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Repot a Rare Case of Achalasia Which Present as Mediastinal Cysts or Masses

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1. Abstract

Achalasia is a rare motility disorder of the esophagus and present by failure of lower esophageal sphincter (LES) relaxation during swallow. This rare disease of esophagus with it symptoms can mimic a variety of common illnesses of esophagouse,but not the mediastinum

A man with 39 years old was referred to our chest clinic with complaints of dysphagia to solid foods and liquids food, chest pain and cough after eating, Dyspnea and regurgitation present latter at night with cough. These symptoms present for the past 2 years and in resent three months becoming progressively worse. A CXR and computer tomography (CT)of chest were performed which show right lung cystic or mass lesion with necrosis. After visit by a pulmonologist and a thoracic surgeon, the patient was transferred to the thoracic Surgery department for a planned surgical intervention. After postero-lateral thoracotomy and exploration, the diagnosis was achalasia whit huge megaesophaguse. Esophagectomy and gastric pull-up was performed. The patient died three day post operation because of pulmonary sepsis due to aspiration

2. Introduction

In vast majority of patients, the etiology of achalasia remains unknown [1, 2]. Achalasia is an unusual motility disorder of esophagus which involve the smooth muscle of the esophagus wall and (LES). The incidence of achalasia about 1 in 100,000 of people and the age of most patients on the time of diagnosis are between 25 to 60 years old [1-3]. In more than 80% of patients, major symptoms of achalasia are usually dysphagia to solid and liquid food, regurgitation, weight loss and chest pain [1,4,5]. The cause of These symptoms are the absence or weak of peristalsis in esophageal body, failure relaxation of LES and pressure in the (LES) is higher than normal, on swallowing time [5, 6] One of primary diagnostic methods are CXR and barium swallow in liquid form and then the movement of barium may follow by fluoroscopy to LES [1, 2]. Other methods of diagnosis are an endoscopy end esophageal ultrasonophrapy and manometry [6, 3].

The radiographic investigations were performed with the patient in a supine semi upright or upright position [3, 6]. Measurements of the maximum diameter of the esophageal body and the gastroesophageal junction at its narrowest point were obtained [1,3,6]. Manometry is the best tool for definitive diagnosis. But todays high-resolution manometry has largely preform than the traditional old manometry. Pressure recording is measure by a catheter in multiple site in the esophagus lumen and traverses the esophagus lumen to the LES and show presentation of esophageal body peristalsis and show the best localization of the LES [5,6]. The defini-

tive diagnosis of achalasia is usually delayed for many years [1,2].

Despit of multiple methods for treatment, but there is not an effective treatment to correct the esophageal body peristalsis and motility [1,4]. All efforts aims are a palliate therapy for dysphagia with disruption of hypertonic (LES) muscle [3,6].

Several surgical and nonsurgical methods have been described for disruption muscle of LES [3,6] But in recent years, minimally invasive techniques as laparoscopy or thorascopy is the surgical treatment of choice [6 ,7]. First, cardiomyotomy performed by Heller through an abdominal approach which in this method both anterior and posterior side of LES muscle was divisions [6,1]. Palliation of dysphagia was the first goal of therapy but the motor dysfunction persists after the cardiomyotomy [8]. A modified Heller's myotomy performed at present time and is the most efficient for long-term solution [6,8]. In some reports recommended often as the first line of treatment [9]. When the esophagus was re-evaluation with barium after more than 10 years, diverticulum formation may present through the myotomized area, and the size of diverticulum depend to the length of the myotomy [8,9]. This complication need for re-operation r (29%) [7,9]. For treatment of this complication, need to take down the Nissen fundoplication or to perform an esophagectomy [7]. The main aim of the esophagogastric myotomy is to reduce the pressure of LES relaxation by division of slingand clasp fibers. One complication of this operation is, pathologic gastero-esophageal reflux and for prevention of reflux a fundoplication procedure remains necessary [10] After the advent of minimally invasive technology, attempts at a pure thoracoscopic approach were problematic and commonly led to incomplete LES division even under endoscopic guidance [2]. More recently, laparoscopic myotomy with partial fundoplication has gained popularity for the treatment of achalasia [3]. Controversy still persists regarding the ideal length of myotomy to treat esophageal achalasia [2,8]. In sigmoid or megaesophagousin form, esophagectomy and gastric pull-up or colon bypass indicated [4,12]. The aim of this case report is to show the rare clinical presentation and imaging finding in achalasia. Because with our knowledge and review of literatures we don't find such case. With this imaging presentations and very huge megaesophagousin.

