

Megaesophagus as Complication of Achalasia Diagnosed as Incidental Finding in Motorvehicle Accident Patient with Suspected Esophageal Injury

Ikhwan SM, Maznah MZ, Zainira WZW, Zaidi Z, Hassan S

Department of Surgery, School of Medical sciences, Universiti Sains, Malaysia

Publication History

Received: 03 December 2013

Accepted: 06 February 2014

Published: 26 February 2014

Citation

Ikhwan SM, Maznah MZ, Zainira WZW, Zaidi Z, Hassan S. Megaesophagus as Complication of Achalasia Diagnosed as Incidental Finding in Motorvehicle Accident Patient with Suspected Esophageal Injury. *Medical Science*, 2014, 5(18), 72-76

ABSTRACT

Achalasia is a disease of hypertensive lower esophageal sphincter causing incoordination of peristalsis activity within the esophagus. The patient usually presented with typical history of dysphagia, regurgitation and nocturnal cough. Asymptomatic achalasia with complication of megaesophagus is very rare. We presented a case of a 59-year-old man who involved in motorvehicle accident which sustained severe head injury and long bone fracture. He was referred to us in view of suspicious ruptured esophagus after Ryles tube seen coiled within the right hemithorax. Ct scan revealed megaesophagus with area of stricture seen at the level of C5, C6. OGDS confirmed the diagnosis of Achalasia after we ruled out carcinoma with biopsy. As this patient already had severe head injury that requiring him tracheostomy and poor GCS, we proceed with feeding gastrostomy. Unfortunately this patient succumbed to death after almost 2 months in the hospital.

Keywords: megaesophagus, achalasia and feeding gastrostomy.

1. INTRODUCTION

Achalasia is defined as failure of organized peristalsis in oesophagus and failure of relaxation at the level of lower oesophageal sphincter. The actual cause is unknown but it can be categorized as (a) primary (idiopathic) achalasia, due to abnormality of oesophageal aurbachs plexus and (b) secondary achalasia (pseudoachalasia) caused by malignant tumor at the gastro-esophageal junction or less commonly, by benign conditions such as chagas' disease. It can occur at any age with male and female equally affected but it commonly diagnosed in age between 25 to 60 years.

The main symptom of achalasia is dysphagia, typical from solid to liquid; it is a slow progressive disease which developed weeks to years and increases as the disease progress. Its leads to weight loss and complication further as food entering trachea causes bronchopulmonary infections and even aspiration pneumonia in 7% to 8% of the patients (Farrokhi et al. 2007). Usually the etiology is unknown. Some familial cases have been reported, but the rarity of familial occurrence does not support the hypothesis that genetic inheritance is a significant etiologic factor.

