DIFFICULT HELLER MYOTOMY DUE TO PROMINENT LARGE LEFT LIVER LOBE

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ABSTRAK

Achalasia Esofagus adalah gangguan *motilitas esofagus* yang ditandai dengan kegagalan peristaltik dan ketidakmampuan *sfingter esofagus* bagian bawah (LES) untuk relaksasi. Hal ini disebabkan oleh degenerasi *pleksus* saraf *mienterikus* pada dinding *esofagus.* Dalam kasus ini, kami melaporkan seorang pasien laki-laki berusia 24 tahun dengan *achalasia esofagus*. Tantangan teknis dalam kasus ini adalah adanya lobus hati kiri yang besar dan menonjol. Pasien laki-laki berusia 24 tahun tersebut dirawat di Rumah Sakit Umum Prof. IGNG Ngoerah dengan keluhan utama *disfagia*. Pasien mengeluhkan kesulitan menelan selama tiga bulan terakhir. Awalnya pasien merasakan ada yang tersangkut setiap kali makan dan minum. Keluhan tersebut memburuk dalam sebulan terakhir hingga pasien tidak dapat menelan makanan padat sama sekali. Pada saat masuk rumah sakit, pasien masih bisa mengonsumsi makanan cair. Tidak ada kelainan yang ditemukan pada pemeriksaan fisik dan hasil laboratorium. Evaluasi *oesophagogram* menunjukkan hampir total *stenosis* pada *esofagus distal* yang mencapai setinggi CV Th12 yang menyebabkan dilatasi pada *esofagus proksimal*. Hasil tersebut juga menunjukkan adanya dilatasi pada *esofagus.* Kemudian, pasien menjalani prosedur *laparoskopi Heller Myotomy,* namun karena adanya lobus hati kiri yang besar dan menonjol, prosedur tersebut diubah menjadi laparotomi dengan *Dor fundoplikasi*. Pasien mengalami pemulihan tanpa komplikasi selama tindak lanjut. *Achalasia esofagus* berhasil diatasi dengan *Heller Myotomy*. Lobus hati kiri yang besar dan menonjol dapat menjadi tantangan dalam prosedur ini.

Kata kunci **:** *dor fundoplication, esophageal achalasia, heller miotomy*

ABSTRACT

Esophageal achalasia is a motility disorder of the esophagus, characterized by failure of peristalsis and the lower esophageal sphincter (LES) is unable to relax.[2] It is caused by degeneration of the myenteric nerve plexus in the esophageal wall. In this case, we present a 24-years-old male patient with esophageal achalasia. Technical challenge is this case is prominent and large left liver lobe. A 24 years old male was admitted to Prof. IGNG Ngoerah General Hospital with a chief complaint of dysphagia. The patient had difficulty of swallowing for three months. Initially the patient felt something was stuck every time the patient eat and drink. Complaints worsened in the last one month until the patient could not swallow solid food at all. Upon admission, patients can take liquid foods. There were no abnormalities on the physical examination, and laboratory results. Evaluation of oesophagogram showed near total stenosis of the distal esophagus projecting as high as CV Th12 which caused dilatation of the proximal esophagus. The result also showed dilatation of the esophagus. Then, the patient underwent laparoscopy Heller Myotomy but due to prominent large left liver lobe, the procedure converted to laparotomy with Dor fundoplication. The patient had an uneventful recovery during follow up. Esophageal achalasia was successfully treated with Heller Myotomy. Large and prominent left liver lobe can be challenging to overcome.

Keywords ***:*** *dor fundoplication, esophageal achalasia, heller miotomy*

INTRODUCTION

Esophageal achalasia is a disorder in the form of an inability of the valve in the gastroesophageal junction to relax, so that only a small amount of swallowed food can enter the stomach (Duffield et al., 2017). Esophageal achalasia is a motility disorder of the esophagus, characterized by failure of peristalsis and the lower esophageal sphincter (LES) is

unable to relax (Rachmanio & Alam, 2019). It is caused by degeneration of the myenteric nerve plexus in the esophageal wall (Nikaki et al., 2019a) . This condition leads to a progressive dilation of the esophagus above the tightened LES, resulting in symptoms such as difficulty swallowing (dysphagia), regurgitation of undigested food, chest pain, and weight loss. The exact cause of the degeneration is still unknown, but it is thought to involve autoimmune factors or chronic inflammation triggered by viral infections.4 Achalasia is a rare condition, with an estimated annual incidence of 1 in 100,000 people, and it can occur at any age but is most commonly diagnosed in middle-aged individuals. Early diagnosis and appropriate treatment, such as pneumatic dilation, Heller myotomy, or peroral endoscopic myotomy (POEM), are crucial to improve patient outcomes and prevent complications like esophageal perforation or aspiration pneumonia.