3. Case Presentation

A man with 39 years old was referred to our chest clinic with complaints of dysphagia to solid foods and liquids food, chest pain and cough after eating, Dyspnea and regurgitation present latter at night with cough. These symptoms present for the past 2 years and in resent three months becoming progressively dysphagia, was overcome by drinking water, a constant weight loss of 23 kg, which had developed over the past two years. Additional symptoms were retrosternal pain, regurgitations, night coughing, dyspnea, right side chest pain, heaviness of chest wall and recurrent respiratory infection. While the laboratory results did show anemia (9mh /l), (albumin 2mg/l) others was normal, a CXR and computed tomography (CT) scan of the chest show a huge cystic lesion with air fluid level in right hemithorax and widening of the mediastinum up to 10 × 8 cm in diameter accompanied by a slit shaped of compress and mild deformity of cranial and narrowing of the trachea (Figure1-6). Differential diagnosis in radiologist report was complicated hydatid cyst, bronchogenic cyst and hiatal hernia, esophageal duplication or mediastinal necrotic masses which compress the esophagus and trachea. Upper endoscopy and bronchoscopy did not performed because the patient cannot tolerated this procedures. Whit above diagnosis, we performed a right postero-lateral thoracotomy whit double lumen for prevention of aspiration during operation, thoracic cavity in 5th intercostal space was opened, during exploration we found a very dilated esophagus which in the proximal portion was very dilated from than other portion of esophageouse. The dilated portion was opened and the content of cavity was full of debris and remnant of swallowing food, after we find two lumen one above and one in the lower portion of this cavity. Very huge megaesophagouse due to achalasia was the definitive diagnosis (Figure 7,8). Esophagectomy was performed, next the position of patient was changed to supine, after laparotomy, for esophagus reconstruction , gastric tube was made, than gastero-esophagostom was performed in the left side of the neck. A feeding joujnostomy tube was performed and abdomen, chest and neck incision was closed. Patient referred to intensive care, three day post-operative the patient was died because of pulmonary sepsis due to aspiration problem.

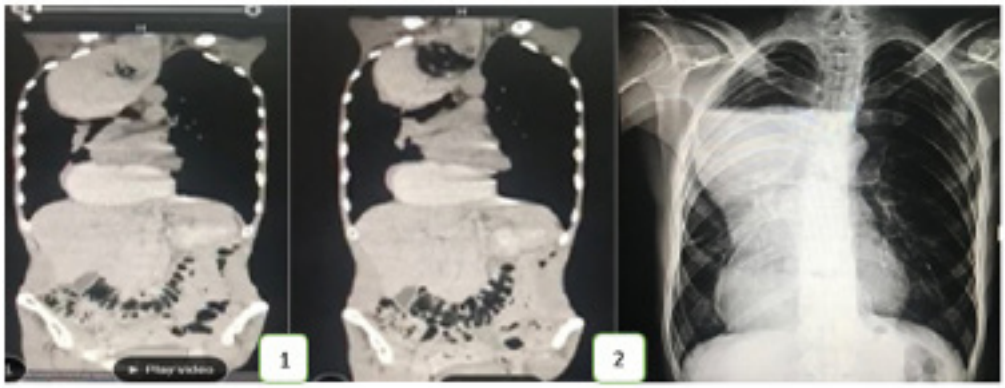

Figure 1, 2: Sagital CT-scan of the chest show mediastinum and cystic lesion with air fluid level

