Approach for Diagnostic and Treatment of Achalasia

Andree Kurniawan, Marcellus Simadibrata**,*
*Prima Yuriandro***, Lie Khie Chen*****
* Department of Internal Medicine, Faculty of Medicine, University of Pelita Harapan
Siloam General Hospital Karawaci, Tangerang
** Division of Gastroenterology, Department of Internal Medicine, Faculty of Medicine
University of Indonesia/Dr. Cipto Mangunkusumo General National Hospital, Jakarta
*** Department of Internal Medicine, Faculty of Medicine, University of Indonesia
Dr. Cipto Mangunkusumo General National Hospital, Jakarta
**** Division of Tropical and Infectious Disease, Department of Internal Medicine
Faculty of Medicine, University of Indonesia
Dr. Cipto Mangunkusumo General National Hospital, Jakarta

ABSTRACT

Achalasia is a rare motor disorder of the esophagus and lower esophageal sphincter. The incidence is approximately 1/100,000 per year and the prevalence rate is 10/100,000. Achalasia is quite difficult to establish because the symptoms might be insidious and therefore not many people come to seek medical attention until it deteriorates to final stage of the disease. There are several modalities that can be used as diagnostic tools such as manometry, barium esophagogram, esophagoduodenoscopy, esophageal CT-scan, until the recent one, high-resolution manometry that can classify achalasia into three different types. The treatment options are the pharmacologic intervention, endoscopic treatment, minimal invasive surgery, and radical surgery.

We reported a case of 20 year old female with achalasia who came with dysphagia symptom since three years before. The diagnosis was made by historytaking, physical examination and barium meal and esophagogastroduodenoscopy. The patient underwent pneumatic dilatation and since then the symptom was relieved.

Keywords: *achalasia, diagnostic, treatment*

ABSTRAK

Akalasia merupakan gangguan motorik esophagus dan sfingter esofagus yang jarang. Insiden berkisar 1/100,000 pertahun dan angka prevalensinya 10/100,000. Gejala awal penyakit ini terselubung karena itu pasien baru berobat setelah stadium lanjut. Terdapat beberapa pilihan diagnosis seperti manometri, barium esofagogram, esofagoduodenoskopi, CT-scan esophagus dan akhir-akhir ini manometri resolusi tinggi dapat mengklasifikasikan akalasia menjadi berbagai tipe. Pilihan terapi akalasia antara lain intervensi farmakologi, terapi endoskopi, bedah minimal dan radikal.

Dilaporkan seorang perempuan usia 20 tahun dengan akalasia yang datang dengan gejala disfagia sejak tiga tahun sebelumnya. Diagnosis ditegakkan berdasarkan anamnesis, pemeriksaan fisik dan pemeriksaan barium meal, serta prosedur esofagogastroduodenoskopi. Setelah pasien menjalani tindakan dilatasi pneumatik kondisinya membaik tanpa keluhan lebih lanjut.

Kata kunci*: akalasia, diagnostik, tatalaksana*

INTRODUCTION

Achalasia is a rare motor disorder of the esophagus and lower esophageal sphincter. Primary idiopathic achalasia is a quite rare disease, with an incidence of approximately 1/100,000 per year and a prevalence rate of 10/100,000.[1] In Indonesia there were only 48 cases during 5 years period (1984-1988) that were documented by Division of Gastroenterology, Department of Internal Medicine, Cipto Mangunkusumo Hospital.[2]

Achalasia is quite difficult to establish because the symptoms might be insidious and therefore, not many people come to seek medical attention until it deteriorates to the final stage, which more drastic intervention such as esophagectomy. There are several modalities that can be used as diagnostic tools. There are esophageal manometry, barium esophagram, esophagoduodenoscopy, esophageal CT-scan, until the recent one, high-resolution manometry, which can classify achalasia into three different types (type I, II and III).[3,4] It is important that we can diagnose achalasia in its early stage, so we can reduce the morbidity as well as cost-burden for the patient.

Those are several treatments available for achalasia nowadays. The options are the pharmacologic intervention, endoscopic treatment, minimal invasive surgery, and radical surgery. Each of the treatment has its own advantages, so as a clinician we must know what the best treatment of choice for each patient considering patient's characteristics included age, the severity of the illness, previous treatment, and socioeconomic status.

