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(CASE REPORT)

Acute respiratory failure in achalasia with megaesophagus

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Abstract

Introduction: Achalasia is one of the most common causes of dysphagia. Typical symptoms include difficulty in swallowing, regurgitation, weight loss, and chest pain. Megaesophagus is a complication of advanced achalasia. This condition rarely causes tracheal compression with acute dyspnea.

Case Presentation : A 51 years old man came to the emergency room and had complaints of intermittent shortness of breath accompanied by a cough for 1 week, chest pain that came and went, nausea (+), vomiting (+), difficulty in swallowing food, and fever (-). The medical history showed that the patient went to gastroenterology polyclinic to examine his dysphagia and esophageal sphincter dilatation 2 months ago. The patient received treatment in the ward, but the complaints he suffered was getting worse, so he was transferred to the ICU with a diagnosis of acute respiratory failure.

Conclusion: The patient was then intubated and decompressed for the megaoesophagus he suffered with the aim to reduce the compression of the trachea and lungs by the esophagus. The patient was getting better and could be extubated on the 3rd day of treatment.

Keywords: Achalasia; Megaesophagus; Dyspnea; Dysphagia

1. Introduction

Achalasia is the main cause of dysphagia. It is reported that the incidence is found in 1 per 100,000 individuals per year. Both men and women have the same possibility to suffer from achalasia as the increase in their age. Achalasia is divided into primary and secondary achalasia. In primary achalasia, the exact etiology is not yet known, possibly caused by a neutropic viral infection resulting in a lesion of the dorsal vagal nucleus in the brainstem and a mesenteric ganglion in the esophagus. Heredity also has a role in this disorder. While the secondary achalasia is caused by infections, intraluminar tumors such as cardidia tumors, extraluminal pressure from pancreatic pseudocysts, anticholinergic drugs, or postoperative vagotomy(1).

The esophagus has the main function of delivering food swallowed from the pharynx to the stomach using peristalsis. The top and bottom of the esophagus have a sphincter that is normally in a tonic or contracting condition to prevent acid reflux of the stomach to the esophagus (1).

Under normal conditions, the esophagus has two types of peristalsis, namely primary and secondary peristalsis. Primary peristalsis is a continuation of peristalsis in the pharynx which is able to deliver food to the stomach in about 8-10 seconds and it can be faster if in a standing position due to gravitational force. If the primary peristaltic fails to transfer food to the stomach, secondary peristalsis appears.

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Histological analysis reveals that achalasia is the result of degeneration of ganglion cells in the myenteric plexus of the esophageal corpus and the lower esophageal sphincter (LES), which causes the esophageal body to remain immobile and LES cannot relax.

Today, the treatment options are orienting to symptoms and reducing the pressure from unsustainable LES. Further genetic research may improve our understanding of the etiology and pathogenesis of achalasia and may lead to better treatment options in our toolbox in the future (2).

A recently published genetic association study showed that immune processes are involved in the pathophysiology of achalasia. Treatments currently can be provided are Laparoscopy of Heller myotomy (LHM) with partial fundoplication, per oral endoscopy myotomy (POEM), pneumatic dilation (PD), and injection of endoscopic botulinum toxin (EBTI), each of which has its own advantages and disadvantages. While typical symptoms, such as dysphagia, regurgitation, weight loss, and chest pain are the most common complaints obtained at the beginning of diagnosis. The case report describes a rare case of adult patients suffering from megaoesophagus because of clinically manifested achalasia of acute respiratory failure.

2. Case report

Mr. A, 51 years old came to the emergency room (ER) with intermittent shortness of breath accompanied by a cough for 1 week, intermittent chest pain, nausea, vomiting, difficulty in swallowing food. The medical history showed that the patient went to gastroenterology polyclinic to examine his dysphagia and esophageal sphincter dilatation 2 months ago. The patient was in conscious condition, but with shortness of breath, blood pressure of 103/69 mmHg, pulse of 126x/min, breath of 26x/min, temperature of 36.7 ^{0}C, and SpO2 of 84%. In the ER, he received O2 NRM therapy of 15L/min, RL infusion of 20 tpm, Ceftriaxon injection of 2x1 gr, Omz of 1x40mg, Ondansenntron of 3x4 mg, MP of 2x 62.5 mg, and oral tablets of N-acetylsitein.

