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Diagnosis and Management of Achalasia : A Case Report

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ABSTRACT

Background: Achalasia is a condition characterized by the absence of peristaltic in the lower esophageal body and hypertonic of the lower esophageal sphincter (LES), caused by degeneration of the myenteric plexus ganglia, leading to inadequate relaxation during swallowing.

Case Presentation: A 33-year-old woman complained of dysphagia when consuming solid or liquid food since last year. The patient also reported heartburn and significant weight loss. Chest X-ray finding was unremarkable. However, endoscopic examination revealed a narrowed esophagus with food residue and a rosette appearance. Contrast-enhanced abdominal computed tomography (CT) scan showed luminal obstruction at the level of CV Th12, resulting in dilation of the proximal esophagus and food retention. After failure of oral medication therapy, definitive therapy in the form of heller myotomy was performed. The patient showed significant improvement after the procedure with good oral intake tolerance.

Discussion: The exact etiology of achalasia remains unknown. The gold standard for diagnosing achalasia is manometry, but this modality is not available. Diagnosis of achalasia in this patient started with clinical symptoms of dysphagia, heartburn, and weight loss, supported by endoscopic and contrast-enhanced abdominal CT scan findings. Other management options include pneumatic dilatation and peroral endoscopic myotomy (POEM), but heller myotomy remains the recommended management with a higher success rate.

Conclusion: Achalasia is a rare motor disorder of the esophagus and the exact etiology of achalasia remains unknown. The diagnosis of achalasia was concluded from clinical symptoms, physical examination, and supportive investigations (endoscopic examination and contrast-enhanced abdominal computed tomography). Management of achalasia in this patient consisted of administering oral medication and definitive therapy in the form of heller myotomy with good results.

Keywords: *Achalasia, endoscopy, heller myotomy, lower esophageal sphincter*

INTRODUCTION

Achalasia is derived from the Greek word that means unable to relax.[1] Achalasia is characterized by the absence of peristalsis in the lower esophageal body and hypertonic lower esophageal sphincter (LES) which disable complete relaxation during swallowing.[2] This abnormality is suspected to be caused by the degeneration of myenteric plexus ganglia. The classic appearance of achalasia is hypertensive LES and no LES relaxation which leads to food stasis and esophageal dilatation.[3]

Achalasia is rarely found. It does not affect certain ages, races, or gender.[4] The incidence of achalasia between men and women is similar, which is 1 out of every 100,000 lives per year with a prevalence of 10 out of 100,000 lives, found in the 30-60 years group.[2,5,6] Data from the gastroenterology division of FKUI/RSCM Department of Internal Medicine found 48 cases within 5 years (1984-1988).[3] There is no achalasia epidemiology data in Indonesia yet, only case reports or small-scale studies in certain hospitals.[2]

The etiology of achalasia is still unknown.[5] It is suspected to be caused by primary and secondary causes. The former may be caused by a neurotropic virus which forms a lesion in the dorsal vagal nucleus in the brain stem and esophageal myenteric plexus ganglia. The latter can be caused by infections, such as the Chagas disease (*Trypanosoma cruzi* infection), intraluminal tumors such as cardiac tumors, or extraluminal such as pancreatic pseudocysts, anticholinergic drugs, and post vagotomy.[2-4,7]

Achalasia is suspected to occur due to an imbalance of parasympathetic excitatory and inhibitory pathways which innervate LES smooth muscles. Excitatory neurotransmitters include substance P and acetylcholine, and inhibitory neurotransmitters are vasoactive intestinal peptide (VIP) and nitric oxide (NO). The role of inhibitory neurotransmitter is to modulate LES pressure and relaxation. Imbalance causes hypertensive and not relaxed LES.[4,5] Another study found that viral infection trigger autoimmune process, which will trigger inflammatory cascade that damage and cause the lost of nitrite oxide release in myenteric plexus.[8]

The primary clinical symptom is dysphagia for both solid and liquid food. Other common symptoms include regurgitation, weight loss, and substernal chest pain.[2,3] Regurgitation can occur immediately or several hours after eating, especially when the patient is lying down. The

food regurgitated has not been digested. Chest pain is caused by esophageal irritation due to stomach acid or toxin resulting from lactate fermentation by bacteria in the esophagus.[2]

Some uncommon symptoms include hiccups or difficulty burping. Complication due to food retention is shown by coughing and aspiration pneumonia. Physical examination provides little help.[3,4] Common extraesophageal symptoms include structural or functional lung abnormalities due to repeated aspiration or tracheal compression from a dilated esophagus. A bullfrog neck sign can appear due to severe dilatation and cervical esophagus distortion, obstructing the trachea and stridor.[4]