CASE ILLUSTRATION

A 20 year old female, came with a chief complaint of experiencing difficulties in swallowing since three years before admission. The patient was referred from Abdul Moeloek Hospital Lampung to undergo endoscopic pneumatic dilatation in Cipto Mangunkusumo Hospital Jakarta. Since five years before admission, patient had complained frequent runny nose, even though she did not have any common cold, mucus came out from her nose especially when lying on side. The patient consulted to the doctor and she was given some medications for sinusitis, including anti allergic and antibiotic but she did not feel any improvement. Since three years before admission, the patient experienced difficulties in swallowing solid and soft food. There was a discomfort sensation, dull pain inside the chest, and sensation like something was stuck inside her chest every meal.

Since one year before admission, the patient began to experience difficulties in swallowing water as well as solid food, frequently experience dry cough, and felt like something crawling up her throat with like burn sensation. The patient coughed especially when lying on back. Later on that year, the patient coughed some blood out and was diagnosed as a tuberculosis (TB) case and was given TB medication for almost 2 months and her doctor stop the medication because she did not have any improvement. Since ten months before admission, the patient had more difficulties in swallowing. She had to flush the food with water in order to swallow, frequently vomited after eating or drinking and began to lost her appetite. The patient only eat baby food and she lost about 40 kg in 2 years.

One month before admission, the patient went to Abdul Moeloek Hospital and being told to undergo a radiologic examination and an endoscopic procedure. Later, the doctor told that she had some narrowing in her esophagus and being referred to Cipto Mangunkusumo Hospital for further treatment.

In the past history of illness, the patient never had any history of exposure to chemical material, no history of another cause for hospitalization, asthma nor allergic to food or medication and any lung disease. In family history, her mother has hypertension but no history of diabetes, malignancy, or similar illness. The patient did not smoke, no use of alcohol, drugs, and herbal medicine.

From physical examination, we found that patient looked moderately ill with vital sign blood pressure 110/60 mmHg, pulse rate 84 times per minutes, respiration rate 18 times per minutes, body temperature 36.7^{o}C, normal color of conjunctiva and sclera and jugular venous pressure was 5-2 cmH_2O. On chest examination, lung and heart were unremarkable. From abdominal examination, we found that the abdomen was flat, smooth, no tenderness on epigastrium. Liver and spleen were not palpable and bowel sound was normal, there was no shifting dullness on examination. On the extremities there was edema in both legs, felt warm and capillary refill time was below 3 seconds.

From laboratory examination, it was found that hemoglobin level was 12.8 g/dL, hematocrit 38%, platelets 234,000/mm^3, leukocyte count 8,630/mm^3 with differential count basophil 0.2%, eosinophils 1.7%, neuthrophils 73.5%, lymphocytes 10.9%, monocytes 15.7%, erythrocyte sedimentation rate 7 mm/hour, ureum 27 mg/dL, creatinin 0.7 mg/dL, AST 20 U/L, ALT 23 U/L, random blood glucose 98 g/dL,

natrium 140 mEq/L, kalium 3.55 mEq/L, chloride 106mEq/L, coagulation result was normal.

From chest X-ray examination, we found that the cardio thoracic ratio was less than 50%, there were no infiltrations, and there were no sign of pleural effusions. From the electrocardiography there was no abnormality. The result of barium meal examination from the hospital before (Figure 1), there was a narrowing in the distal esophagus and the contrast could only pass through it slightly, there was also a 'beak shape' appearance and the esophagoduodenoscopy (EGD) which was performed that the mucosa was normal, the vascularization was also normal, the lower esophageal sphincter (LES) was narrowing, but the scope can still went through LES. The impression from those examinations was achalasia.

