CASE REPORT

Minimally Invasive Total Esophagectomy For Giant Epiphrenic Diverticulum With Megaesophagus

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ABSTRACT

Background: Esophageal diverticulum is a rare condition, and its pathophysiology varies according to its anatomical location in the esophagus (i.e., Zenker's, Rokitansky, and Epiphrenic). Among the three, epiphrenic diverticulum is the rarest and is also known as pulsion diverticulum. It is caused by the herniation of mucosa and submucosa through the esophageal muscular layers as a result of increased intraluminal pressure, involving the distal third of the esophagus within 10cm of the esophagogastric junction.

A case of a 62-year-old lady with an obstructive esophageal diverticulum secondary to achalasia and megaesophagus that underwent minimally invasive total esophagectomy due to a dysfunctional esophagus.

Case Presentation: A 62-year-old lady presented with a history of progressively worsening solid food dysphagia associated with a significant loss of weight. On examination, the patient was malnourished (SGA B). No remarkable findings were noted on abdominal, systemic, or laboratory investigations. Following a series of imaging examinations with manometry showing raised lower esophageal sphincter pressure and absence of peristalsis, the patient was diagnosed with a giant epiphrenic diverticulum with megaesophagus. After a length of nutritional built-up, the patient underwent minimally invasive total esophagectomy and gastric pull-up. Post-operatively, the patient was cared for in the intensive care unit and was discharged well on day 10 of post-operation. The patient was reviewed back again two weeks upon discharge and was noted to be well.

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Discussion and Conclusions: Patients with epiphrenic diverticulum are mostly asymptomatic, and surgical interventions are reserved for progressively worsening symptomatic patients. It is mainly diagnosed by gastroscopy, contrasted studies, and manometry. Traditionally, diverticulectomy with myotomy is the treatment of choice for isolated esophageal diverticulum. Esophagectomy is an alternative approach and is the preferred surgical in this case when diverticulectomy with myotomy is not suitable. The surgical approach includes open or minimally invasive, with minimally invasive preferred. We proceeded with oesophagectomy since she was diagnosed with a giant epiphrenic diverticulum secondary to achalasia and megaesophagus

BACKGROUND

The esophageal diverticulum is a rare condition, and its pathophysiology varies according to its anatomical location in the esophagus (i.e., Zenker's, Rokitansky, and Epiphrenic). Among the three, the epiphrenic diverticulum is the rarest, comprising 0.015 to 2% of esophageal diverticula cases. Epiphrenic diverticulum, also known as pulsion diverticulum, is caused by the herniation of mucosa and submucosa through the esophageal muscular layers as a result of increased intraluminal pressure, involving the distal third of the esophagus within 10cm of the esophagogastric junction. [1].

We report a case of a 62-year-old lady with an obstructive esophageal diverticulum secondary to achalasia and megaesophagus that underwent minimally invasive total esophagectomy due to a dysfunctional esophagus.

CASE PRESENTATION

Chief complaints

A 62-year-old lady presented to the surgical outpatient clinic complaining of dysphagia of solid food for six months.

History of present illness

The patient's dysphagia was progressively worsening and associated with regurgitation. The patient also had a loss of weight of 10 kg within 10 months.

History of past illness

No significant medical or surgical history was noted.

Personal and family history

No significant personal or family history was noted.

Physical examination

The patient was noted to be malnourished with temporal wasting, mid-arm circumference of 28cm, and thinning of lumbrical muscles. PG – SGA score: B. score: B, indicating moderate malnutrition.

No mass or tenderness was elicited on abdominal examination.

Other systemic examinations were unremarkable.

Laboratory examination

Blood analyses such as full blood picture, renal profile & liver function test were unremarkable.

Imaging examination

Figure 1: Gastroscopy: Narrowing of the cardio-oesophageal junction with a huge esophageal epiphrenic diverticulum on the left with food bolus within. (Figure 1A)

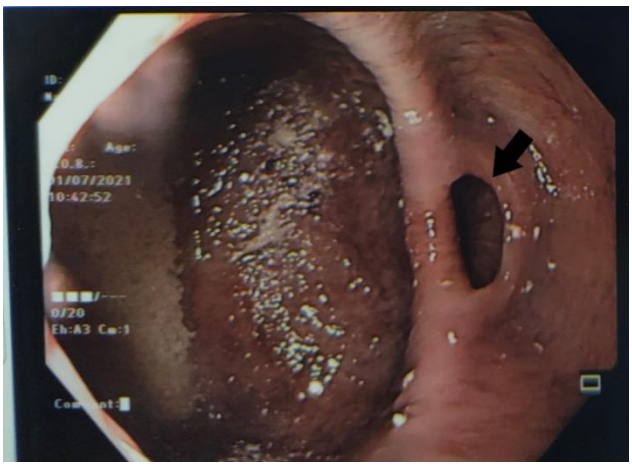


Figure 2: Esophagogram (barium swallow): A large diverticulum measuring 9.6 x 8.4cm at the distal esophagus, proximal to the gastroesophageal junction, in keeping with epiphrenic diverticulum, with a horizontal shelf-like filling defect diverticulum suggestive of a web. (Figure 2B)

