

# Spontaneous esophageal intramural hematoma in a young man wrongly diagnosed as achalasia

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## ABSTRACT

**Intramural hematoma of the esophagus is a rare but well described type of acute injury of the esophageal wall and it is more frequently being recognized throughout the world. Patients usually present with acute retrosternal or epigastric pain, minor hematemesis and dysphagia. The condition is mostly seen in women with abnormal coagulation and it can either occur spontaneous or induced by trauma or transeophageal procedures. It is associated with food impaction and vomiting. Esophageal intramural hematoma has also been reported in young and healthy patients. Case reports with coexisting achalasia are limited. Management is conservative and its course is benign.**

**Keywords:** Esophageal Intramural Hematoma; Achalasia; Endoscopy; Computed Tomography

## 1. INTRODUCTION

Esophageal intramural hematoma (EIH), also known as esophageal dissection, is a rare condition which sometimes occurs spontaneously. Known predisposing factors include coagulation disturbances, endoscopic interventions, foreign body ingestion and food-induced injury. It is most commonly seen in elderly women and a couple of hundred cases have been described worldwide since its first report in 1957 [1]. Intrinsic esophageal disease is uncommon in patients with EIH.

EIH is generally associated with retching and vomiting, which causes a sudden increased transmural pressure, and the hemorrhage occurs subsequently within the submucosal tissues. This entity is thus different from the well-known Mallory-Weiss and Boerhaave syndromes.

The diagnosis of EIH is mostly made by contrast CT-scan, endoscopic ultrasound (EUS), or magnetic resonance imaging [2]. It has been suggested that endoscopy is relatively contraindicated in the evaluation of EIH as air insufflation could worsen the injury. Conservative

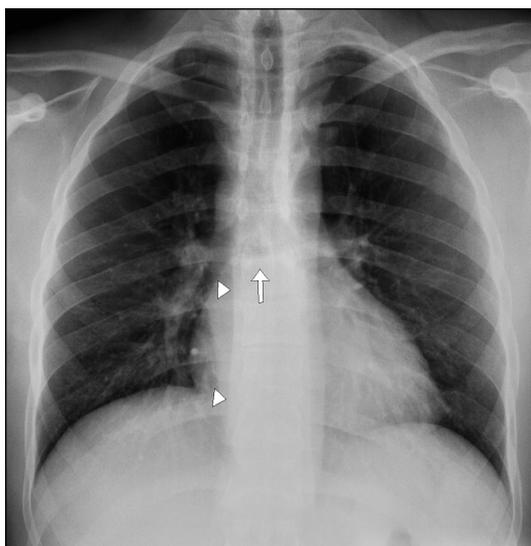
therapy is the treatment of choice in almost all cases. Surgery is indicated only in patients presenting with massive hematemesis or in patients with severe mediastinitis. Conservative treatment consists of fasting for several days and the administration of intravenous fluids, total parenteral nutrition, anti-emetics and proton pump inhibitors. Rarely, when the dysphagia persists under conservative measures, endoscopic treatment can be considered to resolve the hematoma.

The present case describes a young man with acute thoracic and epigastric pain, grave nausea and fever. The diagnosis of spontaneous esophageal intramural hematoma (SEIH) was established and the patient was successfully treated conservatively.