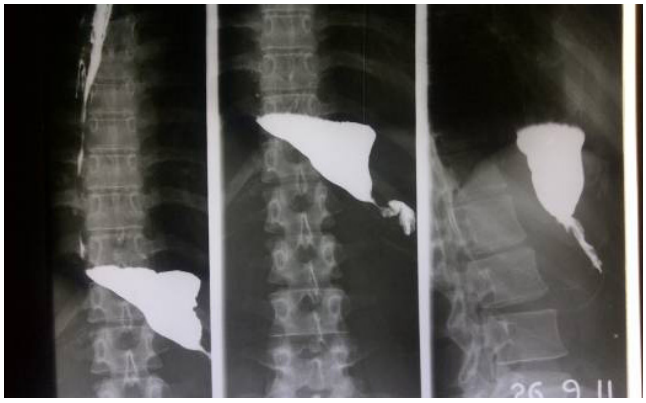

Figure 1. Barium meal examination revealed a narrowing in the distal esophagus and the contrast could only slightly pass through and also a 'beak bird shape' appearance

From the history taking, physical examination and radiologic procedures the patient was diagnosed with achalasia and the patient was scheduled to undergo an endoscopic dilatation procedure using a pneumatic balloon. We tried to alleviate the symptoms of dysphagia, regurgitation and the burn sensation inside the chest by giving medications such as nifedipine 10 mg three times a day before meal, omeprazole 20 mg twice a day, and sucralfate four times a day. The patient only felt slight improvement and still had a rough time in swallowing compact food. Therefore, we changed the diet into the combination between soft diet and milk, combining 1,900 kcal/day. The patient response was quite good.

The first time the patient went for the pneumatic dilatation (PD) (Figure 2), it failed because the scope could not get a clear view because the patient still had many leftover of food inside her esophagus, proximal from the LES, and the patient was hard coughing, so the procedure was canceled and rescheduled. The patient then began to fast 2 days prior to the procedure, we gave total parenteral nutrition at that time and we put a nasogastric tube in the middle of patient's esophagus with flushing 3 times a day, to clear out any food left over.

The patient then underwent the PD with good preparation and it succeeded. The dilatation was done by the balloon. Post dilatation,the stenosis region disappeared and there was only small amount of bleeding. The patient was scheduled for an evaluation 1 month later after PD.

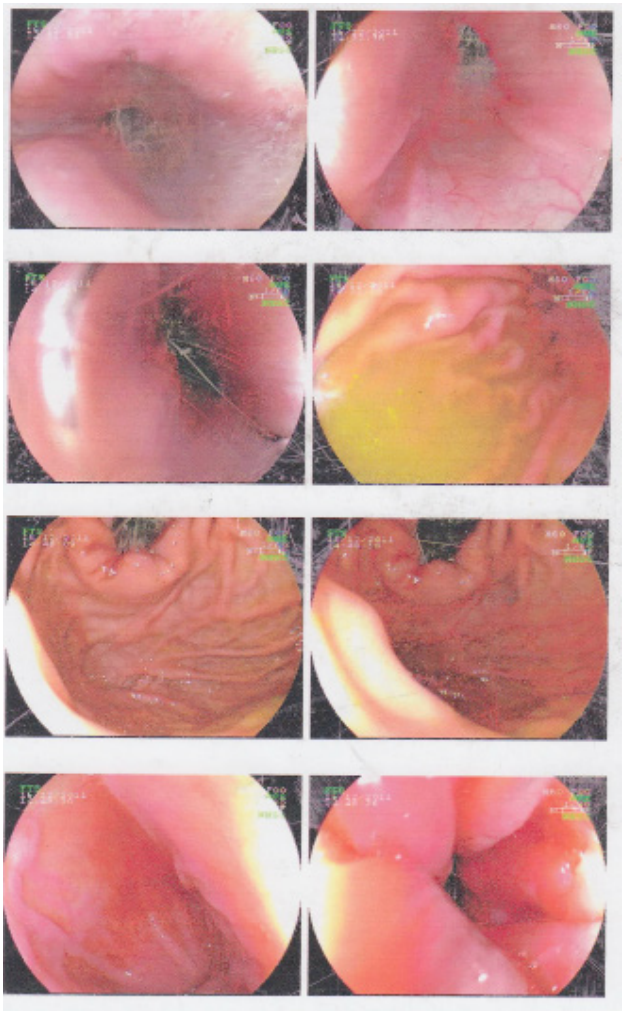

Figure 2. Esophageal appearance before the first dilatation showed narrowing of lower esophageal sphincter with retention of some food

The patient had the second dilatation in the following week after the first one. The second dilatation was also a success. She was still being hospitalized for a couple of days in order to observe if there was any complication of dilatation and to evaluate the response of dilatation. She could eat properly thereafter without experiencing any difficulties while eating or drinking, such as chest discomfort, pain or vomit. There were no more episodes of running nose or like burn sensation inside the chest.