The patient was treated by the doctor who was on duty in the ward for 1 day but the complaint of shortness of breath was getting worse and then he was transferred to the ICU. The patient arrived in the ICU in a conscious condition with blood pressure of 130/93 mmHg, pulse of 137x/minute, breath of 39x/min, temperature of 36.7^{0}C, SpO2 of 84% then he was intubated. After intubation, patient saturation only increased to 86% with ventilator setting: Volume Control mode, Tidal Volume of 380 cc, PEEP 5, Respiration rate of 16, FIO2 of 100%. On auscultation of the patient's lungs, rhonchi were found in both lung fields. After that, suction was performed through the endotracheal tube and the fluid was successfully cleared, yet the saturation did not increase. The nasogastric tube with a depth of 55cm was placed, syringe test was performed and there was not “whoosh sound” in patient’s stomach. So, we estimated that the nasogastric tube could not reach the stomach and could only enter the megaesophagus. Decompression was performed via nasogastric tube and 1000cc of residual fluid was obtained. After the decompression, the patient's saturation increased to 100%.

After the patient is admitted to the ICU for 3 days on a ventilator, the patient can be weaning off and extubated. The patient was in the ICU for 7 days. On the the 5th day of treatment, a laparatomic operation *of Heller myotomy* was performed.

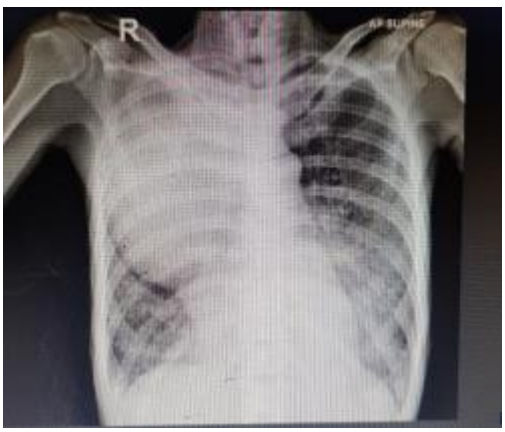

Figure 1 Thorax X-ray without contrast at Emergency Room

- Reading
  - Heart : CTR < 56%
  - Lobulated opacity in the right paratracheal area is seen as high as VT 2-10
  - Pulmo: Increased bronchovascular markings, visible patches in the middle and lower pulmonary fields bilaterally
- Impression:
  - Suspected mediastinal mass
  - Bronchopneumonia
  - Right pleural effusion
  - normal heart size

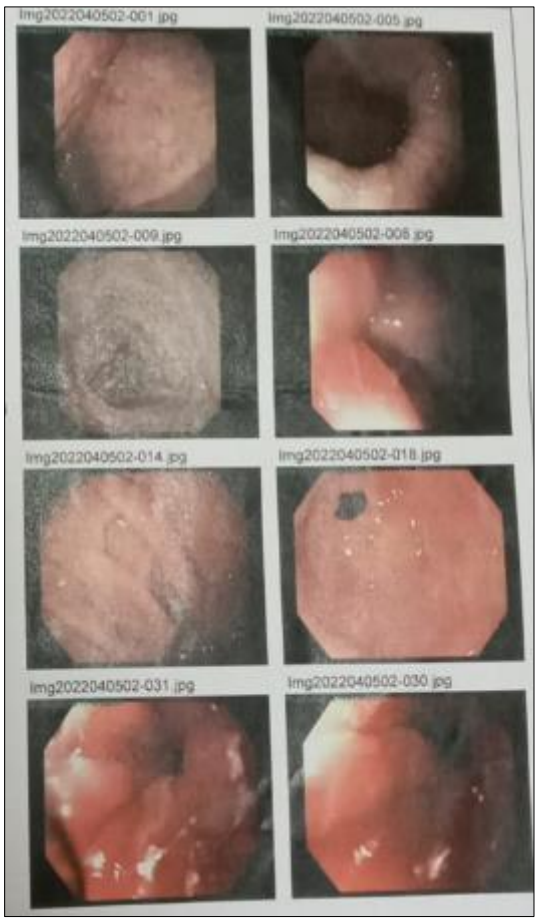

Figure 2 Endoscopic results of Mr. A

Esophagus: The diameter of the esophagus is very wide and there is a structure of the esophagogastric junction with strong resistance. However, it needs force to make the gastroscope enter the stomach, the esophagus is dilated with dilatation traction maneuver 3 times and there are several mucosal breaks on the esophagogastric junction and the gastroscope can enter the stomach. without a force.

