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Topic-Long-Standing Achalasia with Megaesophagus Mimicking Hiatus Hernia and Presenting as Hematemesis

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ABSTRACT

Achalasia is a rare esophageal motility disorder characterized by impaired relaxation of lower esophageal sphincter and esophageal aperistalsis due to parasympathetic denervation. Achalasia usually presents in childhood or early adult life with slow-onset dysphagia. It is very rare for achalasia to present in late adult life, and that too with profuse upper gastrointestinal bleed as the first symptom. We report a case of achalasia with megaesophagus that appeared as a large hiatus hernia on imaging and endoscopy. A 57-year-old man presented with hematemesis and shock. No definite history of prior dysphagia was available. After initial resuscitation, the chest radiograph showed mediastinal widening with a huge dilated viscus in the right hemithorax. CT suggested herniation of the stomach through the esophageal hiatus. Endoscopy showed multiple ulcers with frank blood in a grossly dilated viscus thought to be hiatus hernia. Diagnosis of achalasia was confirmed intraoperatively when a normal stomach and hiatus was found. Heller's cardio myotomy with partial anterior fundoplication procedure was performed. The patient is being systemically optimized for definitive esophagectomy.

INTRODUCTION

Achalasia is an idiopathic primary esophageal motility disorder and incomplete relaxation of the lower esophageal sphincter [1-3]. Destruction of the nerve to lower esophageal sphincter is primary pathologic process and the degeneration of neuromuscular function of body of esophagus is secondary. Achalasia is relative rare disorder with incidence rate of 1 in 100000 and prevalence of 1 in 10000 in United States with equal distribution in males and females [2,4]. Classical triad of achalasia consists of dysphagia, regurgitation and weight loss. Patients with achalasia are at high risk for developing esophageal ulcer, bleeding, esophagitis, aspiration pneumonia spontaneous rupture of esophagus [5]. In extremely rare situation, long-standing primary achalasia can lead to esophageal hemorrhage due to underline mucosal irritation and ulcer formation [6, 7]. We report a rare case of achalasia with upper gastrointestinal bleeding associated with esophageal ulcer formation with megaesophagus that mimics hiatus hernia.

CASE REPORT

A 57-year-old male presented with complains of hematemesis with regurgitated food and water. His symptoms accompanied by sweating, fatigue. Patient had no history of regular use of NSAIDs, aspirin, immunosuppressant, any weight loss, Malena and hoarseness of voice. On physical examination patient had pale conjunctiva with decrease capillary refill time. Routine blood test showed Hemoglobin- 9.6 gm/dl, normocytic normochromic anemia which was significantly lower than baseline, white blood cell counts of 10 k/ml, platelet count 140 k/ml and potassium level 2.7 mmol/L. Chest radiograph reported as mediastinal widening with dilated viscus occupying most of right hemithorax (**Figure 1**). HRCT chest reported large tortuous dilated viscus occupying right hemithorax compressing right lung parenchyma suggesting of large rolling hiatus hernia (**Figure 2**). An upper GI endoscopy showed a dilated tortuous lumen, possibly of the esophagus. The entire scope was accommodated within this tortuous viscus. There were multiple punched-out ulcerations present till the maximum visibility of the scope (**Figure 3**). The lumen was further filled with frank blood with large hemorrhagic tissue adhered to the walls (**Figure 4**). The provisional diagnosis was a large, complicated rolling-type of hiatus hernia through the esophageal hiatus.

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The patient was planned for exploratory laparotomy. Intra-operatively, to our surprise, the stomach found normal intraabdominal (**Figure 5**). There was slight dilatation of the esophageal hiatus and of the abdominal part of the esophagus, however, no obvious herniation was identified. On passing a nasogastric tube from the head end, there was copious drainage of dark hemorrhagic contents. The tube was not palpated in the stomach or the abdominal esophagus. A diagnosis of achalasia and megaesophagus was then considered, bearing in mind that the nasogastric tube of small caliber was rolled-up inside the megaesophagus. A wide-bore (36 Fr) gastric calibration tube (GCT) used in bariatric procedures was then passed. Failure of the GCT to pass across the lower esophageal junction (LES) was considered to be confirmatory for the diagnosis of achalasia. A Heller's cardiomyotomy was performed over the LES (total of 8 cm) and gastric fundus, which enabled the GCT to pass into the stomach. An anterior fundoplication (270°) was used to strengthen the defect and the nasogastric tube was passed till the pylorus.