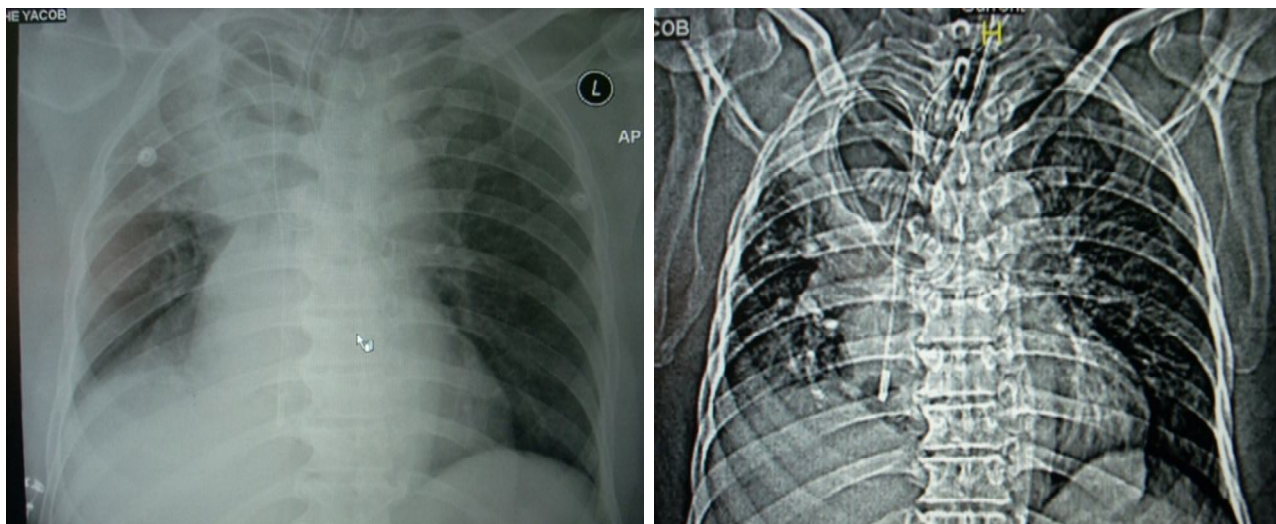


Figure 1

Noted the R/T coiled in the right hemithorax, unable to visualise the tip of R/T in the stomach. There's also right upper mass lesion with widening of the mediastinum



Figure 2

Dilated esophagus occupy right side of mediastinum, coiling of the R/T seen within dilated upper thoracic esophagus with tip seen within the distal part, no evidence of perforation. Focal area of narrowing seen at C5-C6.

Association of achalasia with viral infections and auto-antibodies against myenteric plexus has been reported, but the causal relationship remains unclear.

The diagnosis is based on history of the disease, radiography (barium esophagogram), and esophageal motility testing (esophageal manometry). Endoscopic examination is important to rule out malignancy as the cause of achalasia. There is no cure for achalasia, the goal of treatment is relief of symptoms and to improve esophageal emptying. Current medical and surgical therapeutic options (pneumatic dilation, surgical myotomy, and pharmacologic agents) aimed at reducing the LES pressure and facilitating esophageal emptying by gravity and hydrostatic pressure of retained food and liquids. Although it cannot be permanently cured, excellent palliation is available in over 90% of patients.

2. CASE REPORT

A 59-year-old man alleged in motor vehicle accident sustained severe head injury and closed fracture of right femur. He came in with poor GCS (5/15) and was intubated in casualty. After resuscitation, he was stable.

CXR post intubation and ryles tube (R/T) insertion, noted (R/T) coiled in right hemithorax (Figure 1). At that time esophageal perforation secondary to trauma was diagnosed. In view of possible esophageal injury, CT scan of the thorax and abdomen was done (Figure 2). Diagnosis of achalasia cardia was made. On further questioning, patient's wife only can gave a history of dysphagia for 6 months and unable to noticed any changes in patient's diet or any other symptoms (Figure 3). After 48 hours and confirmed no contrast extravasation to indicate perforation, we proceed with OGDS (Figure 4) which showed dilated proximal esophagus and tight narrowing at lower esophagus. No mass seen. With this evidence, diagnosis of achalasia was confirmed. He was treated accordingly- neurosurgery proceed with extraventricular drainage (EVD) and intracranial pressure (ICP) monitoring and subsequently tracheostomy as patient remained poor GCS, orthopedic – did open reduction and plating of right femur for the femur fracture. As for surgery- we proceed with feeding gastrostomy. Biopsy taken revealed non specific esophagitis. After almost 2 months in the ward patient succumbed to death due to sepsis secondary to hospital acquired pneumonia.