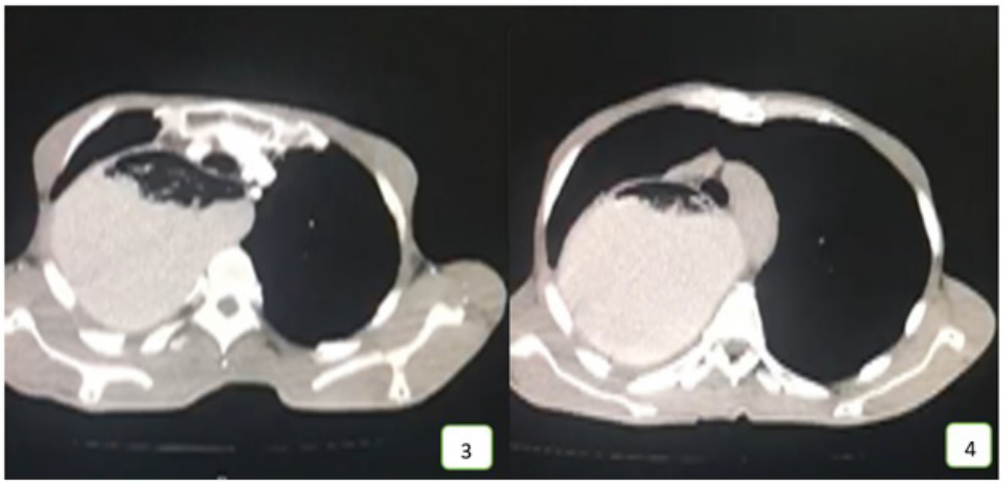

Figure 3, 4: CT-scan of the chest show cystic and solid lesion with air fluid level and compress the trachea

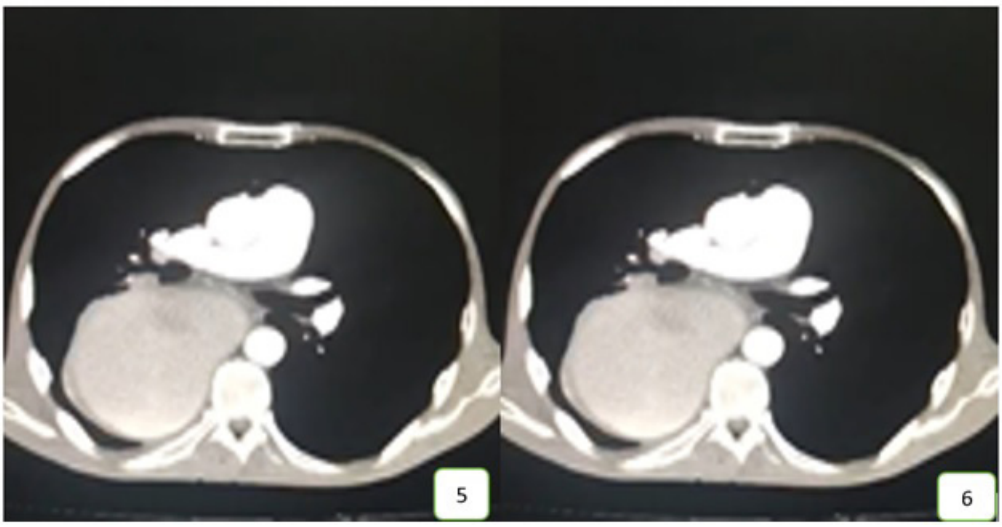

Figure 5,6: CT-scan of the chest show cystic and solid lesion with air fluid level and compress the trachea and carina

