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Case Report

Undiagnosed aorto-esophageal fistula causing intramural hematoma of the esophagus

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ABSTRACT

Aorto-esophageal fistula (AEF) is a rare, but life-threatening cause of intramural hematoma of the esophagus (IHE). Typical clinical presentation of AEF includes mid-thoracic pain and sentinel hemorrhage followed by massive, often fatal, hematemesis, with the period between sentinel hemorrhage and massive hematemesis generally varying from hours to days. This is a case of a 61-year-old male who presented with chest pain after development of an aorto-esophageal fistula and associated intramural hematoma of the esophagus. The fistula and associated hematoma were initially mischaracterized on imaging, and went undiagnosed for approximately 2 weeks before being iatrogenically disrupted during endoscopy. Though this case was successfully treated, aorto-esophageal fistulas are associated with a high mortality, and aorto-esophageal fistula/intramural hematoma of the esophagus should always be considered in the differential of an esophageal mass.

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Introduction

Aorto-esophageal fistula (AEF) is a rare, but life-threatening cause of intramural hematoma of the esophagus (IHE). Typical clinical presentation of AEF, often remembered as the Chiari Triad, includes mid-thoracic pain and sentinel hemorrhage followed by massive, often fatal, hematemesis. The period between sentinel hemorrhage and massive hematemesis varies, but is generally hours to days. Presented here is a case of undiagnosed aorto-esophageal fistula causing intramural hematoma of the esophagus which remained stable for at least 2 weeks before being iatrogenically disrupted during endoscopy.

Case report

A 61-year-old male presented to the emergency department (ED) with dysphagia and chest pain extending into his throat for 1 week, worsened with inspiration and swallowing. Two hours prior to arrival, he had noticed swelling in his neck and prominence of his neck veins. He also reported several weeks of night sweats, and a 20 lb (9 kg) unintentional weight loss. On arrival, the patient was mildly tachycardic, but hemodynamically stable, and work-up for acute coronary syndrome was negative. A CT angiogram of the chest showed a descending thoracic aortic aneurysm with areas of irregularity just above the level of the diaphragm, concerning for rupture and pseudoaneurysm (Fig. 1), as well as homogeneous soft tissue density encasing the esophagus, extending from the thoracic inlet to the diaphragmatic hiatus (Figs. 2 and 3). There was also mild mediastinal and left hilar adenopathy and two

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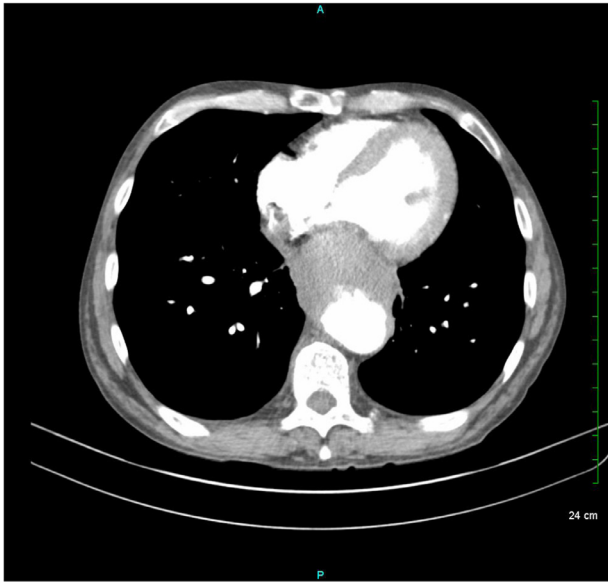


Fig. 1 – Axial CT image at the level of the distal descending thoracic aorta demonstrates irregularity of the aortic wall and surrounding soft tissue density, concerning for aortic pseudoaneurysm and hematoma.

spiculated right upper and left lower lobe nodules. The initial radiology report described the soft tissue mass around the esophagus, the adenopathy, and the spiculated nodules, but unfortunately failed to mention the aortic pseudoaneurysm. As a result, the care team's efforts initially concentrated on characterizing a suspected malignancy, with subsequent subcarinal lymph node needle aspiration, bone scan, and CT of the brain yielding inconclusive results.

Approximately 1 week after presentation to the ED, the patient was transferred to another institution for further workup of the suspected malignancy, and after several other negative tests, endoscopy was performed, with multiple forceps biopsies taken of the distal esophagus (Fig. 4). Several hours following biopsy, the patient experienced massive hematemesis, with a precipitous drop in blood pressure. A CT angiogram of the chest was repeated which showed a descending thoracic aortic pseudoaneurysm with a large mediastinal hematoma compressing and displacing the esophageal lumen (Figs. 5 and 6). The patient underwent emergent TEVAR for repair of the pseudoaneurysm, and after a successful recovery, was discharged home.