Gastric: Normal mucosa

Duodenum: The scope cannot enter the duodenum because the length of the scope has reached its maximum to the gastric antrum

Endoscopic diagnosis: Achalasia with megaesophagus is successfully dilated

3. Radiology examination 4 month before respiratory distress

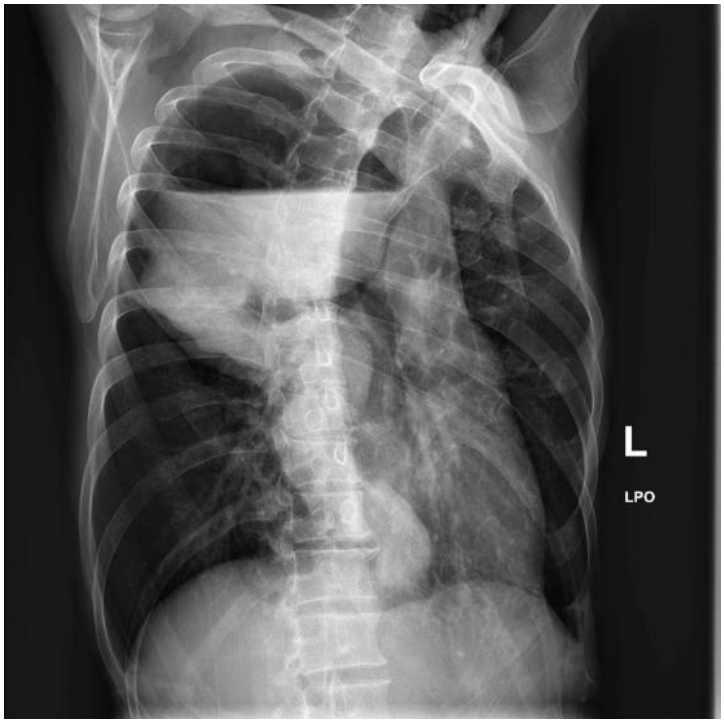

Figure 3 Chest X-ray with contrast, Left Posterior Oblique (LPO) Position

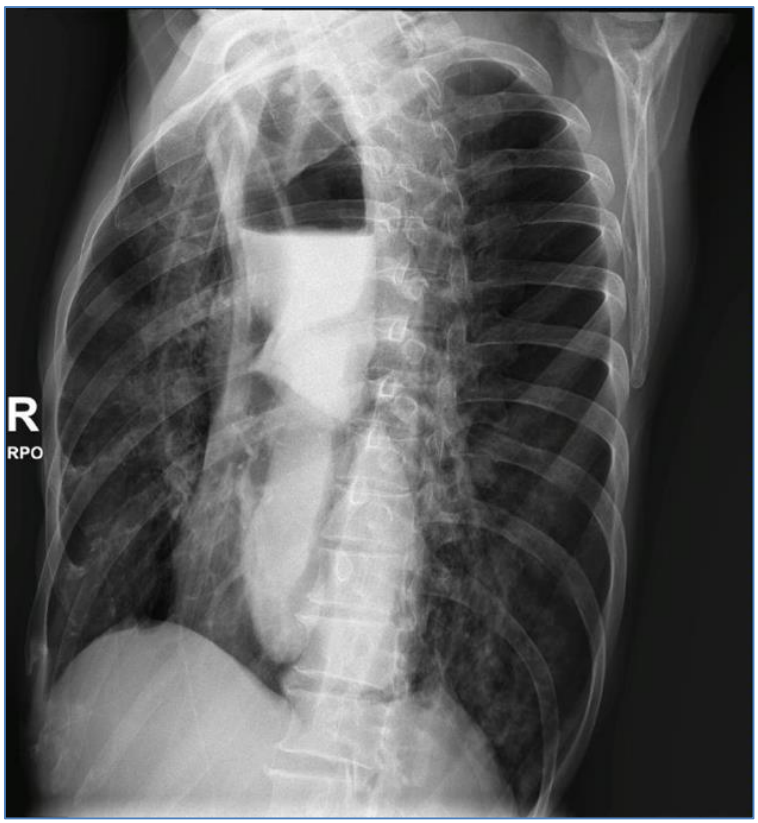