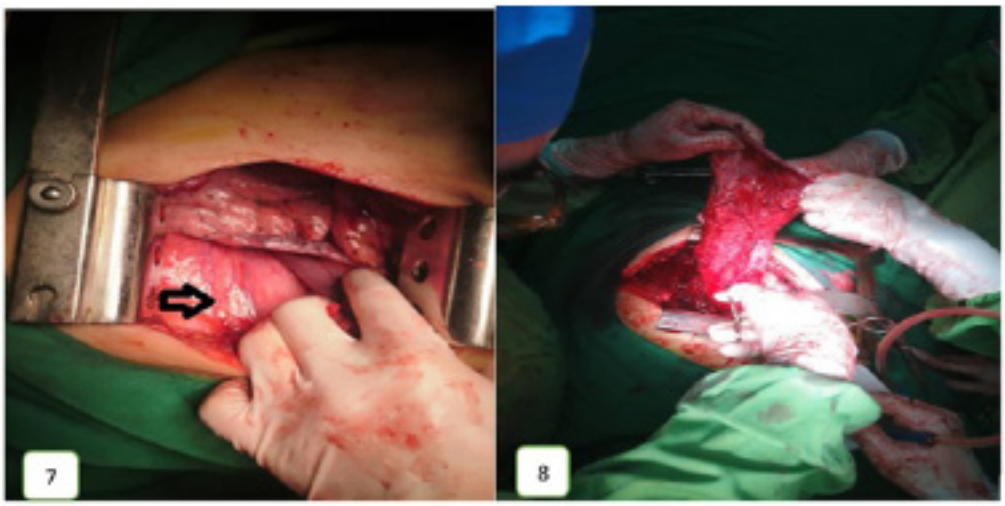

Figure 7,8: intera - operation picture show megaesophagouse and gastric pull-up

4. Dicussuion

The majority (80%) of patients with achalasia complain of dysphagia to solids and liquids food and these symptoms may be the primary compliant in the achalasia patients [1,4]. In 40-60% of patients weight loss, reurgitations, chest pain, cough and heartburn are reported [2,4]. Occasionally, respiratory complications, such as

aspiration pneumonia, lung abcess and bronchiectasis, are another complications of achalasia [3,4]. In advanced stages of achalasia in the few publications report dyspnea, stridor and swelling of the neck due to dilatation of esophagouse [6]. In the literature However, acute total airway compression and death are reported [11].

In this case, the patient present with high weight loss of 21 kg over two years, respiratory problem and hypoalbonomia which show the achalasia is very advanced with delay diagnosis and treatment [4,12]. In this case the CT scan of chest show an extensively widening of mediastinum and a cystic and solid mass with air –fluid level with compression mild of the trachea and carina and bronchial system, respiratory system was significantly compromised. However, in such case difficulties intubation may happen and should be take care of aspiration during induction of anesthesia and before or during and poste surgical intervention, for prevention of sever and fatal aspiration, we insert a tracheal tube in the esophagus and aspiration all content of esophagus In our experience we used this method in four case with successfully.

Diagnosis tools are esophageal manometry, and Chest-X-ray or barium swallow and perform an endoscopy. Another diagnostic method swallow barium should in liquid form. And radiologist will then track the barium's movement down your esophagus through X-rays [1-4]. In our case we used computer- tomography of chest, neck and CXR, but patient can.t tolerate, esophacoscopy, bronscopy and barium swallow. Several types of treatment can either temporarily or permanently reduce symptoms of patients and alter the function of the LES. However most achalasia treatments involve of the LES [7-10]. The main aim of achalasia treatment is to reduce patient's symptoms and to improve their quality of life [1,2,4]. Achalasia is an irreversible disease, and the restoration of esophageal peristalsis is unreliable [4]. Therefore, the ultimate goal of therapy is to relieve the obstruction at the level of the EGJ by either drugs which can produce LES relaxation, botulinum toxin injection, by pneumatic dilatation, per oral endoscopic myotomy, surgical techniques laparoscopic, thorascopy, open thoracotomy or laparatomy Heller myotomy [8-12].