The prevalence of esophageal achalasia is around 10 cases per 100,000 population where the incidence ratio of this disease is the same between men and women. Even though this disease is rare, doctor still have to be able to recognized and treat this disease because the complications that arise from this disease are very life-threatening, such as respiratory tract obstruction (Riyadi et al., 2019). Therefore, it is very important for us to identify the diagnosis of esophageal achalasia and the appropriate treatment. This case report discusses a case of esophageal achalasia in a 24 year old male with no previous history of difficulty swallowing. Esophageal achalasia often presents with a gradual onset of symptoms, including dysphagia, regurgitation, and weight loss, but atypical cases without prior swallowing difficulties, as seen in this report, can delay diagnosis. In some instances, patients may only present with nonspecific symptoms such as chest pain or nocturnal coughing, which can mimic other conditions like gastroesophageal reflux disease (GERD) or cardiac issues.

Advanced diagnostic tools, such as high-resolution esophageal manometry, barium swallow studies, and endoscopy, are essential in confirming the diagnosis. Treatment approaches, including pneumatic dilation, Heller myotomy, or botulinum toxin injections, should be tailored to the patient's age, symptom severity, and overall health. The case of a 24-year-old male with no prior history of dysphagia underscores the importance of maintaining a high index of suspicion for esophageal achalasia, even in atypical presentations, to prevent complications and ensure timely management.

METHODS

This case report was based medical record on from a 24-years-old patient with complaints of difficulty swallowing through anamnesis, physical examination, and supporting examinations in to Prof. IGNG Ngoerah General Hospital.

RESULT

Case Presentation

A 24 years old male was admitted to Prof. IGNG Ngoerah General Hospital with a chief complaint of dysphagia. The patient had difficulty swallowing for three months. Complaints worsened in the last one month until the patient could not swallow solid food at all. Upon admission, patients can take liquid foods. The patient denies any weight loss, there is no problem speaking or breathing. The patient had no prior history of chronic disease such as hypertension and diabetes mellitus, also there were no allergies. However, the patient had a history of tonsillectomy procedure back in 2015.

The patient had normal vital signs. Physical examination of the patient revealed no abnormality, with BMI within normal range (24.49 kg/m2). Blood works were normal. Oesophagogram was done (09/12/23 at Prof IGNG Ngoerah General Hospital) (Figure 1) with

300 cc of water soluble contrast which had been diluted and swallowed slowly. The contrast appears to fill the esophagus smoothly. The contrast filled the pars cervicalis, pars thoracalis esophagus and filled part of the stomach. Irregularities were seen in some of the mucosal walls of the esophagus, narrowing projected at the level of CV Th12 along +/- 2 mm with the narrowest caliber measured +/- 0.8 mm accompanied by dilatation of the esophagus proximally with the widest diameter measured +/- 59 mm. There was still visible contrast flow to the distal aspect to fill the stomach. Esophageal peristaltic movements appeared good and no filling defects were visible. The results of the esophagography were concluded with near total stenosis of the distal esophagus projecting as high as CV Th12 which caused dilatation of the esophagus proximally, there was irregularity in some of the mucosal walls of the esophagus, suggesting a picture of esophagitis. Based on the results of the exams, we concluded the diagnosis as esophageal achalasia. The patient then decided to do Laparoscopy Heller Myotomy (LHM) with fundoplication.

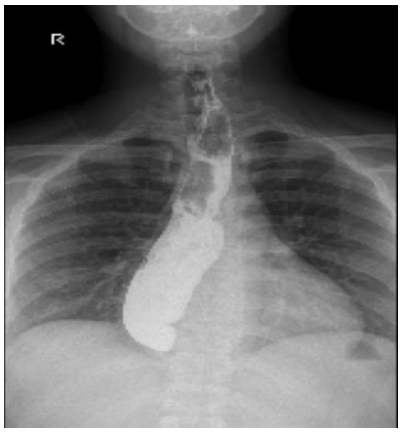

Figure 1. Oesophagogram Result ((09/12/23 at Prof IGNG Ngoerah General Hospital)