After several days of observation the patient was discharged and being told to have a routine check up to other gastroenterologist in her home town, at least one month after the procedure or sooner than that if she felt any more symptoms like she had experienced before. On the follow up, the patient still sometimes experiences chest discomfort while eating only solid food for the last 2 months. She can still eat like usual; there is no vomiting or weight loss.

DISCUSSION

There are several terms of achalasia: primary achalasia, secondary achalasia and pseudo-achalasia. Primary achalasia is a primary esophageal motor disorder of unknown etiology characterized manometrically by insufficient lower esophageal sphincter (LES) relaxation and loss of esophageal peristalsis, esophageal dilation, minimal LES opening with a "bird-beak" appearance, and poor esophageal emptying of barium.[1] Secondary achalasia shares clinical features with primary achalasia, but there is always an identifiable cause. Worldwide, the most common cause of secondary achalasia is protozoal infection by *Trypanosomacruzi*, found in Central and South America.[1] When achalasia is caused by infiltration of the lower esophageal sphincter, by malignancy, or by diseases such as amyloidosis, or as a result of a paraneoplastic syndrome, the term pseudoachalasia is used.[1,2] This case will mainly focus in discussing primary type achalasia.

Distal esophageal wall and LES are innervated by postganglionic neurons, consisting of excitatory and inhibitory neurons. The excitatory neurons release acetylcholine while the inhibitory neurons release nitric oxide (NO) and vasoactive intestinal polypeptide (VIP), resulting in esophageal and LES contractions and relaxations, respectively.

Achalasia results from the degeneration of neurons in esophageal wall. The NO and VIP releasing inhibitory neurons are the target cellsin idiopathic achalasia.[6] Histologic examination shows the decrease amount of neurons (ganglion cell) in myenteric plexuses. The ganglion cells are surrounded by lymphocytes and less prominently by eosinophils. However this inflammatory degeneration relatively spares the cholinergic neurons (causing contraction of smooth muscle in LES) and preferentially affects nitric oxide producing inhibitory neurons (causing relaxation of smooth muscle).[6,7] There is also one hypothesis about the role of interstitial cells of Cajal (ICC) that has been identified in LES (serving as pacemakers, as generators of a smooth muscle hyperpolarizing factor, asmechanicsensors, and as mediators of neurotransmission). In achalasia, there is also a reduction in number of ICC in LES.[8]

The hallmark of clinical presentation in achalasia is dysphagia for solid and liquids.[1,2,3,4] Most achalasia patients are asymptomatic for years before seeking medical attention. The other symptoms are: 1) regurgitation, mostly in recumbent position, the patient awake from coughing and choking; 2) chest pain or discomfort located in the xiphoid area that may mimic angina by location and character, but not being aggravated by exercise or relieved by rest; 3) the sensation of heartburn because of gastroesophageal reflux disease (GERD) or the result of production of lactic acid from retained food or exogenous ingested acidic materials such as carbonated drinks; 4) weight loss, the degree is usually mild (5 to 10 kg), but profound weight loss can be found; 5) various maneuvers, including lifting the neck, throwing the shoulder back, or drinking carbonated beverages to help empty the esophagus; 6) globus sensation, refer to lump-like sensation in the throat; 7) hiccups as a result from obstruction in distal esophagus.

In regard to patient's history of present illness, we could find many symptoms that could lead us to achalasia for our working diagnosis. First of all, the patient chief complaint was having difficulties in swallowing either food or liquid since three years before admission. It means that there was an abnormality in her digestive system, particularly in her esophagus. We have to suspect every patient that comes with dysphagia symptoms to have achalasia as their underlying disease. The other symptoms that can lead us to diagnosis of achalasia in this patient were regurgitation that she experienced while she was lying on her back, resulting in coughs. Then she also experienced chest discomfort, more like dull pain inside her chest whenever she ate. The sensation like something stuck inside her chest that patient had been complaining referred to globus sensation. There was also heartburn sensation in this patient that could be the result of GERD or the lactic acid produced by theretained food in her esophagus. She also had a maneuver that helped her swallowing food, which was drinking water immediately after swallowing some food, in order to flush them down passing through the LES. Weight loss was also an issue in this patientabout 40 kg in 2 years. All those findings led us to achalasia, the next step was to establish thediagnosis and to classify which type of achalasia the patient had.

