment of sigmoid oesophagus. A great debate exists as to which surgical option is appropriate in an achalasia oesophagus with axis deviation.

Oesophagectomy is regarded by a majority of surgeons, as the conventional and most appropriate option for the treatment of sigmoid oesophagus. Devaney et al. published the largest series of oesophagectomy for treatment of achalasia and regarded sigmoid disposition of the oesophagus as one of the indications for oesophagectomy[51]. This is position is held by many surgeons[8,40, 43, 48, 52 – 55]. It is based on the thinking that even an adequately done cardiomyotomy (which is the most common and an effective surgical treatment for uncomplicated achalasia or even a mega-oesophagus without tortuosity) would still not enhance oesophageal clearance or relief symptoms in a tortuous oesophagus and that the mortality and morbidity rates associated with total oesophagectomy was acceptable in proficient hands. Also proponents of oesophagectomy cite the reported rate of development of malignancy is of about 3-7%[50], hence an added advantage in resectional treatment. Various degree of oesophageal resection has been described, ranging from total oesophagectomy to varying degree of distal oesophageal resection[51, 53, 55]. Oesophagectomy for end stage achalasia has its peculiar challenges. These include deviations of the tortuous oesophagus into the right hemithorax, enlarged and tortuous arterial supplies to the oesophagus of aortic origin which can cause troublesome intra-operative haemorrhage and there may be adhesion between the exposed submucosa and the aorta in patients that have undergone previous myotomy[8, 51]. Several surgical approaches have been described for oesophagectomy in achalasia. These include the transhiatal, transthoracic, thoracoabdominal and more recently laparoscopic transhiatal oesophagectomy. The most popular of these is the transhiatal oesophagectomy which although involves blind dissection, has been noted to be a very safe surgical approach, with the advantage of avoiding the complications associated with a thoracotomy [8,40,43,51,52,55,56].
The transthoracic route has the advantage of direct visualisation and better control of haemorrhage[53, 57]. Minimal invasive oesophagectomy for sigmoid oesophagus is yet to gain wide approval, with limited data on its outcome [58,59]. The options for establishing gut continuity includes gastric pull up with cervical oesphago-gastric anastomosis, long or short segment colonic interposition and jejunum conduit[40,43,51,52,55,56, 59].

Reported successful outcome, defined as significantly improved symptoms or ability to swallow normal diet, ranged from 75-96%. The mortality rates from most series ranged from 0 to 9% and morbidity rate was 13-36%[8].

**Laparoscopic Heller’s cardiomyotomy**

Cardiomyotomy was previously regarded as an inappropriate surgical treatment for sigmoid oesophagus by many thoracic surgeons[38, 43]. However there are increasing reports of its use as the first option in the treatment of advance achalasia. Patti and co-workers noted that there was excellent outcome when patients with axis deviation and dilatation were treated with laparoscopic Heller’s myotomy[42]. This was also supported by other reports[60 - 65]. Their rationale was that firstly, myotomy being a less invasive procedure capable of producing good symptom relief should be the first primary treatment modality even for sigmoid oesophagus, reserving oesophagectomy for failed cases, recurrent symptoms or in those with high grade dysplasia or co-existing malignancy. Secondly that the myotomy did not preclude a future oesophagectomy if the need arise. It is also a very useful option in frail patients not fit for extensive surgery[62]. The reported satisfactory treatment outcome ranged from 90 – 100% with minimal morbidity. This was disputed by Devaney et al. and numerous other researchers who claimed that myotomy does not provide a reliable relieve of dysphagia in sigmoid oesophagus[51]. It must also be noted that there are no report of long term follow up in these patient who had myotomy and most of the studies had rather limited number of patients.

**Peroral Endoscopic Myotomy (POEM)**

Peroral endoscopic myotomy (POEM) has also been reported as a useful and effective tool in the management of sigmoid oesophagus. In addition to its advantage of scarless and minimally invasive procedure, it has been shown to produce excellent objective outcome. In a large review of 500 patients treated with POEM for achalasia, Inoue and co-workers observed that POEM produced satisfactory outcome even in patients with sigmoid oesophagus[66].

**Outcome**

Oesophagectomy, surgical myotomy and endoscopic myotomy have all been reported to produce satisfactory outcomes in the treatment of an achalasic sigmoid oesophagus but there no comparative studies to compare their outcomes. POEM has shown promising result but long-term outcome is yet to be ascertained. However oesophagectomy still remains the last option when all other options fail.

**Conclusion**

There still exist great debates as to what is appropriate in the management of sigmoid oesophagus however there is a gradual shift towards less invasive therapy for the initial management of sigmoid achalasia. Oesophagectomy still remain the only option for many patients with sigmoid oesophagus, or those in whom myotomy and other treatment modalities had failed. There is need for large multicentric and possibly randomized control studies to provide definitive data as to the role of various treatment options in the management of this condition.

**References**


