Primary Aorto-Esophageal Fistula: A Rare Cause Of Gastrointestinal Hemorrhage

A Tak, R Klizek, J Campion

Citation

Abstract
Aorto-esophageal fistula is a rare but often fatal entity causing upper gastrointestinal bleeding. Although this entity was first reported in 1818, only a few successful treated cases have been published. The diagnosis is rarely made before death. We describe a case of aorto-esophageal fistula in a 76-year old white female who presented with hematemesis and was found to have a primary aorto-esophageal fistula due to thoracic aortic aneurysm. To our knowledge, this is a very rare case and only very few such cases have been reported in literature. The occurrence of primary aorto-esophageal fistula remains predominantly a post mortem diagnosis.

BACKGROUND
In 1818, Debreuil, a French naval surgeon, reported a case of primary aorto-esophageal fistula occurring in a 28 year old soldier after ingesting a beef bone fragment that died after exsanguinating hemorrhage. (1) Sir Ashley Cooper described a typical case of this entity in 1829. (2) Primary aorto-esophageal fistula is a rare entity with an incidence, in necropsy series, of 0.04-0.07 %. Aorto-esophageal fistulas may be primary or secondary. (3) The majority of these fistulas are secondary and occur in a setting of prior aortic reconstructive procedures with prosthetic material mostly from the duodenum downwards. Pathology of aorta represents 80% of all cases of primary aorto-esophageal fistulas. The etiology of primary aorto-esophageal fistula is uncertain but proposed theories include direct wear and inflammatory destruction triggered by infection, foreign bodies, and erosion. (4) Despite a characteristic presentation with almost invariably fatal course, primary aorto-esophageal fistula is overlooked and is barely mentioned in major gastroenterology texts.

CASE REPORT
A 76 year old white female with history of hypertension, COPD, hypothyroidism and osteoporosis was admitted with an acute spell of weakness, dizziness and light headedness accompanied by a single episode of hematemesis. There was no history of peptic ulcer disease, GERD, or use of NSAIDS. Past history was negative for sexually transmitted diseases, including syphilis. Many months before, she had tried weekly Alendronate a few times. This gave her heartburn, and therefore Alendronate was changed to Evista. Other medications included Norvasc, Synthroid, and vitamin and calcium supplements. Physical examination was significant for tachycardia and guaiac positive stools, and nasogastric aspirate revealed coffee ground blood. There were no clinical features of acute vasculitis. The initial hemoglobin level was 8.7 with hematocrit of 25.5. RPR and vasculitis workup was not ordered. The endoscopy on the day of admission revealed an adherent clot at the GE junction which could not be dislodged after repeated washings. Although mild esophagitis was present, there were no esophageal varices and duodenum and stomach appeared normal. The patient was given blood transfusions and her hemoglobin remained stable. Her hospital course was complicated by cellulites of her left arm which required IV antibiotics and prolonged her inpatient stay. However, on the eighth day of admission, she had a massive episode of hematemesis, became hypotensive, and required multiple blood transfusions and was stabilized. Repeat endoscopy revealed stomach full of fresh blood with normal distal esophagus and minimal esophagitis. At the 26 cm mark from the opening of the mouth, there was a large clot with well circumscribed, sharply margined mucosal defect with pulsatile extrinsic compression presumed to be an ulcer or a fistula. Stat CT scan revealed a focal descending thoracic aortic aneurysm leaking into the azygo-esophageal recess, dissecting into adventitial and sub-mucosal layers of esophagus. The aneurysm was located at the level of carina.
The patient was transferred to a center with higher level of care with fatal outcome.

DISCUSSION

Primary aorto-esophageal fistula is an uncommon but life threatening cause of gastrointestinal bleeding, which often causes death by massive exsanguination. Transient self limited bleed called “herald bleed” may precede fatal exsanguination and is often a significant feature of disease process. This may sometimes allow an opportunity for diagnosis and appropriate management. Diagnostic workup may include CT scan, upper GI endoscopy, RBC scan and angiography but no single modality has good sensitivity and specificity for definitive diagnoses. A pulsating sub mucosal mass is typical and blood clots may obscure the site of fistulation, making diagnosis challenging. All authors agree that treatment of primary aorto-esophageal fistula is surgical. Despite surgical treatment, the mortality rates are very high. The diagnosis is mostly based on high clinical suspicion, supplemented by judicious use of endoscopy and appropriate investigations. Timely thoracic surgery consultation may result in a favorable outcome. We believe that suspicion of aorto-esophageal fistula should be considered in elderly hypertensive patients with sudden unexplained hematemesis.

References

Author Information

Abdul Momin Tak, M.D.
Department of Internal Medicine, Brooks Memorial Hospital

Ronald D. Klizek, M.D.
Department of Radiology, WCA Hospital

James P. Campion, M.D.
Department of Gastroenterology, WCA Hospital