The patient then was prepared for surgery. During the surgery, he was in supine position. Disinfection and draping are carried out with a sterile sheet, then placed two supra umbilical and epigastric trocars. Evaluation showed large size of the liver, the hiatus is not visible because of the liver size even though the liver was lifted, the esophageal hiatus was not visualized well, then it was converted to laparotomy (Figure 2). It started with upper midline abdominal incision, deepening layer by layer. Then mobilizing the left lobe of the liver by dividing the triangular ligament to expose the lower esophagus. After that, the peritoneum was separated over the esophagus and stomach, clamped the gastrohepatic ligament to allow mobilization anterior to the esophagogastric junction. Next, cut the phrenoesophageal ligament and esophageal fat pad, open the fat pad to reveal a healthy esophagus and the narrowed part (Figure 3). A myotomy is performed by dividing the circular and longitudinal muscle fibers proximal to the narrowing, extend the incision 6 cm proximally and 3 cm distally, performed a puncture on the esophagus, entered a 24 Fr Folley Catheter, did a dilatation with a balloon. Then fundoplication was performed. Treat bleeding, wash the surgical wound afterwards, installed a subhepatal drain with NGT No. 18 Fr, set the wound layer by layer, and the surgery was done.

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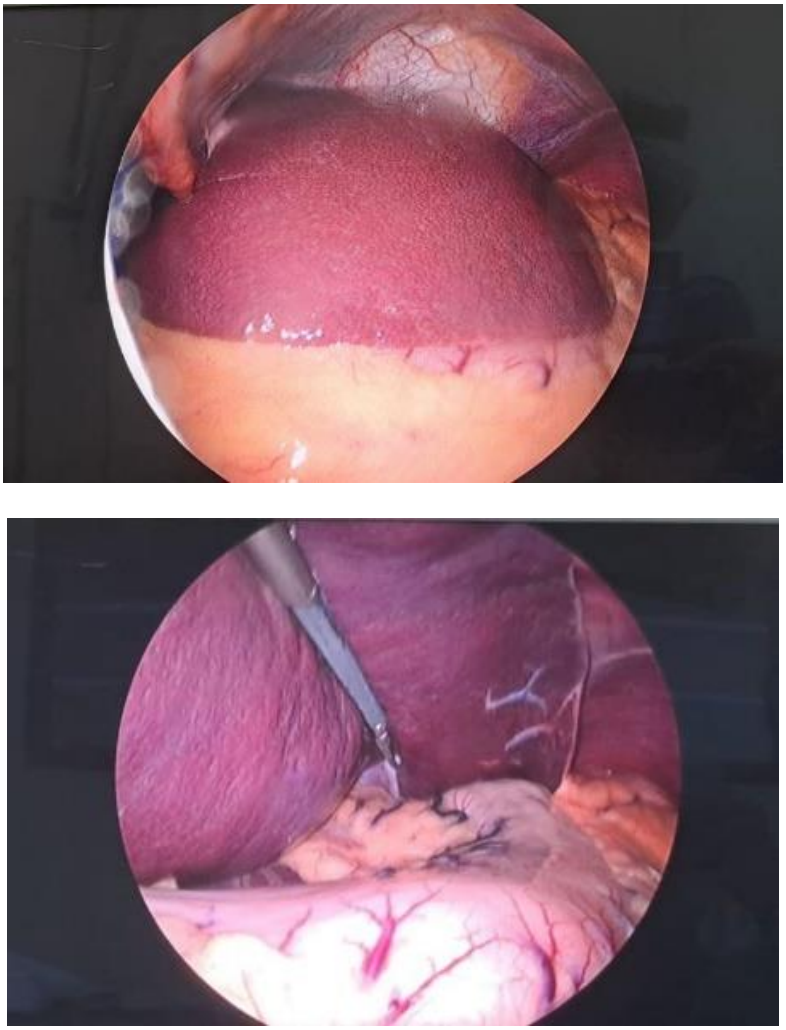
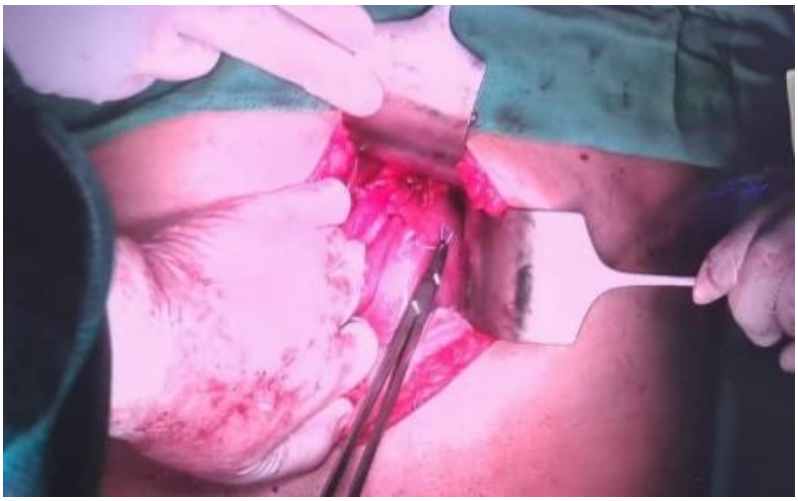