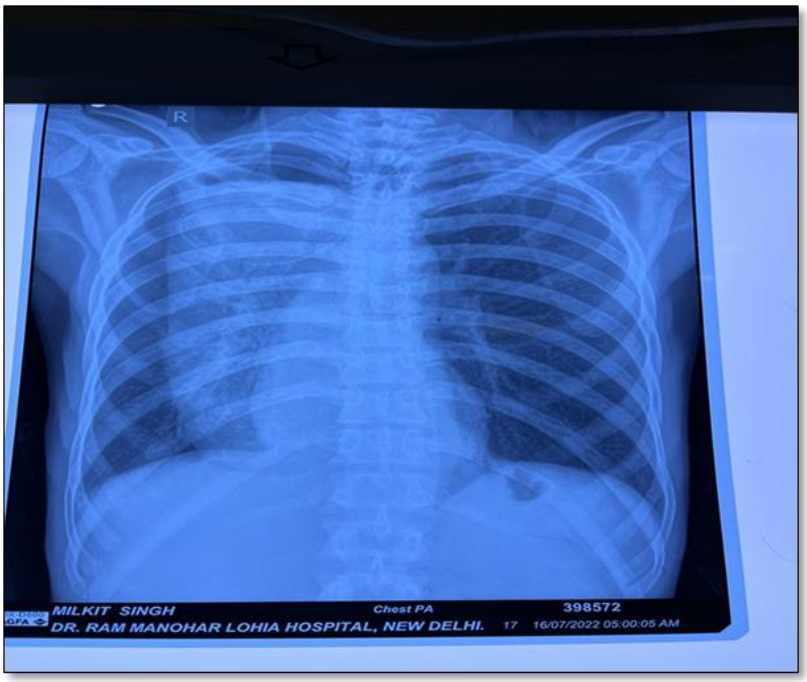

Figure 1. Chest Radiograph shows mediastinal widening with detailed viscus occupying Right hemothorax.

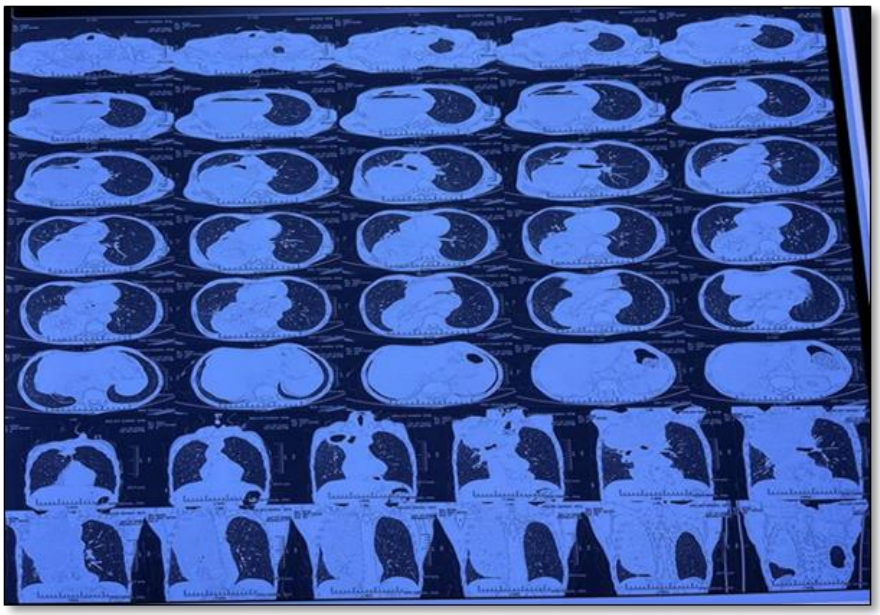

Figure 2. HRCT Chest shows large Tortuous detailed viscus occupying right hemithorax.