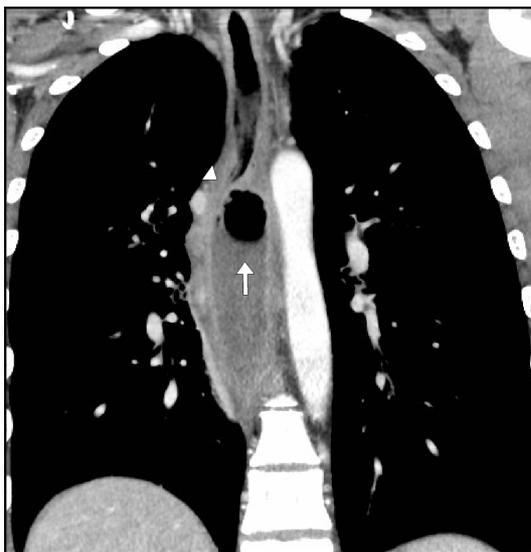
## 2. CASE PRESENTATION

A 29-year-old man was admitted to the emergency department experiencing nausea, epigastralgia and retrosternal pain after eating a snack. His symptoms had started three days before and did not improve with ranitidine. There was a regular reflux of fluids, stained with some blood. In his medical history we only noted mild asthma for which he uses a short-acting B2-adrenergic receptor agonist by inhalation when needed. On admission, the patient showed the following vital signs: blood pressure 128/70 mmHg, pulse rate 100 bpm, temperature 39.5°C and a 100% transcutaneous oxygen saturation. Clinical examination revealed a painful epigastric region upon palpation with local rebound tenderness. Cardiopulmonary assessment was normal. Blood analysis showed marked inflammation with a C-reactive protein (CRP) level of 293 mg/L (normal: <5 mg/L) and a fibrinogen level of 914 mg/dL (normal: 180 - 400 mg/dL). His hemoglobin level was normal upon admission and the white blood cell count was mildly increased at  $10.9 \times 10^3/\text{mm}^3$  with 85% of neutrophils. The prothrombin time was out of range (51%) while the aPTT was normal. The electrocardiogram was consistent with sinus tachycardia. The abdominal X-ray showed no abnormalities.

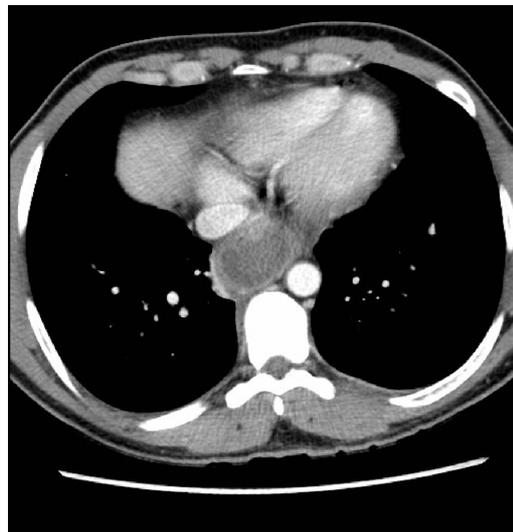
An urgent abdominal CT scan suggested achalasia, based on the pronounced dilatation of the distal esophagus, and the heterogeneous collection that was thought to be caused by food stasis. The chest X-ray presented an extra paravertebral line at the right and a small infracarinal fluid level (**Figure 1**). A computed tomography of the chest was then performed, revealing an intramural collection with deviation of the esophagus. A clear deviation of the true esophageal lumen was visible at the proximal end of the intramural collection (**Figures 2 and 3**). There were no radiographic signs of mediastinitis



**Figure 1.** Chest X-ray presenting a right-sided paravertebral line (arrowheads) and a subcarinal air-fluid level (arrow).



**Figure 2.** Chest CT scan, coronal reconstruction, demonstrating the air-fluid level in the intramural collection (arrow) and deviation of the true esophageal lumen to the right (arrowhead).



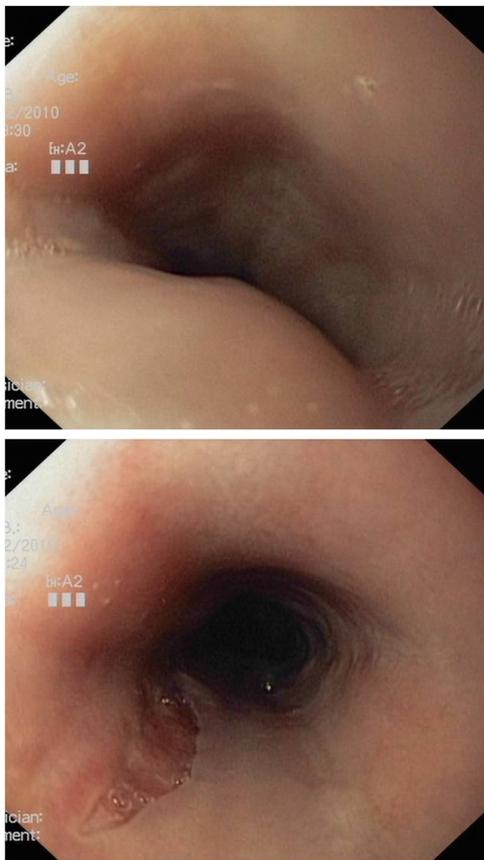
**Figure 3.** Axial view of the lower chest, showing a distended distal esophagus mimicking achalasia.