A follow-up CT of the chest 3 weeks later demonstrated stable repair of the aortic rupture (Fig. 7). It also showed an air-filled false esophageal lumen communicating with the true lumen, in place of the now nearly resolved mediastinal hematoma (Figs. 8 and 9), indicating the hematoma had been primarily within the esophageal wall. Ultimately, the findings supported the conclusion that the patient developed an aorto-esophageal fistula (AEF) prior to his presentation to the ED that resulted in an intramural hematoma of the esophagus (IHE). This fistula/hematoma then remained stable for nearly 2 weeks until it was biopsied, when it ruptured into the esophageal lumen.



Fig. 2 – Axial CT images at the level of the upper and mid esophagus demonstrate homogenous soft tissue density encasing the esophagus, initially characterized in the report as a soft tissue mass concerning for malignancy, possibly lymphoproliferative in origin.

Discussion

Both intramural hematoma of the esophagus (IHE) and aorto-esophageal fistula (AEF) are rare conditions, and have overlapping symptoms including chest pain, hematemesis, dysphagia, and odynophagia [1,2]. IHE is generally a self-limited process involving dissection of hemorrhage between the mucosa and the submucosa. It is part of a continuum of esophageal injury including Mallory-Weiss tear and Boerhaave's Syndrome [3]. Dissection through the mucosa can also be seen, resulting in hematemesis, usually low volume, though there are case reports of recurrent massive hematemesis [4]. Causes of IHE include Valsalva maneuver, heavy weight lifting, vomiting, coughing, sneezing, or direct injury from ingested contents, external chest trauma, or endoscopic iatrogenic injury. There are also reports of spontaneous idiopathic IHE [5]. For IHE from these causes, conservative treatment is highly effective. AEF however, while a rare cause of IHE, carries a high mortality rate, and requires surgical treatment [6]. For this reason, AEF should always be considered in cases of esophageal intramural hematoma.

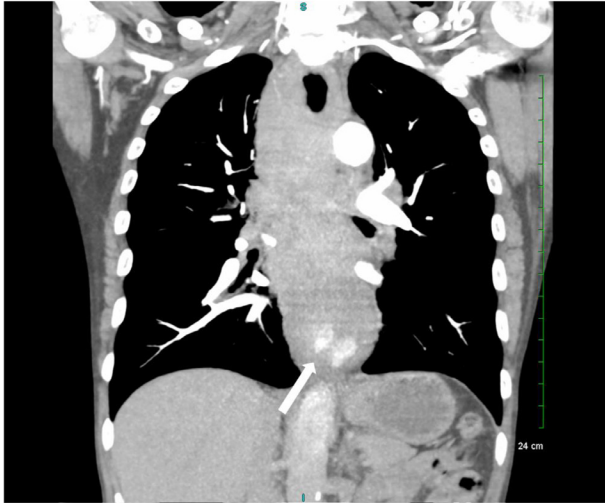


Fig. 3 – Coronal CT MIP image demonstrates the extent of the para-esophageal soft tissue density, extending from the thoracic inlet to the diaphragm. Aortic pseudoaneurysm can be partially seen just above the diaphragm (white arrow).

