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Case Report

Surgery Section

Laparoscopic Heller's Myotomy for Hypercontractile Oesophagus: A Case Report

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ABSTRACT

The oesophageal motility disorder is a wide range of diseases, extending from acontractile to Hypercontractile Oesophagus (HE), each having a unique pathophysiology. Hence, their management is also individualised. However, a rare subset of this disorder, namely, a HE, presents a management conundrum. There is no evidence of superiority of one treatment modality over the other within the spectrum of available treatment modalities, such as pharmacological, endoscopic, and surgical therapy in view of symptomatic relief and chances of recurrences. Hereby, the authors present a case report of 81-year-old male patient of HE successfully managed by laparoscopic Heller's myotomy with Dor's fundoplication. Over 20 postoperative months, the patient remains symptom-free.

Keywords: Dor's fundoplication, Endoscopic therapy, Oesophageal motility disorder, Pharmacological therapy

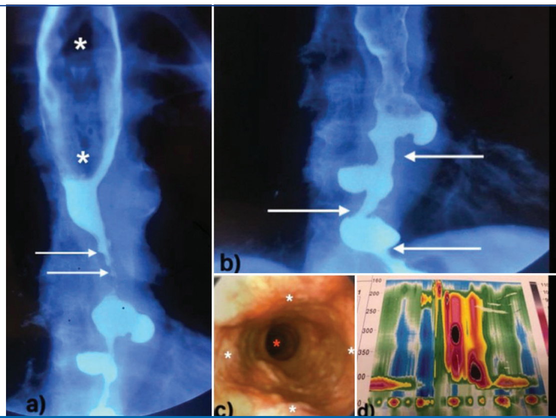
CASE REPORT

An 81-year-old male with no known co-morbidities presented with recurrent chest discomfort and dysphagia over three years. Initially, he had occasional chest discomfort associated with food intake and dysphagia, which was more pronounced with solids. Gradually, he experienced worsening of symptoms over the last six months, including more frequent severe spasmodic chest pain with no association with his oral food intake, dysphagia for both solids and liquids, nausea, and significant unintentional weight loss of approximately 7-8 kg over the last six months. Physical examination was unremarkable.

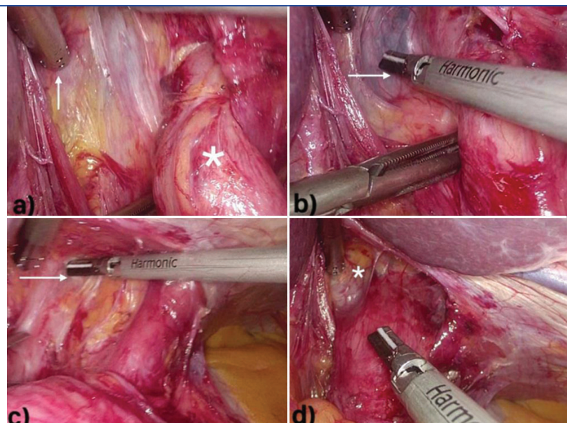
On evaluation, the Electrocardiogram (ECG), 2D Echocardiogram (ECHO), and the cardiac markers were within normal limits, thereby ruling out a cardiac cause. A barium swallow study with oral contrast demonstrated diffuse uncoordinated contractions predominantly in the lower oesophagus and dilated proximal oesophagus [Table/Fig-1a,b]. Upper Gastrointestinal (GI) endoscopy revealed no abnormality with a minimal short-lived spasmodic resistance at Gastro-oesophageal (GE) junction; however, the scope could be easily negotiated beyond the episode [Table/Fig-1c]. Then, a High Resolution oesophageal Manometry (HRM) showed evidence of high-amplitude contractions predominantly in the mid to lower body of oesophagus, with occasional swallow showing normal peristalsis, oesophageal pressures were low, and the mean amplitude of contraction exceeding 200 mmHg, suggestive of HE, previously called as nutcracker's oesophagus [Table/Fig-1d].

In view of the severity of the symptoms, the patient's age, the predominantly lower oesophageal location of the disease, availability of advanced laparoscopic setup, an experienced laparoscopic surgeon, and the necessity to prevent reflux after the myotomy, it was decided to proceed with the Heller-Dor procedure.

At laparoscopy, the lesser sac was entered after dividing the gastro-hepatic ligament, followed by division of the oesophagophrenic ligaments to achieve adequate exposure of the lower oesophagus at the oesophageal hiatus. The anterior vagus nerve was identified and preserved. The GE junction was identified, and the mediastinal dissection was performed up to the inferior pulmonary vein [Table/Fig-2a-d]. Then, a myotomy of approximately 13-14 cm was performed using a harmonic scalpel, caudally extending approximately 2-3 cm onto the gastric side below the GE junction [Table/Fig-3a-d]. Following this, the right and left crus of the diaphragm were sutured