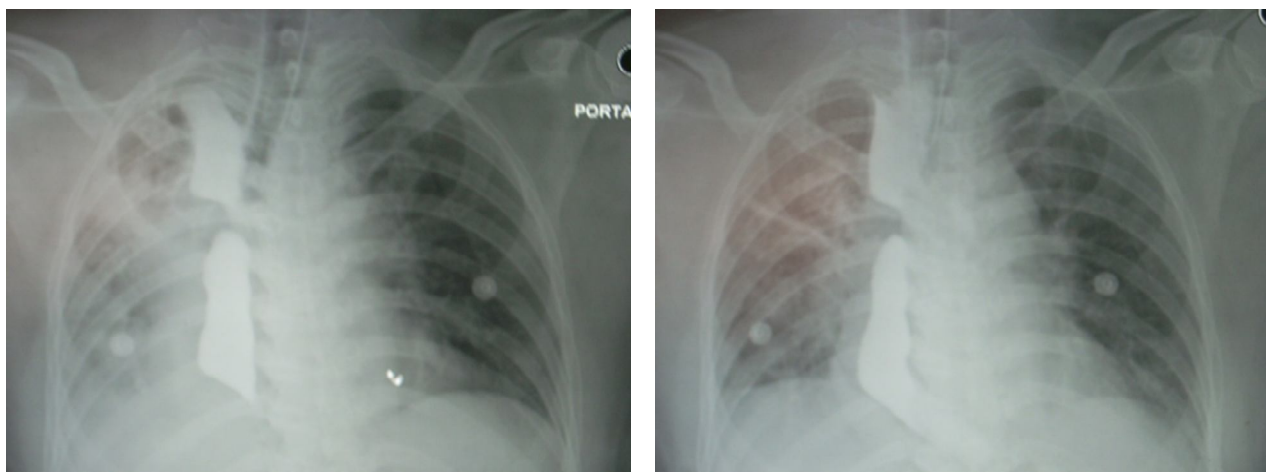


Figure 3

Contrast was injected through R/T showed contrast seen layering the dilated esophagus and focal area of narrowing seen at C5-C6. Contrast seen in the stomach



Figure 4

Proximal esophagus grossly dilated with tight stricture 40 cm from the incisor with mucosal reaction, unable to proceed further. Biopsy taken.

3. DISCUSSION

Achalasia was first recognised by Sir Thomas Willis in 1674; where he suggested that the disease is due to the loss of normal inhibition in the distal esophagus and the etiology of achalasia is remain poorly understood. Epidemiologically, achalasia is a rare disease, with an incidence of 1:100 000 per year (Richter et al. 2010). The onset is insidious and usually the patient seeks attention after presenting the symptoms for many years. As a result, achalasia is not diagnosed until averages of five years after initial complaint begin. This leads to an unnecessary loss of quality of life and to complications such as megaesophagus and esophageal carcinoma. The etiology of achalasia is unclear however Clark et al 2010 suggested an underlying autoimmune process is the cause when histologic resection of the achalasia have revealed inflammation around the myenteric plexus which is predominantly contains T-lymphocytes (CD3 positive).

Clinical presentation of achalasia is often late, most of the patient are symptomatic for years before seeking medical attention. Majority will come with dysphagia for

solids and liquids, regurgitation and chest pain. As many as 70–97% of patients with achalasia have dysphagia for both solids and liquids at presentation (Mikaeli et al. 2009). Over the years, patients learn to accommodate to their problem by using various maneuvers, including lifting the neck or drinking carbonated beverages to help empty the esophagus or walking around after a meal (to accomplish bolus passage through the aperistaltic esophagus and across the lower esophageal sphincter barrier). This is the reason why patient turn up late to seek for help. However, patients with rapid onset of dysphagia (< 6 months), weight loss and age >50 years, pseudoachalasia should be ruled out by endoscopic examination and a CT-scan (Gockel et al. 2012). Regurgitation which is not responding to proton pump inhibitor (PPI) therapy also a characteristic of achalasia. Regurgitation of material retained in the dilated esophagus, especially during supine position at night, may lead to aspiration. Weight loss (usually between 5 to 10 kg) is not present in all patients (Farrokhi et al. 2007) Chest pain is another presenting symptom of achalasia (17%–95%). The occurrence of this symptom is unrelated to the LES pressure (Mikaeli et al. 2009).