The diagnosis of achalasia is based on the results of gastroscopy, manometry, and timed barium esophagography. When the diagnosis of achalasia is suspected, a barium esophagram with fluoroscopy is the single best diagnostic study. This test will reveal loss of primary peristalsis in the distal two-thirds of the esophagus with to-and-fro movement in the supine position. In the upright position, there will be poor emptying with retained food and saliva often producing a heterogeneous air-fluid level at the top of the barium column. Early in the disease, the esophagus may be minimally dilated, but more chronic disease is associated with sigmoid-like tortuosity and sometime massive dilation of the esophageal body. There is a smooth tapering of the lower esophagus leading to the closed LES, resembling a "bird's beak". The films are taken at 1, 2 and 5 minutes after the last swallow of barium; the purpose of 2 min film is to assess interim emptying.[3,6,8] When findings of achalasia are present on barium studies, a narrowed distal esophageal segment longer than 3.5 cm with little or no proximal dilatation in a patient with recent onset of dysphagia should be considered highly suggestive of secondary achalasia, even in the absence of other suspicious radiographic findings.[9]

Manometry is the gold standard means for establishing the diagnosis of achalasia. We require esophageal aperistalsis to be present in order to diagnose achalasia. The LES displays high pressure at rest and fails to relax, or relaxes only partially with swallowing. Up to 40% of patients with achalasia have normal LES pressure (10–40 mm Hg).[6] However, low pressure LES is not seen in untreated achalasia patients. Since the emergence of high resolution manometry (HRM) with pressure topography plotting, esophageal achalasia can be classified into three subtypes.[3,10] In type I achalasia (classic achalasia), impaired LES relaxation but no significant pressurization within the esophageal body is observed. In type II achalasia (with compression), swallowing of water causes rapid panesophageal pressurization. This may exceed LES pressure, causing the esophagus to empty. Type III achalasia (spastic achalasia) is also associated with rapidly propagated pressurization; however, the pressurization is attributable to an abnormal lumen, obliterating contraction.[3]

All patients with suspected achalasia should undergo upper gastrointestinal endoscopy to exclude pseudoachalasia. At endoscopy, the esophageal body may look normal, or dilated, atonic and often tortuous. The mucosa looks normal, but sometimes it is thickened or friable with even superficial ulcers secondary to chronic stasis or candida esophagitis. The LES is closed even with insufflations of air, but the endoscope can easily pass this area with gentle pressure.[3,4,6] If excess pressure is required, the presence of pseudoachalasia should be highly suspected, the gastroesophageal junction and cardia must be closely examined, and biopsies should be taken. Endoscopic ultrasonography may prove to be useful in patients with a non diagnostic endoscopy and a high degree of clinical suspicion for pseudoachalasia, but it is not recommended as a routine test.[11]

The suspicion of achalasia in this patient was established from the result of barium esophagram and endoscopic procedure. Although the main key test for establishing achalasia, manometry, had not been done, still we could be sure of that the main underlying disease for patient dysphagia was achalasia. It is said that barium esophagram is the single best diagnostic study because it can provide us with the specific appearance, especially if the disease is not in its early state. From barium esophagram we could find the "bird's beak" appearance, which meant that there was a smooth tapering of the lower esophagus leading to the closed LES. The endoscopic procedure in this patient was compulsory. Its purpose was not to reconfirm the diagnosis of achalasia that we had found from barium esophagram, but to distinguish between achalasia and pseudoachalasia. As many literature have stated, that all patients with suspected achalasia should undergo upper gastrointestinal endoscopy to exclude pseudoachalasia.[1,6,11] From the first gastroscopy procedure in Abdoel Moeloek Hospital, we knew that there was a narrowing of LES but with the appliance of gentle pressure the scope can still pass through it. The diagnostic of achalasia is less likely to result from gastroscopy so there was no indication to do any biopsy or to have another examination such as CT scan or endoscopic ultrasonography, therefore the diagnosis of pseudoachalasia could be excluded. From gastroscopic findings, we could also know that there was no other condition that could worsen the condition of the patient such as ulcers or candida esophagitis secondary to chronic stasis.[6]