nor perforation. The patient was put on nil per os, total parenteral nutrition, intravenous fluids, anti-emetics, antipyretics and pantoprazole intravenously. Because of the elevated inflammatory markers in the blood and the high fever, broad-spectrum antibiotics (piperacillin/tazobactam) were also started. Multiple blood cultures were negative. The hemoglobin level of the patient dropped to 12.0 g/dL on day 1, coming from 15.5 g/dL. The symptoms improved significantly within a couple of days. Five days after admission a follow-up CT scan showed spontaneous involution of the hematoma, which was even more obvious on day 11. Blood tests confirmed the positive evolution with normalisation of both CRP and PT levels. Ten days after admission the patient was allowed to drink liquids. Upper endoscopy was eventually performed on day 13 which revealed residual bulging and linear ulcerations of the distal esophagus (**Figure 4**). The patient was then put on a semi-liquid diet and oral proton pump inhibitors. He was discharged from the hospital in an excellent condition on day 14.

### 3. DISCUSSION

EIH is a rare cause of acute chest pain and, as the present case report demonstrates, it can occasionally be seen in young and healthy men with no obvious underlying cause (spontaneous esophageal intramural hematoma, SEIH). CT scanning confirmed the diagnosis revealing a marked intramural collection with deviation of the esophageal lumen to the right, consistent with stage IV hematoma [3].

Interestingly, the initial abdominal CT scan suggested possible achalasia in the present case. The coexistence of EIH and achalasia has been described recently by Chu YY *et al.* in a case series of five patients [4]. These au-



**Figure 4.** Upper endoscopy on day 13 revealing bulging of the hematoma in the esophageal lumen at 25 cm (up) and linear ulcerations at 33 cm from the mouth (bottom).

thors recommend conservative treatment for EIH and classical dilatation for achalasia. The clinical evolution of our patient however did not confirm the diagnosis of achalasia. It was when a CT scan of the thorax was performed that a deviation of the esophagus was spotted and an esophageal mucosal dissection suspected.

The triad of chest pain, dysphagia and hematemesis is present in 35% of patients [5]. Fever is also reported in previous cases [6]. Our patient presented with high fever which pointed in direction of secondary mediastinitis. Yet, the CT scan detected no free air and the general condition of the patient remained favorable. We believe that the fever and the elevated CRP level could be explained by superinfection of the hematoma. Piperacillin/tazobactam was chosen as a broad-spectrum antibiotic and the fever faded quickly. The coagulation disorder, which recovered spontaneously, was most likely caused by the disseminated inflammatory response leading to transient factor VII deficiency [7]. Follow-up CT scans were used to evaluate the hematoma and an upper endoscopy one month after discharge showed fully healed esophageal mucosa.

In the literature we found 96 cases of SEIH, 74% of which were female with a mean age of 59 years [4-6,8-34]. Nineteen% of the cases had an abnormal coagulation, either acquired or induced by medication. 58% presented with hematemesis, while only 11% of the cases had a fever. Virtually all presented with severe chest or epigastric pain.

Currently, there are no guidelines for the management of SEIH, although an algorithm has been suggested by Beumer *et al.*, proposing conservative treatment in all hemodynamically stable patients without signs of perforation on imaging, with initiation of broad-spectrum antibiotics [30]. Patients should fast until clear clinical improvement occurs and realimentation should then be initiated gradually and with caution. Endoscopy seems to be indicated when symptoms last for more than a week but should be performed with caution as perforation of the esophagus has been reported [26]. However, at least three papers describe successful endoscopic treatment by incision of the mucosal bridges in patients whose symptoms did not resolve spontaneously after several weeks [31-33]. One patient developed an esophageal stricture after this treatment which had to be endoscopically dilated. Mucosal healing is thought to be complete within three to four weeks [34]. SEIH is a benign condition with an excellent prognosis, in contrast to the high mortality rates observed with Boerhaave syndrome.

#### 4. CONCLUSION

Spontaneous esophageal intramural hematoma is a rare disorder, and therefore often a difficult and delayed diagnosis. Typical symptoms include chest pain, nausea, dysphagia and hematemesis. Occasionally, it is accompanied by fever and an inflammatory response of uncertain origin. Conservative management is efficacious in most cases.

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