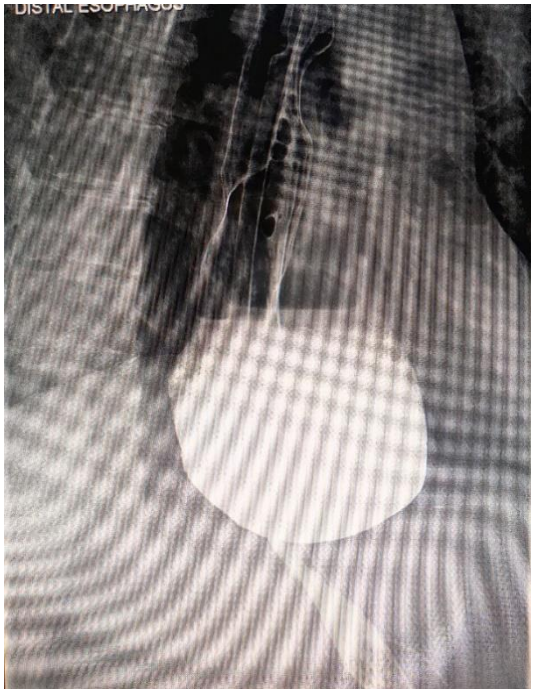
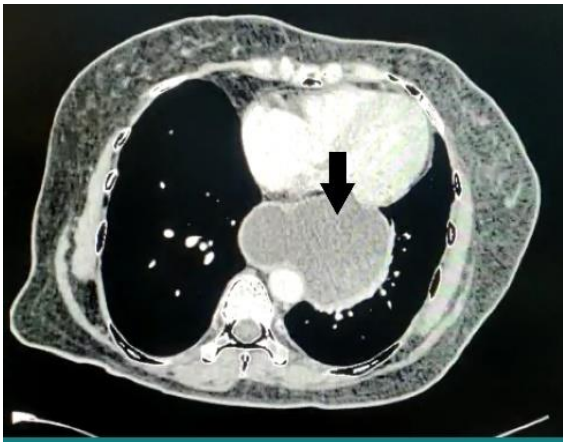


Figure 3: Contrasted CT Thorax/Abdomen: Distal esophageal giant diverticulum 65 x 68 x 73mm with dilated esophagus up to the upper thorax. (Figure 3C)



Manometry: Raised lower esophageal sphincter pressure and absence of oesophageal peristalsis.

Echocardiogram: Ejection fraction of 66%, good left ventricular function, normal chamber size, and trivial mitral regurgitation.

Lung function test: normal with FEV1/FVC of 98%.

Final Diagnosis

Giant epiphrenic esophageal diverticulum secondary to achalasia and megaesophagus.

Treatment

Optimization of nutrition was done via a gastroscopy wire-guided nasogastric tube insertion. The patient was started on polymeric feeding, beginning with 10kcal/kg, and gradually increased to 25kcal/kg, achieving 90% of the total energy requirement. Protein was kept at 1.5g/kg.

After one month of nutritional optimization (malnourished with SGA B), the patient underwent laparoscopic total minimally invasive esophagectomy and gastric pull-up reconstruction, and specimens were retrieved from the thoracic inlet. Intra-operatively, there was an epiphrenic diverticulum with a tight cardio-oesophageal junction and dilated esophagus. (Figure 4D)

Figure 4: Intraoperative finding showing epiphrenic diverticulum.

Post-operatively, the patient was sent to the intensive care unit and noted to have developed a cardiac arrhythmia, which was resolved by giving amiodarone.

At 48 hours post-operation, a gastroscopy was done to assess the conduit and anastomosis, which was normal.

The patient was extubated at 72 hours post-operation, was allowed oral feeding at 96 hours post-operation and started to ambulate at day 7.

As the patient was previously malnourished, the team kept her for another week to monitor and further build up her nutrition.

Outcome and follow-up

The patient was discharged home well two weeks after the operation.

The pathological report revealed a benign epiphrenic diverticulum.

Upon review two weeks post-discharge, the patient was noted to be well.

DISCUSSION AND CONCLUSIONS

An epiphrenic diverticulum is a false tract located within 10 cm of the gastroesophageal junction due to weakness of the muscularis propria at this junction .[2].

Only a small percentage of patients with esophageal diverticulum is symptomatic, and the usual complaints are dysphagia, regurgitation, anorexia, nocturnal cough, dyspepsia, and weight loss. Life-threatening circumstances, such as aspiration pneumonia and malignancy, have been reported, and are uncommon but can occur in situations with delayed diagnosis. [2,3].