Several adjunctive examinations can aid in establishing the diagnosis of achalasia, including radiological examination (esophagogram), upper gastrointestinal endoscopy, and manometry.[3-5] Esophagogram examination will show winding and elongated esophageal dilatation with a tapered distal end and a smooth surface like a bird's beak.[3] Several cases show air-fluid levels and no intragastric fluid.[4]Aside from diagnostics, endoscopy can also be used for therapy. Most patients show normal mucosa, but some have mild hyperemia to diffused distal esophagus and white spots, erosion, or ulcers in the mucosa due to food retention. Endoscopy is recommended for all achalasia patients to rule out pseudoachalasia.[1,2] Esophageal manometry is the most sensitive test to diagnose achalasia and is the gold standard. Manometry will show incomplete LES relaxation as a response to swallowing, lack of peristalsis in the lower esophageal body, and increased LES pressure.[4]

Achalasia treatment is aimed to improve LES relaxation function through oral medicaments, LES dilatation or stretching, esophagotomy, and injection of botulinum toxin (botox) to the esophageal sphincter.[1,2] The oral medicaments used include nitrate (isosorbide dinitrate), calcium channel blockers (CCB) such as nifedipine and verapamil, and phosphodiesterase-5 inhibitors (sildenafil).[4] CCB group and nitrate are usually consumed 30-60 minutes before meal. CCB works by inhibiting calcium absorption by cells because intracellular calcium is required for LES contraction to induce relaxation.[7] Nitrate works against the reduction of the inhibitory neurotransmitter, nitric oxide. This reduces LES tone, followed by reduced LES pressure.[4,7]

In achalasia, transit time is longer and esophageal emptying is delayed. Thus, the absorption and effectivity of oral medications cannot be calculated. These drugs should be used sublingual,

such as sublingual nifedipine 10-30 mg for 30-45 minutes before meal and sublingual ISDN 5 mg 10-15 minutes before meal. These medications reduce LES pressure to 50%.[2] Oral medicaments aim to relax LES. However, this therapy can only effectively reduce LES pressure and not improve relaxation.[2] Oral medicaments are mostly non-responsive and have many adverse effects, such as peripheral edema, headache, hypotension, and short-term use.[1,2] The indication of oral medications is early condition without esophageal dilatation, patients refusing invasive procedures, and severe achalasia with chest pain.[3]

Incremental dilatation treatment will temporarily reduce symptoms. A simple approach is using Hurst's Mercury Tube. The success of this treatment is 50% without recurrence, 35% with recurrence, and 15% failure. Pneumatic dilatation is more recommended with the best results in 75-85% of cases. Complication is rare, in the form of esophageal perforation. Surgical intervention is prepared if perforation occurs.[2,3,5] Gastroesophageal reflux is the most common chronic complication after pneumatic dilatation. Therefore, PPI is recommended.[5,7]

Esophagomyotomy (Heller procedure) incises circular muscles surrounding LES. Laparoscopic Heller Myotomy (LHM) is always followed by an anti-reflux procedure to prevent chronic gastroesophageal reflux.[4,9] Conventional surgery involves a long incision in the chest. However, laparoscopy has been used as a minimally invasive concept similar to an open surgery.[2] Peroral Endoscopy Myotomy (POEM) is an effective minimally invasive alternative. Dissection of LES circular fibers is performed endoscopically, which causes LES relaxation. However, it includes a higher gastroesophageal reflux risk due to no anti-reflux procedure.[4,10]

Treatment with Botulinum Toxin injection to weak LES via endoscopy is a safe therapy, albeit short-term, and requires repeated injections.[3] The recurrence rate is higher than 50% within 6 months. Botulinum Toxin-A used for achalasia therapy works by breaking SNAP-25 protein molecules in presynaptic membranes, inhibiting acetylcholine release and exocytosis to synaptic areas. This will cause temporary muscle weakness by inhibiting cholinergic stimulation to LES.[2,5] This choice of therapy is beneficial for patients with a high risk of surgery or the elderly.[3]

CASE PRESENTATION

A 33-year-old woman reported to the Emergency Department of Bangli Regional Hospital with her family with a complaint of continuous nausea and vomiting. This complaint aggravated the last three days. Vomiting occurs after eating and drinking with an interval of 2-5 minutes. The vomit includes food waste and liquid. The patient felt burning around her epigastrium with each vomit. She tends to vomit when consuming solid food. However, vomiting reduces when consuming liquid food. This has occurred for 1 year. The patient also complained of drastic weight loss from 60 kg to 32 kg. Other symptoms include chest pain, epigastric pain, infrequent bowel movement, less than 1 in 3 days. The patient denied having fever, prolonged cough, shortness of breath, and other chronic diseases.