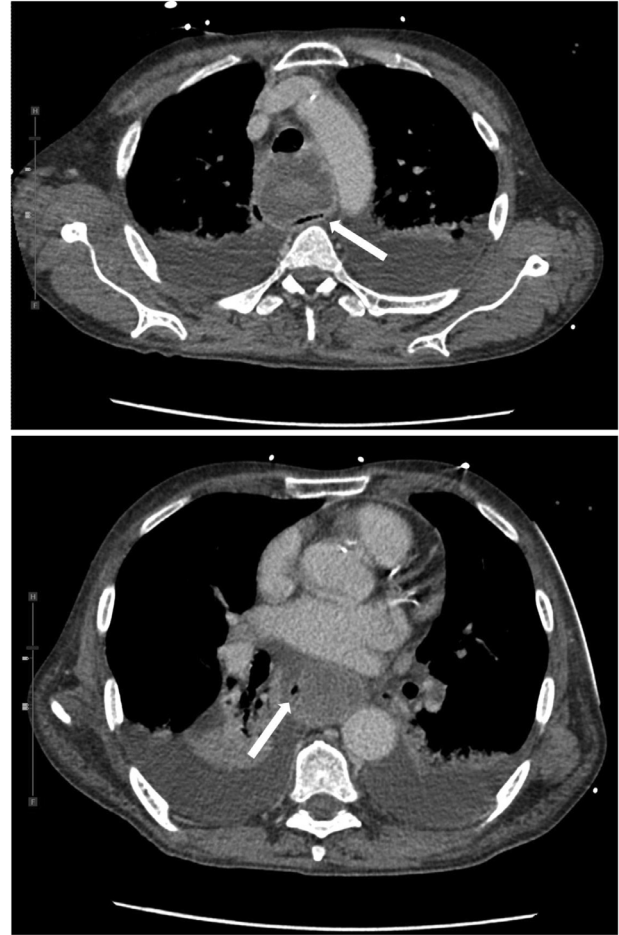


Fig. 5 – Axial CT images at the level of the upper and mid esophagus, approximately 2 weeks after initial presentation, show heterogeneous para-esophageal density, consistent with evolving hematoma. Note the displaced esophageal lumen (white arrows) and bilateral pleural effusions.



Fig. 4 – Endoscopy showed a bulging area of friable mucosa in the distal esophagus which was biopsied. Several hours later the patient experienced massive hematemesis. Biopsy results showed mild esophagitis, but no malignancy.

Aortoesophageal fistula is a rare and life-threatening condition consisting of an abnormal communication between the esophagus and the aorta, allowing blood from the aorta to enter the esophagus at high pressure. This high-pressure flow results in massive hematemesis if the fistula extends all the way to the esophageal lumen, which is the primary cause of death in AEF. Massive hematemesis may also result from other more common processes, including gastroesophageal varices and bleeding upper gastrointestinal tract ulcers. However, these can often be differentiated clinically by the color of the hematemesis, with bright red blood indicating AEF, as opposed to dark red venous blood seen in variceal bleeding, and dark

red arterial blood seen with bleeding ulcers secondary to exposure of the blood to gastric acids [2].

Classic presentation of AEF, known as the Chiari Triad, includes midthoracic pain and sentinel hemorrhage followed by fatal hematemesis. The time between the herald bleed and fatal hemorrhage varies, but typically lasts from hours to days, with the delay thought likely due to facilitation of clot formation by lowered arterial pressure from the initial hemorrhage [7]. As blood pressure returns to normal, the clot is dislodged and bleeding resumes.

The most common causes of AEF reported in the literature are thoracic aortic aneurysm, foreign body ingestion, esophageal malignancy, postsurgical complications, and esophageal ulcers and reflux [2,8–10]. Thoracic aortic aneurysm is by far the most common, representing 51.2% of cases in Hollander and Quick's literature review. By comparison, esophageal malignancy and esophageal ulcers/reflux represented 17% and 2% of cases, respectively. While most aneurysms of the thoracic aorta occur in the ascending

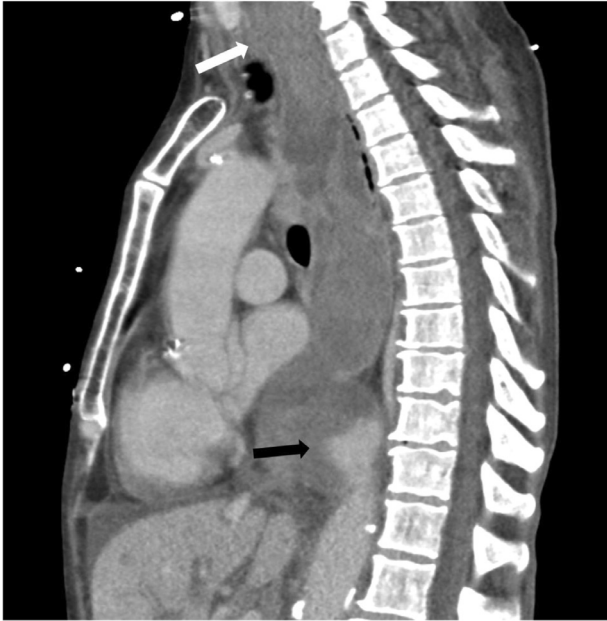


Fig. 6 – Sagittal CT image demonstrates that the intramural hematoma of the esophagus now extends into the neck (white arrow), and again shows the aortic pseudoaneurysm (black arrow).