The diagnosis of the esophageal diverticulum is mostly found incidentally, especially in asymptomatic patients. The standard investigations include a plain chest radiograph, gastroscope, esophagogram, manometry, and CT scan. An esophagogram using barium contrast helps in identifying the diverticula anatomical characteristics. A gastroscope is essential in visualizing the inner lining of the esophagus and identifying esophageal tumorstumours or strictures. Manometry is needed to confirm the presence of oesophageal motility disorder and is considered the ‘gold-standard technique.’ A CT scan can help identify and define a mass lesion. [3,4].

Esophageal diverticulum is commonly associated with motility problems such as achalasia and esophageal dilatation. The type of surgical intervention depends on several factors that ultimately are based on recurrence and whether the esophagus is salvageable.

Traditionally, symptomatic patients with isolated esophageal diverticulum are treated surgically by either an open approach or minimally invasive procedure, of which the latter can be either via video-assisted thoracoscopy or laparoscopy or via both in combination. The procedure includes diverticulectomy with or without cardiomyotomy and fundoplication. [3]. However, a diverticulectomy without myotomy has been reported to have a higher rate of postoperative leak, persistent symptoms, and diverticular recurrence. It has been reported that a minimally invasive approach to epiphrenic diverticulum reduces the risk of the length of hospital stay, morbidity, and mortality when compared with open procedures. [3,4].

Although there is a growing consensus that a myotomy should be performed with a fundoplication, this issue continues to be queried. Despite having no cause-effect connection demonstrable between epiphrenic diverticulum and hiatus hernia, they are, however, anatomical variations that can coincide with pathological acid reflux seen in pH studies but are poorly quantified. [5].

Patients with multiple esophageal diverticula are rare. In 2017, Taniguchi et al. reported a case of a patient with symptomatic multiple huge epiphrenic diverticula with manometry results showing high lower esophageal sphincter pressure and normal peristaltic contractions, surgically treated successfully with a minimally invasive esophagectomy. The team had considered that diverticulectomy had a high risk of leakage as the esophagus was dilated with high intra-esophageal pressure in addition to the multiple and large diverticula. [6]. Postoperative staple line leakage due to the increased intra-esophageal pressure, has been reported to occur 7.7 – 27.2% post diverticulectomy with myotomy for esophageal diverticulum. [7].

In our case, we evaluated clinically as evidenced by an ongoing obstruction, morphologically from gastroscope and contrast studies, and manometry for esophageal functionality. The gastroscope, esophagogram, and contrasted CT showed a giant epiphrenic diverticulum with dilated esophagus while manometry studies showed not only a raised lower esophageal sphincter pressure but a non-functioning esophagus with the absence of peristalsis. We considered that a diverticulectomy with myotomy alone had a high risk of recurrence and leakage, hence decided on a total esophagectomy as a better surgical method to achieve cure.

In a case series published in 2007 by Reznik et al., 39 patients with functional esophageal obstruction were reviewed between the years 1987 to 2005. . [9] Ppatients underwent esophagectomy successfully for unsalvageable esophagus caused by factors such as large diverticula, megaesophagus, history of previous hiatal hernia repair, and unsatisfactory diverticulectomy with myotomy. [2].

Minimally invasive esophagectomy is deemed safe and effective, hence it is the preferred technique for esophagectomy. Compared to open surgery, it results in a lower complication rate including wound infection, pulmonary complications, and reduced intraoperative blood loss. [8].

In conclusion, the main aim of surgically treating esophageal diverticulum is for achieving cure. The type of surgery preferred must take into account the morphology and functionality of the esophagus. Esophagectomy is another surgical choice for patients who are not suitable for diverticulectomy, myotomy, and relief of obstruction. A minimally invasive technique is preferred as it offers an advantage to open esophagectomy for the management of epiphrenic diverticulum, resulting in reduced length of stay, morbidity, and mortality. Despite the large experience with a rare case, there is a need for further case comparison to further assess the outcome in our local setting.

LIST OF ABBEVIATIONS

CT: Computerised Tomography; PG-SGA: Patient-Generated Subjective Global Assessment

DECLARATIONS

Ethics approval and consent to participate

This work adheres to the guidelines and principles of the Declaration of Helsinki and is in accordance with the Malaysian Good Clinical Practice (MGCP) 4th edition 2018. This research is also registered with the National Medical Research Register (NMRR), and ethics approval was obtained from the Medical Research and Ethics Committee (MREC) prior to the initiation of the study.

Consent for publication
Written consent was obtained from the patient.

Availability of data and materials
Not applicable.

Competing interests
The authors declare that they have no competing interests.

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Authors' contributions
CJ collected the patient's clinic information, searched relevant works of literature, and wrote the manuscript. ST was the attending doctor of the patient. ST, HAM, NRK & ATM carried out critical revision and correction of the manuscript. All authors read and approved the final manuscript for submission and publication.

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