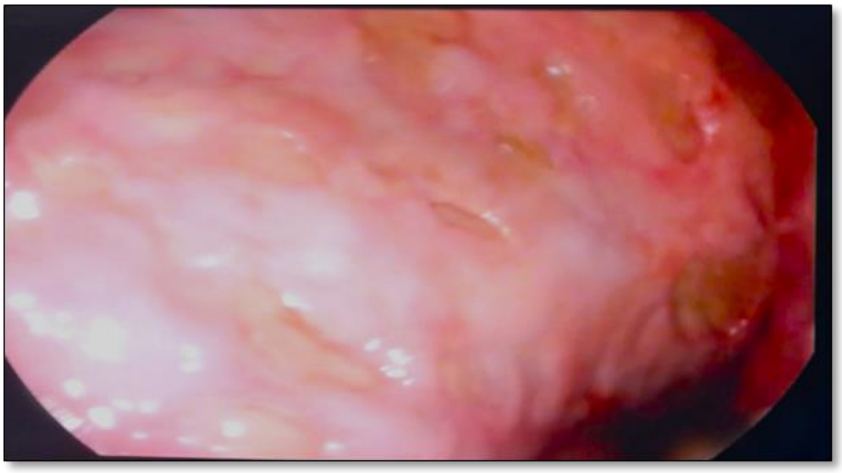

Figure 3. Upper GI endoscopy shows multiple punched out ulcerations.

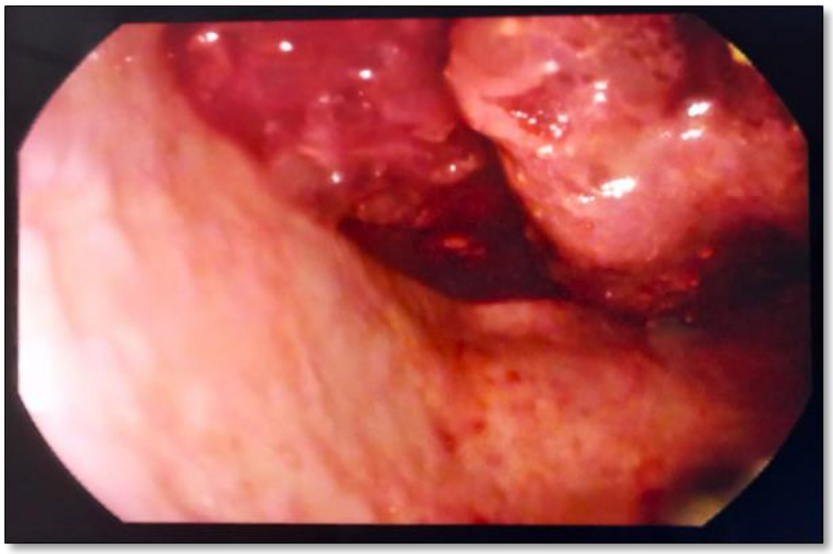

Figure 4. Upper GI endoscopy shows lumen with large hemorrhagic tissue.