Figure 4 Chest X-ray with contrast, Right Posterior Oblique (RPO) Position

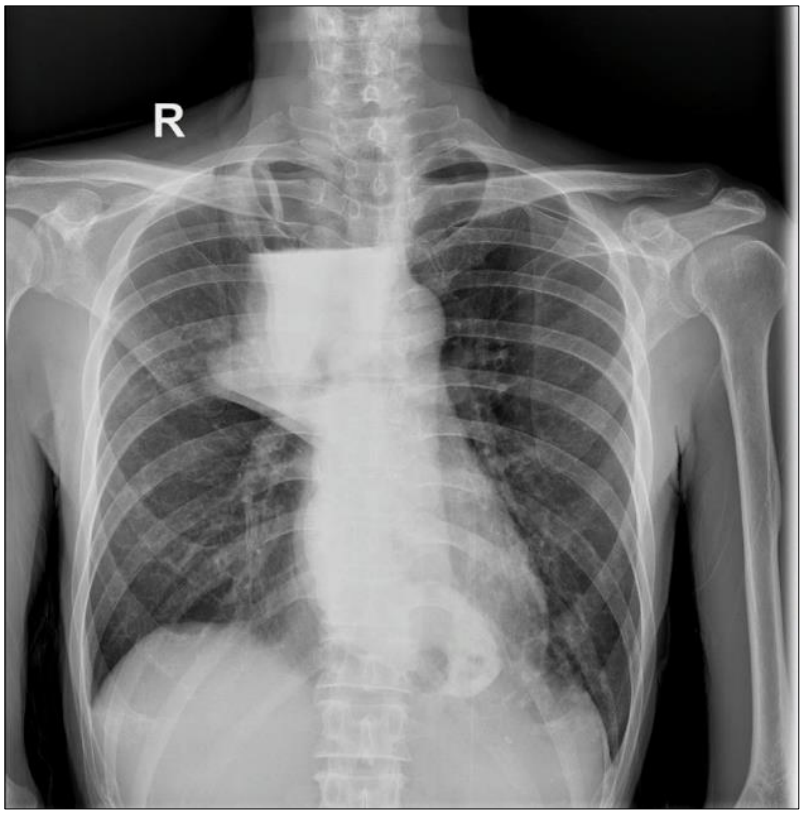

Figure 5 Chest X-ray with contrast, AP Position

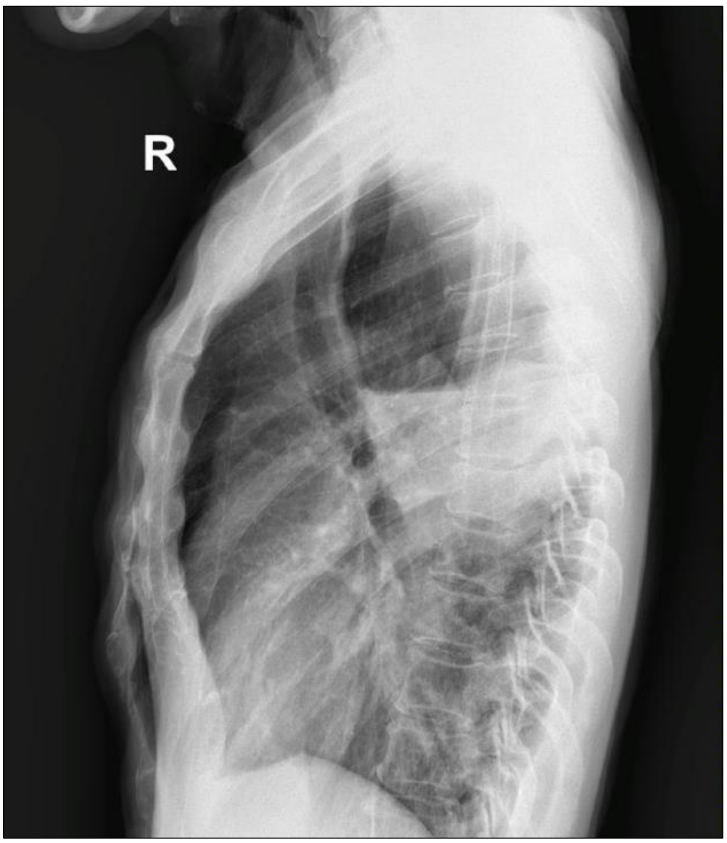

Figure 6 Chest X-ray with contrast, Lateral position

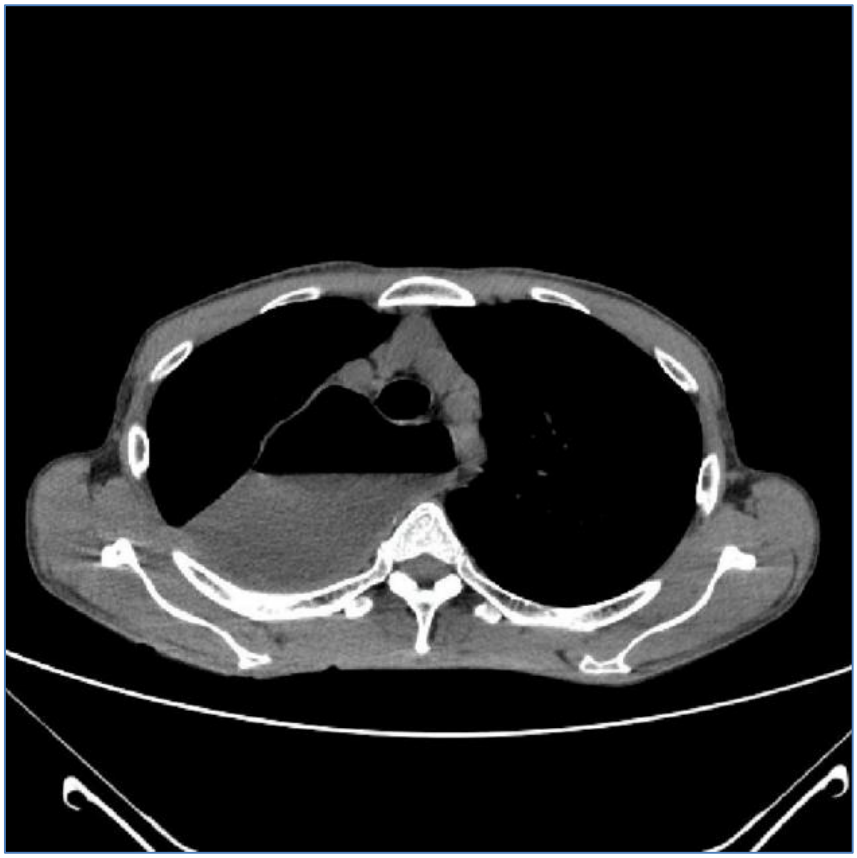

Figure 7 MSCT thorax with contrast

Table 1 Hemodynamics

19/09/22	**10:00**	**11:00**	**12:00**	**24:00**	**20/06/22**	**6:00**	**12:00**	**18:00**	**24:00**
BP SIST	130	115	114	94	BP SIST	98	95	104	100
BP DIAS	93	90	82	69	BP DIAS	68	74	67	68
MAP	105	99	93	78	MAP	79	81	79	79
PULSE	137	139	139	113	PULSE	100	94	98	102
RR	39	24	34	15	RR	14	15	16	13
SPO2	84	86	99	99	SPO2	100	100	100	99
TEMP	36.8	37	37	36.2	TEMP	37	36.5	37	37
	NRM 10L	A/C		SIMV		SIMV		SIMV	
		TV 350		PS 10		PS 10		PS 10	
		PEEP 5		PEEP 5		PEEP 5		PEEP 5	
		RR 16		RR 12		RR 12		RR 12	
		FIO2		FIO2		FIO2		FIO2	
		100		60%		60%		50%	