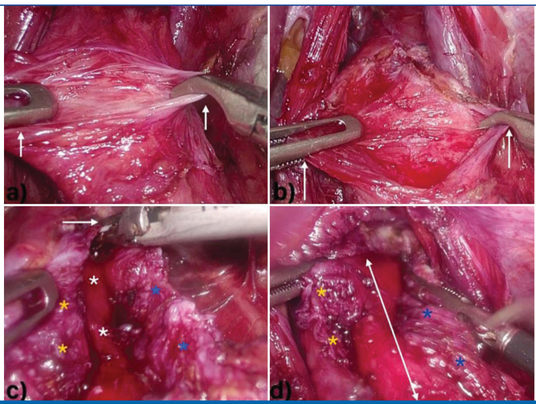
[Table/Fig-1]: Preoperative investigations' images: a,b) Barium swallow study with oral contrast demonstrates diffuse uncoordinated contractions (white arrows) predominantly in the lower oesophagus and dilated proximal oesophagus (white asterisks); c) Upper GI endoscopy showing spasmodic resistance at GE junction (red asterisk) with proximal dilated oesophagus (white asterisks); d) HRM findings suggestive of Hypercontractile Oesophagus (HE).



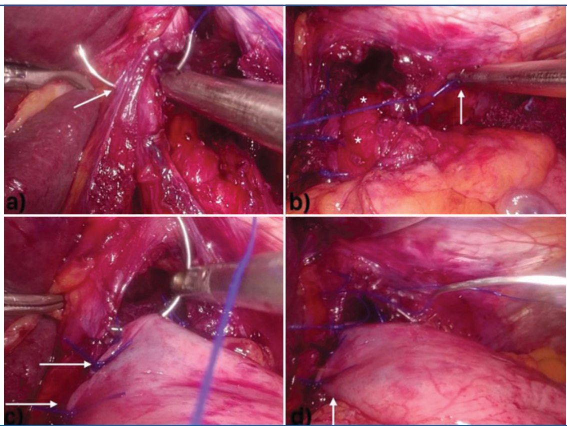
[Table/Fig-2]: Operative pics: a) Shows the oesophagus (white asterisk) and mediastinal dissection being commenced (white arrow); b-d) Shows mediastinal dissection being carried out using blunt dissection and harmonic scalpel (white arrows and white asterisk).

and fixed to the right and left lips of myotomy, respectively [Table/Fig-4a,b]. The and then, the gastric fundus was mobilised to perform a Dor's 180-degree anterior fundoplication [Table/Fig-4c,d].

Postoperatively, the patient was gradually started on liquid diet from Postoperative Day (POD) 1. The rest of the course in the hospital



[Table/Fig-3]: a,b) Myotomy being initiated by splitting both the longitudinal and circular muscle fibres from the underlying mucosa of oesophagus (white arrows); c) Myotomy being extended proximally on to the mediastinal oesophagus using harmonic scalpel (white arrow) exposing the mucosal layer beneath (white asterisks), the right and left lips of myotomy (yellow and blue asterisks respectively); d) The completed myotomy (double-headed white arrow).



[Table/Fig-4]: a) Right myotomy lip being sutured to right crus (white arrow); b) Left myotomy lip being suture-fixed to left crus (white arrow) with exposed esophageal mucosa (white asterisks); c) Fundus of stomach being sutured to right crus as part of anterior fundoplication (white arrows); d) Completed Dor's anterior 180 degree fundoplication (white arrow).

was uneventful, and the patient was discharged on a pureed diet on POD 3. On follow-up, the patient was symptom-free and gradually started on a soft to normal diet after two weeks. A telephonic interview was conducted with him 20 months after the surgery revealed that he continues to be completely symptom-free.

DISCUSSION

The HE is a fascinating yet rare motility disorder of the oesophagus characterised by abnormal contraction and function of the oesophagus [1]. During swallowing, the normal oesophagus has involuntary rhythmic contractions to propel food distally. However, in HE, these contractions become much stronger, more frequent, forceful, and dysrhythmic, failing propagative contractile force and ultimately leading to difficulty in swallowing, non cardiac chest pain, or regurgitation [1,2]. The term “nutcracker oesophagus” has been used to describe this condition but has fallen out of favour in clinical settings. Instead, it is now referred to as HE [1].

The exact aetiology of this condition is uncertain, but there are several risk factors found to be associated with it: age over 60 years, female gender, history of Gastro Oesophageal Reflux Disease (GERD), obesity, and certain medications like opioids, hypersensitivity of the oesophagus leading to eosinophilic esophagitis. In addition to the above-mentioned factors, the inherent oesophageal muscle thickness and function could also play a role [1,2]. A recent study found an increased risk of HE in individuals with spinal injury, hiatal hernia, and inappropriate excitation of the vagus nerve [3]. The exact pathophysiology behind HE is not completely understood and may be because of excessive cholinergic drive with temporal asynchrony of circular and longitudinal muscle contractions [4].