Although the one of fatal complicatin in advanced achalasia during intubation is aspiration. In this patients, if adequate treatment perfome with delay, sever pulmonary complications such as fibrosis of lung occur because of long time of aspiration Tracheal function such as tracheomalacia due to long time of compression might have been possible, compressed of trachea due to mega esophagus might result in life-threatening complication also in earlier courses of postoperative time [3,4,6]. Thus, even more severe problems associated with endotracheal intubation and postoperative extubation, may need for tracheostomy [4,6].

In older patients and possible respiratory or other co-morbidities and, this distinct finding of a slit-shape and For safety reasons, extubation was performed on the intensive care units [8-12]. Therefore, proper therapy may perform in order to restore and prevention of megaesophagus as early as possible [4,6].

Achalasia is a rare esophageal disease such as gastroesophageal reflux and usually diagnosed too late, which often have some difficulty on the time of correct and tratment for many physicians [3,4,6]. However, we recommend that patients with suspicion of achalasia with typical or atypical, especially symptoms should be referred for further diagnostic work-up such as high resolution manometry and barium swallow [4,6,7].

The cause of sever dilated of esophagus (megaesophagous) in achalasia and tracheal obstruction could not be fully discussed yet (4). One reason is esophagus may be displaced posterior to the cricopharyngeal muscle folding [1,4]. Another cause is that the upper esophageal sphincter can not relax during the allowing food [2-4]. Furthermore, in this situation belching reflex not present, because the causes a relaxation of the LES and motive of upper esophageal sphincter (UES) relaxation by belching, (9-700,10-800).

Only very few reported in the literature discuse tracheal compression by megaesophagouse of the esophagus in achalasia and therefore is a rare feature of the disease [6,11,13,14]. The radiologic finding and clinical symptoms in megaesophagouse patients may be multiple and are often unspecific [4]. In the presence of acute airway obstruction in suspected achalasia early endotracheal intubation for prevention of aspiration, early esophageal decompression, or tracheostomy are recommended [9,13]. If surgery is in elective patients, as in our patient, special care should be performed to prevent serious anesthetic complications, especially during intubation [1-4, 6]. Esophagomyotomy is a type of surgery that can help surgeon if the patient have achalasia. The great majority of esophagomyotomy procedures are successful [4]. However, some people have problems after with gastroesophageal reflux disease [4,8-10].

In our case, respiratory symptoms were mild on examination but severely weight loss and decreased breath sound in right side of chest was prominent. Preoperative, achalasia was not our diagnosis with history, physical examination and imaging but intraoperative finding was the cause achalasia. In some patients as our case definitive diagnosis of achalasia was performed on the time of surgery [1-4,6]. Although severe complication during intubation of the patient may were evident [3,4,6]. However, if treatment of patients perform with delay, multiple and serious pulmonary complications would have been happen [4,8,9,10]. Restoration of proper tracheal function in preoperative is important and states such as some complication as tracheomalacia might have been impossible postoperave. Thus, even more with this severe problem associated with endotracheal intubation, postoperative extubation, such as tracheostomy may be need and could have occurred [4,8,9,10].

5. Conclusion

This case report show that achalasia with megaesophagousin, mediastinum widening, cystic lesion and mediastinum mass may be in differentiated diagnosis of megaesophagouse due to achalasia and early treatment need in order to prevent severe and fatal complications, becaused the compression of tracheal due to megaesophagus and sever pulmonary aspiration. If diagnosed by radiologic imaging only, in such patients tracheal obstraction by megaesophagouse or tracheomalacia can difficulty intubation for by anesthesiologists during intubation for surgical interventions and prevention of life-threatening aspiration during surgery, we putt a tube in esophagus and aspiration the content of esophagus.

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