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Massive Hematemesis as Presentation of Congenital Aortic Coarctation With Superinfected Aneurysm and Aortoesophageal Fistula

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Introduction

Aortoesophageal fistula (AEF) is extremely rare in children and usually results from an esophageal foreign body or esophageal surgery.^{1–2} There are two reported cases of AEF from superinfected aortic aneurysms in conjunction with undiagnosed aortic coarctations: an 11-year-old male and a 14-year-old male.^{3,4} Superinfected aortic aneurysms leading to AEF are more common in adults.^{5,6} We report the third and oldest patient with an AEF from an undiagnosed aortic coarctation. This case has recently been published in the cardiology literature;⁷ here we focus on the initial presentation, endoscopic features, and decision making prior to the diagnosis of AEF.

Case Report

A 15-year-old male was transferred to our hospital for acute hematemesis and anemia. Three weeks prior to admission, he developed daily fevers for which he took ibuprofen and naproxen. One week prior to admission, he developed increasing fatigue, a four-kilogram weight loss, diffuse back pain worse in the right scapular area, and non-bloody, non-bilious emesis. The morning of admission, he vomited 3–4 cups of bright red blood. He presented to a local emergency room, where his hemoglobin was 9 g/dL. He was transferred to our hospital, and on arrival, his blood pressure was 148/89 and pulse was 126. A 2/6 vibratory systolic murmur was heard best at the left lower sternal border that radiated upward. He had no back or abdominal tenderness, rash, telangiectasias, splinter hemorrhages or hepatosplenomegaly. His past medical history was negative, including no history of hypertension. Surgical and family histories were non-contributory.

At admission, the patient's white blood cell count was 12,000 cells/uL with 82% neutrophils, and hemoglobin was 7.9 g/dL. INR was 1.4, and PTT and fibrinogen were normal. Erythrocyte sedimentation rate was 111 mm/hr and C-reactive protein was 10.1

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mg/dL. Blood cultures were sent. Differential considerations for his hematemesis were NSAID-induced ulcers or a Mallory Weiss tear, although these did not explain the fever or elevated inflammatory markers. Overnight, he received acid suppression, but hematemesis continued and he received two units of packed red blood cells. Esophagogastroduodenoscopy was performed and showed the stomach filled with blood, but after extensive lavage, no source of bleeding was identified. The mucosa of the esophagus, stomach, and duodenum appeared normal. Upon removal of the endoscope, he vomited blood and went into hypotensive shock with a blood pressure of 50/30. The massive transfusion protocol was activated. Repeat endoscopy again did not identify a bleeding source; a video was obtained while withdrawing the endoscope (viewable at <http://links.lww.com/MPG/A315>). A purple mass was briefly seen during withdrawal of the endoscope, but on continued visualization was not seen again and was thought to be a passing blood clot (Figure 1). Otolaryngologic evaluation did not identify a source of bleeding. An angiogram of the celiac, gastroduodenal, left gastric and superior mesenteric arteries was unremarkable. CT angiogram of the abdomen was normal as well.

Angiography was repeated, and when the thoracic aorta was visualized, a coarctation of the aorta just past the subclavian artery was noted. There was a small pseudoaneurysm that had a jet medially, consistent with an AEF (Figure 2). An aortic stent was placed to cover the pseudoaneurysm and fistula. This was unsuccessful and required subsequent aortic graft placement. In post-case review of the endoscopic video, the purple mass in the mid-esophagus was the aneurysm pushing into the lumen (Figure 1, Video [<http://links.lww.com/MPG/A315>]).

Over his initial 48 hours in the hospital, the patient required transfusion of 25 liters of blood products. Blood cultures grew *Streptococcus pneumoniae*. Sequelae of his hospital course included a mild stroke in the left parietal area, which slowed his processing speed slightly. He developed severe gastroparesis, thought to be from vagal nerve transection during aortic graft placement. After several months on parenteral nutrition, he underwent a pyloroplasty, after which he tolerated enteral feeds.

Discussion

In a previously healthy adolescent with no risk factors, AEF is exceedingly rare. Several key features of this case merit further discussion. First, the initial unexplained hypertension was a clue to the coarctation, but due to ongoing hemodynamic instability and recurrent hypovolemic shock, further investigation was not performed. Second, the endoscopic appearance of a pseudoaneurysm had not been previously described; the lesion was not pulsatile, was not bleeding, and was not easily observed. Third, the awareness of the rapid massive bleeding and hypotension should have led to earlier consideration of an arterial source of bleeding. When radiologic visualization of the abdominal aortic vessels did not identify a cause, imaging the thoracic vessels should have been considered. A bleeding scan was also not considered, and in a brisk bleed of this etiology, it may have been effective. Fourth, survival depends on massive cardiovascular support, rapid diagnosis, and availability of cardiothoracic intervention. Implementation of a massive transfusion protocol with the use of the Belmont Rapid Infuser (Belmont Instrument Corporation, Billerica, MA)

to rapidly administer large volumes of blood products was lifesaving. A massive transfusion protocol was essential to deliver sufficient blood products for survival; the use of crystalloid has been shown to worsen outcomes in the setting of massive blood loss.⁸ Following intubation with ongoing bleeding, placement of an esophageal compression tube could have been used to slow the bleeding in our opinion. Despite a delay in diagnosis of approximately 27 hours after arrival to our hospital, the patient survived with minimal neurologic injury.

In summary, this patient had an undiagnosed congenital aortic coarctation, which created turbulence leading to the development of a pseudoaneurysm. This area of disrupted blood flow became superinfected with *Streptococcus pneumoniae* causing back pain and fever. The infected aneurysm then eroded into the esophagus creating an AEF. AEF has a high mortality; of the two previously reported cases from aortic coarctation, one survived and one died.^{3,4} The pediatric gastroenterologist should consider AEF in context of massive hematemesis. Recognition of the endoscopic findings of AEF, as reported in this case, may be essential for patient survival.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Figure 1.

Review of the endoscopic video (<http://links.lww.com/MPG/A315>) showed a non-pulsatile purple mass protrude briefly into the esophagus, likely the aneurysm. The left image was distal to the mass in the esophagus. The middle image shows the beginning of the mass. The image on the right shows the purple mass in the mid-esophagus.

Figure 2.

An image taken from the angiography of the thoracic vessels. The red arrow identifies the coarctation; the yellow arrow marks the site of extravasation.