Physical examination found good consciousness and orientation. Her blood pressure was 102/86 mmHg, pulse 82 x/minute, breathing 20 x/minute, axillary temperature 36.3°, and SpO_2 97% at room air. Based on nutritional status, the patient was considered underweight with body mass index (BMI) of 14.2 kg/m^2. Physical examination in general is within normal limits with pressure pain in the epigastric region. Laboratory examination found leukocytes at 8.55 10^3/uL (3.5-9.5 10^3/uL), hemoglobin at 14.8 g/dL (11.5-15 g/dL), hematocrit at 43.2% (35-45%), and platelets at 415 10^3/uL (150-350 10^3/uL). Electrolyte assessment found blood sodium level at 140.9 mmol/L (136-145 mmol/L), blood potassium level at 3.87 mmol/L (3.5-5.5 mmol/L), and blood chloride level at 98.5 mmol/L (96-108 mmol/L). The random blood glucose test was 77 mg/dl (70-140 mg/dl). Kidney function test showed ureum level at 19 mg/dL (15-40 mg/dL) and creatinine level at 0.48 mg/dL (0.5-0.9 mg/dL). Mehran score was 1, which is a low risk of contrast-induced nephropathy (CIN).

Thorax radiograph found no abnormalities to the heart with normal bronchovascular markings. No gastric bubble was found. Endoscopic examination found a narrowed esophagus with food residues and a rosette sign. The stomach and duodenum could not be evaluated.

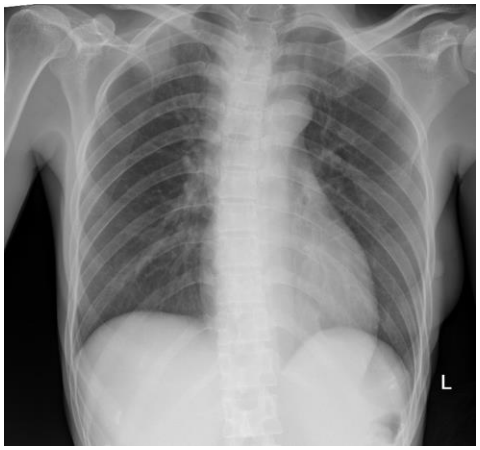

Figure 1. Chest x-ray showed absence of gastric air bubble

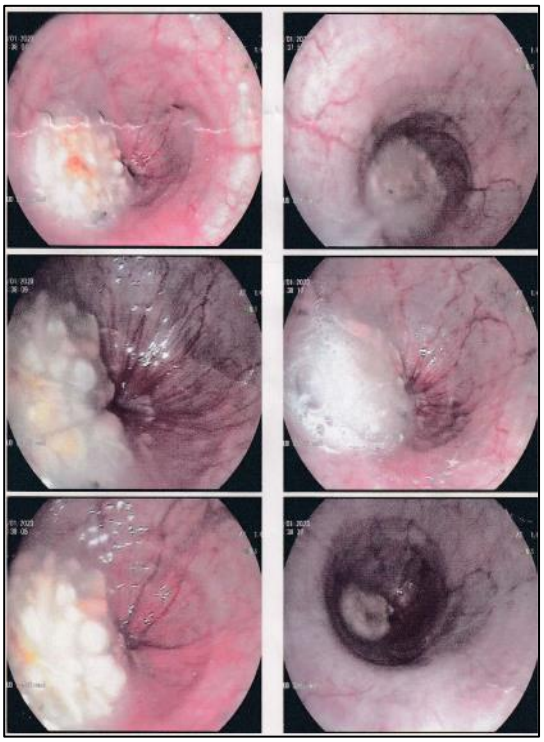

Figure 2. Endoscopic examination showed narrowed esophagus with food residue

The patient was hospitalized in Bangli Regional Hospital. During the stay, she was given intravenous NaCl: D5%: Aminofluid = 1:1:1-20 tpm, lansoprazole inj 30 mg/24 hours iv, ondansetron inj 8 mg/8 hours iv, mecobalamin inj 500 mg/24 hours iv, nifedipine 10 mg/12 hours sublingual, and sucralfate syrup 3x10 cc per oral along with observation of general condition, vital signs, and symptoms.

An abdominal computerized tomography (CT) scan was performed with contrast. The result represented achalasia which creates lumen obstruction as high as CV Th12 and causes

dilatation to proximal esophagus and food retention. The patient was referred to another hospital for definitive surgery of Heller Myotomy. She is a good candidate for definitive surgery.