From a limited history from the patient's wife, it seems that our patient did not manifest all of the classical symptoms and the history is only 6 months. Our speculation is that, this patient might didn't complaint much to his wife and kept to himself as he might manage to accommodate his symptoms as mention above. The diagnosis of idiopathic achalasia is relatively straight forward with a well-documented medical history, endoscopic, radiography,

and esophageal motility testing. A combination of these investigations, which is esophagogastroscope together with barium swallow and manometry, is part of standard diagnostics for suspected achalasia. However, diagnosis can only be established endoscopically in approximately one third of all achalasia patients, and it depends on the stage the disease (Howard et al. 1992). In this case, the esophagus is grossly dilated with resistance encountered at the gastroesophageal junction and unable to overcome the resistance by the endoscope. These indicate that this patient already has an advanced case of the disease on presentation. Endoscopy is also important to exclude other causes of dysphagia, these include esophageal or stomach tumors or strictures (caused by scar tissue or inflammation).

Depending on the stage of the disease, the esophagus usually appears dilated in a barium swallow, shows a loss of peristalsis in the distal portion, and shows a tapered, thread-like area in the cardiac region, called a "bird's beak" because of its shape but it may occasionally be normal (Cameroon 2008). Although a barium swallow reveals abnormalities in almost every patient, they are often nonspecific, so achalasia is successfully diagnosed only in approximately two-thirds of cases (Howard et al. 1992). Manometry must be considered as the gold standard for diagnosis of the disease. Manometry findings consistent with achalasia include incomplete lower esophageal sphincter relaxation, which is present in more than 80% of patients; elevated LES pressure, which is present in some patients; and diminished to absent peristalsis in the distal esophagus. Achalasia is an incurable disease, thus management is mainly for symptomatic relief either medically or surgically managed. The goal of management is to relieve patient symptoms, improve esophageal emptying by eliminate the non-relaxing LES and prevent development of megaesophagus. Medical treatment is aimed at decreasing the LES tone and its include nitrates, calcium channel blocker, however the effect is short term and the outcome is poor (Cameron, 2008). The drug also often has harmful side effects. Thus drug therapy is reserved for patients not willing or unable to undergo any procedure.

Next is endoscopic treatment which further divided into botulinum toxin (BT) injection, pneumatic balloon dilator and the latest is Peroral endoscopic myotomy (POEM). POEM is a novel endoscopic esophagomyotomy. The toxin injection into LES aims to block the release of acetylcholine from cholinergic neurons and this causes chemical denervation and its can last for few months. However it also act as a temporary management as its only last 6-9 months (Seng et al. 2011). The downfall of BT is, it creates an inflammatory reaction at LES and makes the subsequent surgical is difficult. Pneumatic dilation (PD) is the most effective non-surgical treatment option for patients with achalasia. Pneumatic balloon dilatation decreases esophageal resistance by forceful tearing of LES muscle fiber. Good to excellent relief of symptoms are reported in 50% to 93% of patients (Farokhi et al. 2007). The outcome is varies as it depends on the number of dilatation session, inflation time and it is an operator dependent. Immediate and short term relief is excellent but high recurrence rate is reported for long term. Only half of the patient benefits for more than one year (Seng et al. 2011). This treatment is also reserved for patients not willing or unable to undergo any surgical procedure.

The latest endoscopic tool is per oral endoscopic myotomy (POEM) for the treatment of achalasia as describe by Inoue et al 2010. POEM is performed by creating a submucosal tunnel and endoscopic myotomy of circular muscle bundles. He also observed symptoms of achalasia decreased or disappeared in all patients. The LES pressure decreased significantly after the procedure. Even though no specific complications related to POEM were reported but it is associated with serious complication from esophageal perforation. Surgical management of achalasia involves performing a Heller myotomy (HM) as describe by Ernest Heller in 1913, combined with an antireflux procedure (Toupet or Dor). HM, both the anterior and posterior LES were disrupted then a modified version by single anterior myotomy was introduced. Once it performed only through laparotomy or thoracotomy approach but now with laparoscopic era, laparoscopic myotomy has become the standard surgical approach. It has showed to produce excellent long term results in 90%-98%. The antireflux procedure helps to avoid late stricture due to postoperative gastroesophageal reflux (Tadaka et al. 2012). Complication is low around 10-15% and generally mild usually gastroesophageal reflux is the common symptoms but it can be controlled by proton pump inhibitor.