Table 2 Labs Results

	18/6/22	**20/6/22**	**23/6/22**	**24/6/22**	**BGA**	**19/6/22**	**20/6/22**
HB	12		8.3	11.3	PH	7.17	7.21
HCT	40		28	36	PO2	58.8	105.2
LEU	20980		25760	18300	PCO2	55	60.3
TRO	776000		233000	211000	HCO3	19.8	23.7
UR	57.31		39.64		BE	-9.1	-5.1
CR	0.64		0.39		SPO2	84.2	96.8
GDS	85		185		AaDO2	452.8	
SGOT	11						
SGPT	5						
ALB	3.5”	2.98	2.91				
Na	145		136				
K	4.7		4.6				
Cl	106		102				

4. Discussion

The vast majority (90%) of patients suffering from achalasia have complaints of dysphagia for solid and liquid foods as its main symptom. Weight loss, regurgitation, chest pain, and heartburn were reported in 40-60% of patients. Sometimes, respiratory symptoms, such as aspiration leading to pneumonia and bronchiectasis, are associated with this disease. Few publications have reported dyspnea and stridor as well as neck swelling, also referred to as the "*bull frog neck*", are related to advanced fascination. However, there are also cases of acute total airway compression and death mentioned in the literature. In these cases, a low body mass index accompanied by respiratory disorders indicates the severity of the achalasia accompanied by megaesophagus. This patient also wants to vomit when lying down. This can be caused by the regurgitation of fluid stored in the megaesophagus, thereby increasing the risk of aspiration. This condition is getting worse when receiving endotracheal intubation and it is reported that there is a buildup of fluid in the oral cavity. When suction is performed intra ET, quite a lot of aspirate is obtained. In addition, megaesophageal decompression is also carried out to reduce compression of the trachea and lungs. In the presence of acute airway obstruction in suspected or undiagnosed achalasia, early endotracheal intubation with "rapid sequence induction" (RSI; aspiration prophylaxis), emergency esophageal decompression, or tracheostomy with local anesthesia is recommended (3,4).

Megaesophagus is a form of esophageal compensation through dilatation caused by the narrowing of the distal part of the esophagus. Therefore, food and drink consumed will be accumulated in the megaesophagus and they cannot be delivered to the stomach. The causal relationship of massive esophageal dilatation between achalasia and airway obstruction has not been fully explained. The "*pinch-valve*" theory assumes that the dilated esophagus can move behind the crease of the cryptopharyngeal muscle itself. This can lead to one-way valves that trap air inside the esophagus. The second theory mentions the upper esophageal sphincter cannot relax during the swallowing process. Furthermore, the loss of the saltpeter reflex physiologically leads to the simultaneous relaxation of the SLE and the simultaneous relaxation of the upper esophageal sphincter (UES). Only a few cases of tracheal compression caused by the extension of esophageal dilatation on achalasia are reported in the literature because it is a rare disease. These radiological signs and clinical symptoms in patients can be varied and often nonspecific (regardless of the stage of achalasia) and thus they can appear even in the early stages of the disease (5).

5. Conclusion

Acute respiratory failure is rarely observed in patients with achalasia. Early diagnosis and proper treatment of achalasia can restore esophageal and tracheobronchial function, as well as can prevent complications. If the disease is not treated properly, it can be worse rapidly leading to respiratory symptoms such as stridor and acute respiratory failure.

Acute upper airway obstruction due to tracheal and pulmonary compression by esophageal dilatation may occur in cases of akalsasia due to UES dysfunction. Endotracheal intubation and installation of nasogastric tubes into the esophagus for the decompression of megaesophagus can reduce respiratory symptoms.

Compliance with ethical standards

Disclosure of conflict of interest

The authors have no conflicts of interest to declare that are relevant to the content of this article.

Statement of ethical approval

his study was approved by Dr. Margono Soekarjo Hospital ethics and research committee.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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