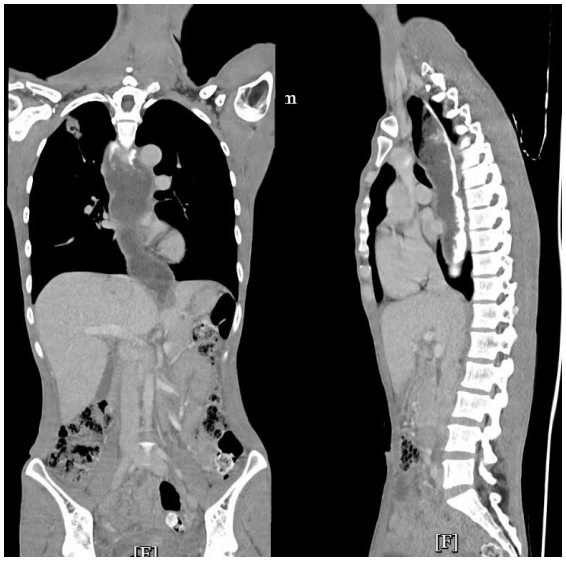

Figure 3. Contrast-enhanced abdominal computerized tomography (CT) scan showed lumen obstruction as high as CV Th12

After surgery, the patient was hospitalized for 3 days in Prof. dr. I.G.N.G. Ngoerah Central Hospital and received a clear fluid diet and milk for 6x100 cc, asering iv 20 tpm, cefoperazone 1 gram/12 hours iv. After treatment, the patient was discharged in good condition and had no difficulty swallowing or vomiting after eating.

DISCUSSION

Achalasia is a condition characterized by the absence of peristalsis in the lower esophageal body and hypertonic lower esophageal sphincter (LES).[2] It is rare with an unknown underlying cause.[4] In this case, the patient was feeling nauseous and continuously vomiting solid or liquid food. Vomit occurred after every meal and drink with an interval of 2-5 minutes. The regurgitated substance was undigested food. The symptoms were accompanied by weight loss and chest pain. The main clinical symptom found in achalasia is dysphagia for solid and liquid food. Other common symptoms include regurgitation, weight loss, and substernal chest pain.[2,3] The patient's chest pain was caused by esophageal irritation due to stomach acid or toxin produced by lactate fermentation by bacteria in the esophagus.[2] Complications due to food retention in the form of aspiration pneumonia were not found in this case.

The recommended adjunctive examination for achalasia is radiology (esophagogram), upper gastrointestinal endoscopy, and manometry.[3-5] Manometry, the gold standard examination, cannot be performed in this case due to unavailability. Endoscopy result on the patient found narrowed esophagus, food residues, and a rosette sign. Endoscopy is recommended for all achalasia patients to rule out the possibility of pseudoachalasia.[1,2] An abdominal computerized tomography scan (CT scan) with contrast showed lumen obstruction as high as CV Th12, causing dilatation to the proximal esophagus and food retention.

Achalasia treatment is aimed at improving the relaxation function of LES through oral medicaments, dilatation, LES stretching, esophagotomy, and botulinum toxin (botox) injection to the esophageal sphincter. The newest approach is the peroral endoscopy myotomy (POEM) method. [1,2]

This patient was treated with oral medicaments, nifedipine 10 mg every 12 hours sublingually to relax and reduce LES pressure. The indication for oral medicaments in achalasia patients is chest pain, early condition, or refusing invasive procedures.[3] Nifedipine works by inhibiting calcium absorption by cells because intracellular calcium is required for LES contraction to induce relaxation.[7] Sublingual administration was chosen because achalasia causes longer transit and delayed esophageal emptying, which inhibit the absorption and effectivity of oral medications.[2] However, oral medications were not effective in this patient. No adverse effects such as leg edema, headache, and hypotension found in oral medications. Studies showed that oral medication is only effective in reducing LES pressure and not increasing relaxation.[2]

Heller Myotomy is a procedure involving the incision of circular muscles surrounding LES. This procedure was chosen for this patient due to high success always accompanied by anti-reflux procedure to prevent chronic gastroesophageal reflux seen in other procedures.[4,9]

CONCLUSION

Achalasia is a rare case with an unknown underlying cause. This patient was diagnosed based on clinical symptoms, and physical and adjunctive examination (endoscopy and abdominal CT scan with contrast). The management of achalasia in this patient includes oral medications and definitive therapy of Heller myotomy with good results.

CONFLICT OF INTEREST

The authors affirmed that there were no conflicts of interest in this study.

FUNDING

The authors were responsible for all research funding without obtaining financial support.

ETHICAL CLEARANCE

Authors have secured informed consent regarding patient medical records for this case report.

AUTHOR CONTRIBUTION

All authors contributed equally in this research and publication of this manuscript.

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