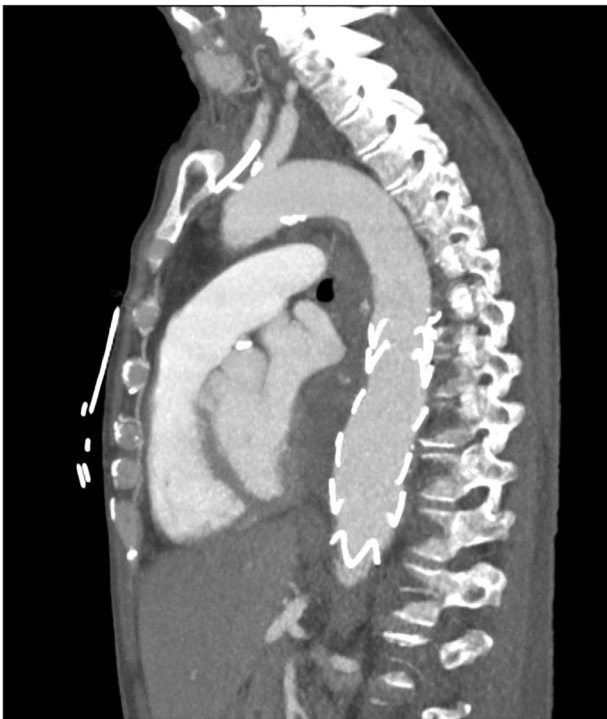


Fig. 7 – Sagittal MIP CT image approximately 5 weeks after initial presentation demonstrates successful repair of the aortic pseudoaneurysm, with some residual hematoma.

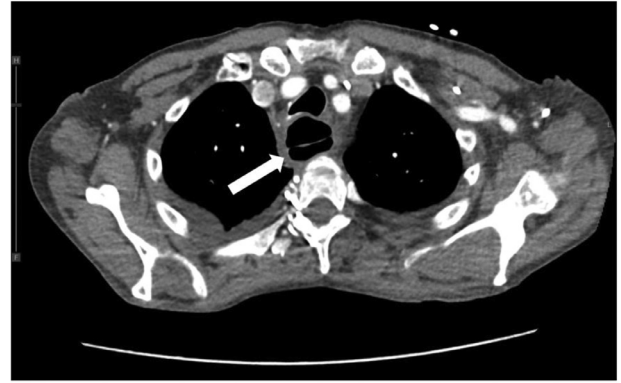


Fig. 8 – Axial and sagittal CT images show the “false” esophageal lumen (arrows) left behind by the resolving intramural esophageal hematoma.

portion, AEF from aortic aneurysm most commonly occurs in the descending aorta, where the aorta and esophagus are closely apposed. However, AEF has been reported in the ascending aorta and aortic arch [2]. Causes of AEF can be classified as primary AEF, caused by aneurysm, esophagitis, etc., and secondary AEF, caused by ingestion, trauma, or iatrogenic causes [7]. Of note, there are reports in the literature of delayed AEF following trauma, some delayed for years [2,11].

Fistula formation in AEF by aneurysm and by esophageal ulcers/reflux is thought to occur by similar mechanisms, both causing an inflammatory process leading to breakdown of the soft tissues of the walls of the aorta and esophagus respectively. Over time, hydrostatic pressure from the aorta causes necrosis and results in propagation of the fistula through the inflamed tissues into the esophagus [10]. AEF from esophageal malignancy is thought to occur by a similar mechanism: in these cases, thrombosis of the vasa vasorum due to



Fig. 9 – Communication between the 2 esophageal lumens was demonstrated by the presence of oral contrast in both spaces (black arrows).

malignancy invasion causes inflammation and ischemia of the aortic wall, leading to erosion and fistulation [2].

Treatment for AEF is invariably surgical, with emergent aortic stenting the standard of care. Owing to contamination with the gastrointestinal tract, up to one quarter become infected. Once the infection is controlled, complex reconstruction then occurs in multiple stages [7,12].

Conclusion

This case represents an unusual circumstance of aorto-esophageal fistula resulting in intramural hematoma of the esophagus that remained essentially stable for at least 2 weeks, from the time of initial presentation to the ED to the time of esophageal biopsy. Given that the patient's symptoms reportedly began a week before presentation, it may have been stable for even longer. In this patient's case, the massive hematemesis associated with the fistula was likely iatrogenic from disruption of the AEF-IHE complex at the time of endoscopy and biopsy.

The definitive etiology of this patient's AEF remains unknown. While the left hilar adenopathy was ultimately determined to be malignant, biopsies of his esophagus showed only mild esophagitis, without evidence of malignancy. Esophagitis itself can cause AEF, but statistically this is a much less common cause of AEF, and the mild degree of the esophagitis makes this even less likely. The most likely cause remains erosion of the aortic aneurysm into the esophageal wall.

Evaluation of the aorta and mediastinum should always include consideration that esophageal wall thickening could represent IHE, and the aorta should be carefully evaluated for the presence of aortic rupture and AEF. This is particularly important as, unlike other causes of IHE, IHE from AEF carries a high mortality rate and requires immediate surgical repair. Any suspicion of AEF should be promptly relayed to the physicians managing the care of the patient to guide treatment and give the patient the best chance of survival.

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