Recurrent dysphagia following surgical myotomy should be investigated to exclude obstructing lesion. Management options include pneumatic dilatation or remyotomy but Zaninotto and colleagues found that most (78%) of the 10 patients with recurrent dysphagia could effectively treated with pneumatic dilatation. They concluded that pneumatic dilatation should be considered as the first line treatment for patients with persistent dysphagia and reoperation should be reserved for those who do not respond. Unfortunately for this patient, we cannot offer him any of these treatment based on his condition, he presented to us with severe head injury which makes the above option is impossible. Feeding gastrostomy is the best option for the patient at the moment.

4. CONCLUSION

There are still many challenges and questions to be answered regarding achalasia and its treatment. We need to understand the triggers leading to the destruction of the esophageal and LES neurons and possibly how to prevent

these insults. If this is due to an autoimmune process, one possible alternative therapeutic approach may be immune modulating drugs. Future large, randomized, prospective trials will need to compare laparoscopic Heller myotomy and pneumatic dilation to address the superiority of one technique to the other over a 5 to 10 year period, or to determine which therapies should be reserved for a certain subset of patients.

REFERENCES

1. Clark SB, Rice TW, Tubbs RR, Richter JE, Goldblum JR. The nature of the myenteric infiltrate in achalasia: an immunohistochemical analysis. *Am J Surg Pathol*, 2000, 24(8), 1153-8
2. Edition JLCt. Current Surgical Therapy- Achalasia of Esophagus. *Surgical Textbook*, 2008, Section 1: Esophagus
3. Farrokhi F, Micheal F Vaezi. Idiopathic (primary) Achalasia. *Orpanet J*, 2007, 2(38), 1750- 1760
4. Giovanni Zaninotto FACS, Giuseppe Portale, Giorgio Battaglia, Dianella Molena. Etiology, Diagnosis and Treatment of Failures after Laparoscopic Heller Myotomy for Achalasia. *Annals Of Surgery*, 235(5), 186-192
5. Gockel I, Muller M, Schumacher J. Achalasi;-a Disease of Unknown Cause That Is Often Diagnosed Too Late. *Dtsch Arztebl Int*, 2012, 109(12), 209-14
6. Howard PJL Maher, Pryde A, Camerron EW, Heading RC. Five Years Prospective Study and Incidences, Clinical Features and Diagnostic achalsia in Edinburg. *International Journal of Gastrointestinal and Hepatology*, 1992, 33(8), 1011-1015
7. Inoue H, Minami H, Kobayashi Y, Sato Y, Kaga M, Suzuki M, Satodate H, Odaka N, Itoh H, Kudo S. Peroral Endoscopic Myotomy (POEM) for Esophageal Achalasia. *Endoscopy*, 2010, 42(4), 265-71
8. Javad Mikaeli FI, Reza Malekzadeh. Achalasia: A Review of Western and Irani Experiences. *World Journal of Gastroenterology*, 2009, 15(40), 5000-5009
9. Katada N, Sakuramoto S, Yamashita K, Shibata T, Moriya H, Kikuchi S, Watanabe M. Recent Trends in the Management of Achalasia. *Ann Thorac Cardiovasc Surg*, 2012, 18(5), 420-8
10. Richter JE. Achalasia - an update. *J Neurogastroenterol Motil*, 2010, 16(3), 232-42
11. Seng Kee Chuah PIS, Keng Liang Wu, Deng Chyang Wu, Weichen Tai. 2011 Update on Achalasia. *World Gastroenterology*, 2011, 18(14), 1573-1578