Figure 2. Durante Op (Laparoscopy Showed Large Liver That Covered the Diaphragm)



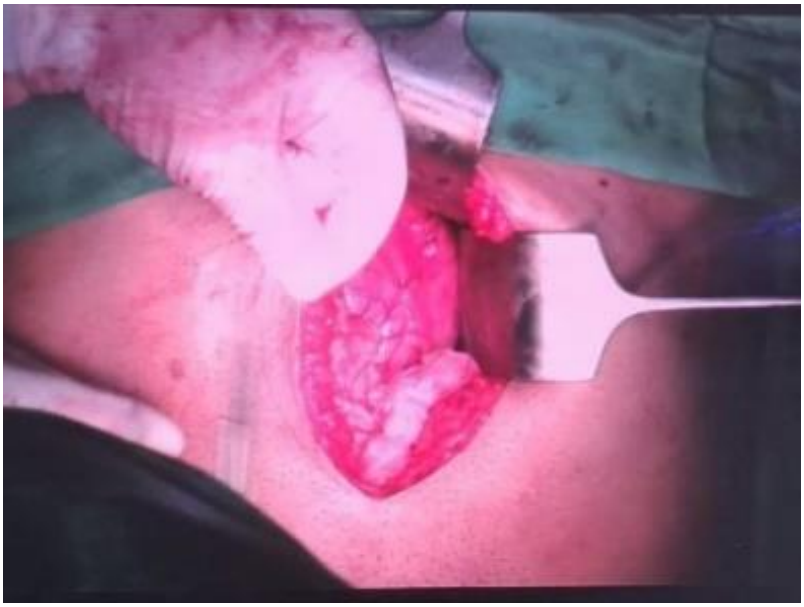


Figure 3. Durante Op (Laparotomy)

After surgery, the patient was given liquid diet and the calorie requirement of 2250 kcal/24 hours was administered via NGT. The patient showed clinical improvement after being treated and was discharged in fifth day. The patient was planned for follow up in the outpatient clinic.

DISCUSSION

In a normal esophagus, peristaltic waves appear every time it swallows something. In achalasia there is a neuromuscular defect characterized by decreased or absent peristalsis. Microscopically, there is visible degeneration of ganglion cells in Auerbach's myenteric plexus. This condition is analogous to megacolon. Achalasia may progress slowly. The incidence of achalasia is very rare, namely 1 in 100,000 people with a prevalence of 10 in 100,000. This occurs most often in individuals aged 40 or older (Ramirez & Patti, 2015). When peristaltic movements pass through the esophagus during swallowing, the receptive relaxation process relaxes the lower esophageal sphincter before the peristaltic movement and allows food to enter the stomach. When the sphincter does not relax, achalasia will occur (Shofiyanti et al., 2023)

The diagnosis of achalasia should be suspected in patients who present with complaints of difficulty swallowing solid or liquid food. Symptoms that may arise are dysphagia, reflux or regurgitation, heartburn, and weight loss.[3] The results of esophageal motility tests in achalasia patients can show that the lower esophageal sphincter pressure does not return to normal at rest and cannot relax properly after swallowing food (Pressman & Behar, 2017) In this case, a 24 year old male patient came with complaints of dysphagia. The patient had difficulty swallowing for eight days. Complaints worsened in the last one month until the patient could not swallow solid food at all. Upon admission, patients can eat soft and liquid foods. The patient denies any weight loss.

On the patient's physical examination, no abnormalities were found, and the patient's BMI was still within normal limits. In cases of achalasia, usually no distinctive features are found on physical examination. So, the doctor will recommend carrying out supporting examinations. Next, the doctor can carry out supporting examinations, such as esophagography**,** to get a detailed picture of the esophagus, stomach and intestines by drinking Barium liquid, then there is manometry, to measure the flexibility and strength of contraction of the esophagus muscles when swallowing, and endoscopy to check the condition of the walls of the esophagus and stomach (Nikaki et al., 2019b). A characteristic finding in manometry is low resting pressure,

but high in the LES because of the failure of the LES to relax when swallowing and reduction in the amplitude of peristaltic waves along the esophagus (Carlson & Pandolfino, 2020)Chest X-Ray can also be used as a supporting examination, although it is less recommended because it does not produce specific results (Kuriyama et al., 2021). Chest X-ray of patients with achalasia can show a dilated esophagus and air-fluid levels. Using barium will show a tortuous and widened esophagus, sometimes narrowing at the cardia like a bird's beak(Abud et al., 2016) In this patient, an oesophagogram was performed with 300 cc of water soluble contrast which had been diluted and swallowed slowly. The results of the esophagography were concluded with near total stenosis of the distal esophagus projecting as high as CV Th12 which caused dilatation of the esophagus proximally, there was irregularity in some of the mucosal walls of the esophagus, suggesting a picture of esophagitis. The picture also showed dilatation of the esophagus.