Currently, there was no cure for esophageal achalasia. The only available therapeutic options are to loosen the LES (focuses on relaxation or mechanical disruption of LES) and treat the symptoms. If achalasia is left untreated, a dilated esophagus with severe bolus transit impairment will develop, and that condition signals high risk for aspiration pneumonia

or perforation. Therefore, the goal in the management of achalasia is an early diagnosis and treatment before reaching this end-stage phase, when surgery becomes mandatory, but usually with a poor outcome.[3,8]

Pharmacologic agents used for achalasia are mostly smooth muscle relaxants that act by reducing LES pressure. Calcium channel blockers and long-acting nitrates are the two most common medications used.[1,5,8,12] Pharmacologic agents rarely yield satisfactory long-term alleviation of symptoms and are now only used in patients who are not candidates for pneumatic dilation or surgery. They also can be used during the planning stage for more effective therapy.[6,8] Calcium channel blockers inhibit cellular uptake of calcium, and because intracellular calcium is necessary for lower esophageal sphincter contraction, relaxation occurs. Calcium channel blockers have been evaluated in the long-term treatment of achalasia, but the use of these drugs often induces tolerance, which severely diminishes the effects over the time. Studies show that the time to maximum effect for sublingual nifedipine is 20 to 45 minutes; therefore, it is recommended nifedipine (10–30 mg) to be given sublingually 30 to 45 minutes before meals and at bedtime. Nitrate therapy acts by counteracting the decrease in the inhibitory neurotransmitter, nitric oxide. This leads to a decrease in lower esophageal sphincter pressure. The effect of nitrate is more rapid than that of nifidipine, but has a shorter duration; thus, sublingually isosorbide dinitrate (ISDN) 2.5-5 mg or orally 10-20 mg is commonly administered only 10 to 15 minutes before meals.[6,8,12]

The patient was scheduled to undergo a PD in Cipto Mangunkusumo Hospital, while she was waiting for it, she still suffered from the dysphagia and chest discomfort every meal time. There was absolutely an indication to give pharmacologic treatment for this patient, first that she was still having symptoms and second, that it was only for short period of time, while she was waiting for more definite treatment, so that we could avoid any unpleasant side effect such as headache, dizziness, and pedal edema.[3] For that reason we chose to use nifedipine sublingually 10 mg before each meal. We also gave proton pump inhibitor, omeprazole 20 mg to be taken twice daily and also sucralfate as mucous protector to be taken four times daily to help alleviate the heartburn sensation.

Another common therapy for achalasia is endoscopic treatment; there were two available options currently, which were botulinum toxin (BT) injection or PD. Endoscopic treatment with BT injection at the terminal nerve endings of myoneural junctions prevents the release of acetylcholine from vesicles. This causes chemical denervation, which may last for several months. As a result of its wider safety range and fewer complications, local injection of BT into the LES muscle of patients with achalasia lowers LES tone, and the patient becomes asymptomatic. This treatment yields excellent immediate responses with success rates of > 90%. However, the results last only 6-9 motnhs on average in most patients, and only half of all the patients benefit for > 1 year.[1,3,8] Botulinum toxin A 80–100 U is injected through a 5 mm needle into the LES, with aliquots of 20–25 U of the toxin injected into the four quadrants of the LES. The patients can go home after they recover from sedation and are allowed to eat soft foods later in the day. Symptomatic improvement occurs gradually and usually peaks 1–3 days later, although this may be delayed even further in the occasional patient.[6,8] Surgeons have reported that Heller myotomy could be more difficult in patients who undergo repeated botulinum toxin A injection due to increased adhesion of the muscular layer.[13] Overall, BT is recommended to be most effective in elderly patients, in whom dilation or surgery represent a high risk, or in patients with comorbid illnesses who are not candidates for PD or myotomy.[1,3,8,13]