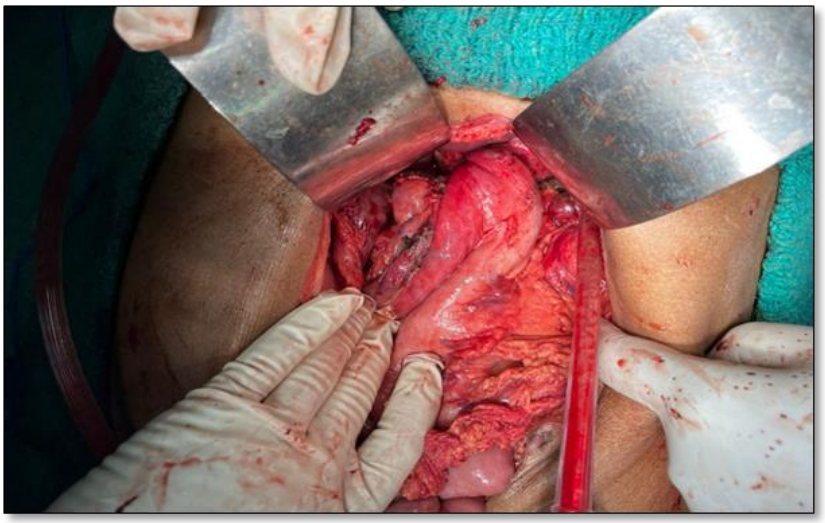

Figure 5. Intraoperatively stomach found Intraabdominal.

The patient recovered over 2 weeks postoperatively with enteral and parenteral nutrition. He was discharged on oral feeding on POD 17, and is being considered for elective transthoracic esophagectomy after optimization.

DISCUSSION

Achalasia is an esophageal motility disorder due to neurogenic degeneration which result in hypertension of lower esophageal sphincter (LES), with consequent failure of LES relaxation. Achalasia is categorized based on etiology - primary or idiopathic, and, secondary if caused by parasitic infections like Trypanosoma's Cruzi seen in chagas disease or, if associated with metabolic disease or tumors [4,5]. Presenting symptoms of achalasia consists of dysphagia, regurgitation and weight loss. The gold-standard test for diagnosis is esophageal manometry which demonstrates esophageal aperistalsis and increase LES pressure.

It is very rare for a patient of achalasia to present with upper gastrointestinal bleeding; the postulated mechanisms are esophageal ulcer or severe esophagitis due to loss of the mucosal protective barrier by the uncontrolled gastric acid reflux and by prolonged stasis [8]. Infections like Candida species, cytomegalovirus, and herpes simplex virus may be contributory in immunocompromised patients. Direct mucosal damage from the ingestion and stasis of caustic agents like tannins, nonsteroidal anti-inflammatory drugs (NSAIDs), bisphosphonates, and certain antibiotics may also complicate a pre-existing achalasia. In extreme cases, achalasia may present as near-fatal esophageal bleeding. Other long-standing complications can be megaesophagus, aspiration pneumonia, esophageal rupture, malnutrition, esophageal squamous cell carcinoma [2,9]. It is evident that patients who develop large episodic hematemesis from underlying ulcers with end-stage complications like megaesophagus, malignancy and cases refractory to medical treatment need to undergo esophagectomy [10].

Our patient presented with upper GI bleed with massive esophageal dilation mimicking hiatus hernia on UGI endoscopy and imaging. The diagnosis of achalasia was confirmed intra-operatively after failure to identify any intra-abdominal pathology. The standard surgical treatment involves pneumatic dilation or Heller's cardio myotomy which both aim to reduce LES resting pressures. We performed Heller's cardio myotomy with anterior fundoplication as a temporizing measure in our moribund patient; we have further planned for elective transthoracic esophagectomy after optimization.

There are inherent challenges in the diagnosis and management of complicated achalasia, as evident from our case. With a large sigmoid dolichol mega esophagus undiagnosed previously, investigations like CT and endoscopy may also be limited in the emergency situation. The endoscope is often limited by the amount of blood, necrotic luminal contents and the increased tortuous length. Multiple endoscopies may be required to clearly diagnose the condition, since manometry is not a feasible option in these situations. This has also been documented in earlier reports [2]. Probably, intra-operative confirmation as we performed is the only feasible method.

CONCLUSION

This case demonstrates rare case of long-standing achalasia with megaesophagus that mimics hiatus hernia. Multiple endoscopies may be required to clearly diagnose the condition, since manometry is not a feasible option in these situations.

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