It is important to know that achalasia is a chronic condition that cannot be cured. The ultimate goals of therapy include reducing symptoms, improving esophageal emptying, and preventing further esophageal dilatation. Current treatment options for achalasia aim to reduce LES hypertonicity. Currently available achalasia treatment options include pharmacological, endoscopic and surgical methods. Esophageal achalasia is a rare motility disorder characterized by the inability of the lower esophageal sphincter (LES) to relax due to degeneration of the myenteric nerve plexus, leading to progressive dysphagia, regurgitation, and other symptoms such as chest pain or weight loss. This case discusses a 24-year-old male patient who presented with a chief complaint of dysphagia for the past three months that progressively worsened to the point where he could only swallow liquids. Radiologic evaluation via esophagogram showed near-total stenosis of the distal esophagus with proximal dilatation, reflecting the classic presentation of achalasia. Although the patient did not show any significant weight loss or abnormalities on physical examination, imaging and evaluation of esophageal function provided a convincing diagnosis. For treatment, the patient was scheduled to undergo Heller Myotomy with Dor fundoplication, which is the gold standard for achalasia. However, challenges arose during laparoscopy due to the large and prominent left lobe of the liver, which obscured the visualization of the esophageal hiatus despite liver lifting. This situation necessitated conversion to laparotomy, which provided better access with mobilization of the liver lobe through division of the triangular ligament. This conversion reflects the importance of flexibility and technical adaptability in dealing with unusual anatomy during surgery. Heller Myotomy was successfully performed by extending the incision proximally and distally to the esophagus to overcome the stenosis, followed by Dor fundoplication to prevent postoperative gastroesophageal reflux complications. The patient had an uneventful recovery and was discharged on the fifth postoperative day. This case highlights the importance of early diagnosis and appropriate surgical approach to overcome the technical challenges in the treatment of esophageal achalasia.

Pharmacological therapy is the least effective therapy in the treatment of achalasia. Examples are calcium channel blockers, and nitrates, which are believed to release nitrous oxide which can cause LES relaxation. Other drugs that are rarely used are anticholonergics, but they cause many side effects such as headache, hypotension, and edema (Vaezi et al., 2020). Heller's myotomy is one of the surgical procedures for achalasia patients. In this surgical process, an incision is made in the esophageal muscle in the valve area between the esophagus and the stomach down to the submucosa of the esophagus (Swanström, 2019). There are several options available, such as esophagectomy. Esophagectomy is performed if previous treatment is unsuccessful,so it is not suitable for this patient who had no history of achalasia treatments before(Wadhwa et al., 2017). This patient underwent surgery, using Laparoscopy Heller Myotomy with Dor fundoplication. However, during the laparoscopy, it was found that the patient’s liver were large and prominent, covering the hiatus area eventhough the liver was

lifted, hence the procedure was converted to laparotomy. A complication of achalasia that can develop as a result of the natural course of the disease is high aspiration due to the uncertain presence of food in the esophagus. If not treated quickly, achalasia patients can also experience complications such as symptomatic gastroesophageal reflux, esophagitis and can even cause death (Bright, 2020) These patients are given appropriate calorie needs to prevent nutritional deficiencies, and are given antibiotics for post-surgery infection prophylaxis. The rest are supportive medications such as PPIs for prophylaxis of increased stomach acid and to prevent stress-related mucosal bleeding (Mari et al., 2021).

CONCLUSION

Achalasia is an esophageal motility disorder that causes disruption of peristalsis in the esophagus and malfunction of the esophageal sphincter which is characterized by difficulty swallowing. Even though the incidence of achalasia is very rare, if it is not treated, it can result in aspiration and even death. Therefore, a diagnosis is needed in order to carry out effective and safe surgical treatment for achalasia patients. Heller myotomy and fundoplication is still considered effective treatment for esophageal achalasia. Large and prominent left liver lobe can be challenging to overcome, if liver lifting is inadequate, consider laparotomy.

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