PD is the most effective non-surgical treatment option for patients with achalasia. It uses the air to dilate the esophageal lumen and disrupt the circular muscle fibers of the LES. Typically, pneumatic dilation is performed over a guide wire under fluoroscopic guidance. This allows for assessment of the progress of dilation as the narrowed "waist" of the lower esophageal sphincter disappears. However, in addition to radiation exposure, this requires bulky and expensive fluoroscopy equipment, which may be in high demand or not available for general use. Performing PD under direct endoscopic visualization is a good alternative to fluoroscopic guidance. After placement of the dilator over the guide wire, the endoscope is reinserted into the esophagus. The balloon placement and dilation effects are observed proximal to the balloon. The number of dilation sessions and the inflation time needed for a successful dilation vary and are operator dependent.[3,8] In a prospective follow-up investigation study conducted by Eckardt et al, only 40% of the patients treated using a single PD procedure remained in remission at 5 years.[14]

This patient underwent PD for a definite treatment. This option was chosen because itwas the most effective therapy compare to the other therapies. It also uncommon to have any complication, the most

severe complication is perforation, and it only occurs in approximately 2% of cases.[3]

The first attempt in this patient failed because there was so many food leftover inside the esophagus even though the patient had fasted 12 hour before the procedure. So the nasal gastric tube (NGT) was placed then, in the middle of patient's esophagus, proximal from the LES, and esophageal lavage was done every 2 hours until it was clear enough. The second attempt was a success, there was no more food leftover, the guide wire then being inserted to help the placement of the dilator, this procedure was done under fluoroscopy, to evaluate the precision of the balloon placement. After the balloon was put in the correct position, gastroesophageal junction fluoroscopically, then the balloon was dilated to obliterate the waist. From endoscopic evaluation after the dilatation we can see that the waist of achalasia was disappeared and there is no sign of any bleeding at that time. The patient then was observed for another couple of days to evaluate if there was any chest pain, bleeding, or difficulty in swallowing before being discharged home. The evaluation after the PD should include a barium swallow to see if there was any esophageal perforation, which was not being done in this patient. Timed barium esophagogram can be used as a better predictor of treatment success after PD. We have found that in almost 70% of the patients, the height of the barium column at 5 minute post-therapy correlates with symptom improvement (concordant group), while in others esophageal emptying was poor despite reports of excellent symptom relief (discordant group). Nearly all patients in discordant group failed the treatment within 1 year after treatment, while 77% of the concordant groups were still in symptom remission after 6 years of follow-up. Therefore, it is suggested that the timed barium esophagogram not only assesses treatment shortly after therapy, it can also predict the poor response to the treatment if the patient has retained barium postpneumatic dilation.[15]

From the follow up one month after, the patient had no difficulty in swallowing or had any chest discomfort like she had used to have before. Although from the follow up 5 months after, the patient began to feel slight chest discomfort again after eating, but this time there was no sign of regurgitation, heartburn sensation, or any extra effort to help the patient in swallowing food. It is generally accepted that the predictors of risk factors for relapse after PD include young age (< 40 - 45 years), male, single dilation with a 3.0 cm balloon, post-treatment LES pressure > 10-15 mmHg, poor esophageal emptying after timed barium swallow, and type I and type III achalasia pattern on HRM.[3] In this patient the risk factors are young age (20 years old) and single dilation.

The patient may need for another dilation session using a wider diameter of the balloon (3.5 cm and 4 cm). If there wasstill any symptom then the patient should undergo a laparoscopic myotomy (Heller myotomy). This technique is associated with the greater decrease in dysphagia, a shorter hospital stay, lower risk of postoperative gastroesophageal reflux, and low complication rates. An entire flux procedure can further reduce postoperative heartburn rates by 80%, as well as the risk of esophagitis and peptic stricture.[8,13] If there is still no improvement and the patient is already in the late stage of the disease, with mega esophagus or there is any complication from the latter myotomy such as adenocarcinoma in Barrett's esophagus then drastic measure need to be done, which is esophagectomy.

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Correspondence:
Andree Kurniawan
Department of Internal Medicine
Siloam General Hospital
Jl. Boulevard Jendral Sudirman Tangerang 15811 Indonesia
Phone: +62-21-54210130 Ext: 3411 Facsimile: +62-21- 54210133
E-mail: andree.kurniawan@uph.edu