The common clinical presentations can be any of the following: chest pain, dysphagia, heartburn, regurgitation, or globus sensation. The chest pain secondary to intense oesophageal contractions can be so severe that it can often mistaken for heart-related pain. As the above-mentioned symptoms are non-specific, patients need to undergo a set of investigations, such as Upper Gastrointestinal Endoscopy (UGI scopy), High Resolution Manometry (HRM), 24-hour pH monitoring, Endoscopic Ultrasound Scan (EUS), Computed Tomography (CT) scan, to rule out all other possible differential diagnoses before labelling it as HE [3].

The use of conventional manometry has become obsolete, as HRM more accurately measures oesophageal contractions. Previously, the term “nutcracker oesophagus” referred to oesophageal contraction pressure greater than 180 mmHg or 216 mmHg when measured using conventional or high-resolution manometry, respectively [5,6]. Currently, HRM is the gold standard modality used to diagnose HE.

According to Chicago Classification version 4.0, HE is defined as an entity with clinically relevant symptoms and a high-resolution manometric calculation of at least 20% of the swallows with a Distal Contractile Integral (DCI) exceeding 8000 mmHg/cm/second, with normal Lower Oesophageal Sphincter (LES), i.e., normal Integrated Relaxation Pressure (IRP) [7]. In this recent update, “Jackhammer oesophagus” is no longer considered synonymous with HE, as three phenotypes of HE were given- single peak hypercontractile swallow, repetitive prolonged contractions (Jackhammer oesophagus), and a very new entity named hypercontractile LES after contraction [7].

Management protocol for HE has yet to be validated, and there is no single randomised, sham, or placebo-controlled prospective study in the literature, as the DCI value of more than 8000 mmHg/cm/s is rarely recorded in otherwise normal healthy control subjects with any of the above-mentioned symptoms [8].

The various treatment modalities include drugs (e.g., nitrates, calcium channel blockers, phosphodiesterase-5 inhibitors, anticholinergic, proton pump inhibitors, and antidepressants), endoscopic approaches (such as botulinum toxin injection, Peroral Endoscopic Myotomy (POEM), and pneumatic dilation), and laparoscopic myotomy [8]. Interestingly, a study by Schupack D et al., revealed that the majority of symptomatic patients (72.5%) showed clinical improvement or symptom resolution at a mean follow-up of 2.8 years, without any drugs or intervention [9]. Also, there is a chance of progression of HE into type II and III achalasia cardia in patients [10,11].

A meta-analysis showed an improvement in clinical symptoms in about 73.6% of patients who underwent medical and endoscopic management. The study also revealed an 82% success rate for POEM specifically [12]. However, a French cohort study published in the same year deemed medical and endoscopic management unsatisfactory, citing poor efficacy and high relapse rates [13]. Given the contrasting results of the above-mentioned studies and the non availability of long-term results to advocate the use of POEM as the prime modality of management, there is still scope for surgical management by laparoscopic myotomy coupled with anti-reflux procedure (to care for the postoperative reflux) in carefully selected patients.

The surgical management was first described by Lortat-Jacob in 1950 for non achalasia primary oesophageal motility disorders and was reserved especially for patients with obstructive symptoms [6]. Furthermore, a report by Nastos D et al., comparing the management of oesophageal spastic disorders by oesophageal myotomy with anti-reflux surgery demonstrated a better postoperative outcome in patients with spastic disorder and an accompanying epiphrenic diverticulum than in patients with pure spastic disorder [14]. The above mentioned report underscores the fact that a subset of patients with oesophageal spastic disorders and normal or low oesophageal sphincter pressure can benefit from surgical myotomy combined with anti-reflux surgery.

A review article by Siddaiah-Subramanya M et al., revealed that although myotomy relieved dysphagia in 77-89% of patients, it also induced gastroesophageal reflux in 31-100% of them [15]. To counter this effect, a complete or partial fundoplication to augment the myotomy was studied, and it was found that the occurrence of reflux was significantly less in patients with added fundoplication. But, Nissen's complete 360-degree fundoplication caused more severe dysphagia postoperatively, and the authors suggested that a partial fundoplication, either a Dor's or a Toupet's, be combined with myotomy to provide an anti-reflux mechanism without causing severe dysphagia [15].

Patient selection for surgical management is of prime importance, as long-term prognosis even without treatment seems to be good for this disease. Further, patient's overall performance status, co-morbidities, anaesthesia-related risks, the possibility of oesophageal perforation while performing myotomy in less experienced hands, and the persistence or aggravation of dysphagia after a complete 360 degree fundoplication all play a role in decision-making and can limit the practice of surgical approach compared to other less invasive endoscopic management like POEM. The authors believe that, due to obvious reasons, a surgical myotomy would have the best results among patients afflicted with HE when the disease affects the distal oesophagus.

CONCLUSION(S)

The HE is still an enigma, as the medical fraternity has yet to completely understand the pathophysiology, the relationship between the symptoms, and their association with the severity of the disease. Further, there are no homogeneous methods of investigation and treatment protocols. Hence, even in the era of endoscopic therapy, surgical management still has scope, especially for a subset of HE patients with involvement of LES, as it can help in the reduction of oesophageal reflux. However, more standardised prospective study models are needed to have a proper algorithm for the management of HE.

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