

Massive Hemorrhage Due to Aorto-esophageal Fistula in an 11-Months-Old Infant: A Case Report

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ABSTRACT

Aorto-esophageal Fistula (AEF) with no prior history of cardiac disease or trauma is an uncommon but a mortal cause of Upper Gastro Intestinal Bleeding (UGIB) in children.

In pediatric patients, AEF is mainly due to a congenital cardiac or vascular anomaly or foreign body ingestion. There are surgical, endoscopic, and interventional radiological treatment options; however, definitive treatment is surgical intervention. Because of the rapid and devastating course, diagnosis and treatment decision should be made quickly.

In this article, we report a case of an 11-months-old infant who presented with hematemesis due to a primary aorto-esophageal fistula. The patient was transported to the operating theater for an emergency endoscopy. He suffered massive hematemesis in the operating room and an emergency laparotomy was performed but no source of bleeding was detected. An undiagnosed AEF was found and repaired during a left thoracotomy. Despite massive volume resuscitation, the patient passed away four hours later in the PICU. The etiology of the fistula remains unknown.

Aorto-esophageal fistula (AEF) is an extremely infrequent problem that is mostly fatal. This issue has not been listed among the common causes of gastrointestinal bleeding in children [1] and is mainly attributed to accidental ingestion of foreign bodies; particularly disc batteries or after cardiothoracic surgery [2]. In recent years the ingestion of disc batteries especially lithium batteries in toys is a growing concern with catastrophic outcome with a high mortality rate of 2% in this group of patients [3]. The causes of UGIB in children are age-related. The most common causes in toddler age include bleeding of varicose veins, esophageal laceration, Mallory e Weiss syndrome, and gastritis [4-5]. In pediatric patients, AEF is mainly due to foreign body ingestion or a congenital cardiac or vascular anomaly [1]. Another cause of UGIB with unknown

etiology, reported in a 2-year old child, was primary AEF which was repaired with cardiopulmonary bypass [6].

In adults, thoracic aortic aneurysm is the most common cause of AEF which can be life-threatening, other causes are esophageal malignancy, foreign body ingestion, trauma (iatrogenic in most cases), postoperative complication and very rare aortitis due to tuberculosis [7-8].

The patient's hemodynamic status, determines whether an investigation should be done or not. In cases that are diagnosed or under suspect, treatment decisions should be made quickly because of the high mortality risk. Options can be surgical, endoscopic, or interventional radiological procedures [9].

In this article, we report a rare case of primary AEF in an 11-months-old infant with no underlying cause that resulted to death due to massive GI hemorrhage.

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Case Report

An 11 months-old male infant was taken to emergency service by his mother due to bloody sputum and hematemesis. He had a history of coughing and sputum for the past three months and no history of illness or medication usage after birth. His mother reports three times low-volume bloody vomiting four hours before admission to the hospital.

On physical examination, he was pale and ill, with no abdominal guarding or tenderness. Vital sign: blood pressure 65/pulse mmHg, pulse rate 130/min, RR=30/min, no hematochezia was observed. There was no history of chronic liver disease or icterus, trauma, foreign body, or corrosive ingestion.

His laboratory tests except that a hemoglobin level of 5.9 g/dl, were unremarkable. The patient was hospitalized, oral intake was stopped, and nasogastric tube inserted. Gastric lavage with 150 ml of normal saline yielded 500cc blood-stained fluid. Transfusion of packed red blood cells plus FFP was started. Consultation with pediatric gastroenterologist and surgeon was made and patient was transported to pediatric ICU. Upper GI endoscopy was delayed until hemodynamic stabilization and resuscitation. Two hours later, GI bleeding was stopped, with suspicious to gastric ulcer, pantoprazole and octreotide was started and he was transported to the operating room for urgent endoscopy.

Unfortunately, the child began to vomit bright red blood and the patient's condition was deteriorated, he had diffuse crackle in auscultation of the both lungs. Vital sign: PR= 155/min, BP=50mmHg/pulse and RR = 28/min, resuscitation with blood, fluids and inotrope drugs was intensified.

With the presumption of gastro duodenal source of bleeding, laparotomy was suggested and rapid sequence anesthesia was induced with ketamine, midazolam and succinyl choline and the patient was intubated by cuffed endotracheal tube.

In explorative laparotomy, the stomach was distended with some clots and fresh bright blood, but no source of bleeding could be visualized in the stomach or duodenum, meanwhile a steady flow of bright red blood from the esophagus could be seen. With the suspicion of esophageal bleeding, a retrograde endoscopy was performed but no lesion, clot, hematoma or inflammation could be found, nonetheless a delicate streak of blood was seen.

During the procedure, the patient sustained ventricular tachycardia resulted in cardiac arrest within minutes. Resuscitation was started with chest compression, epinephrine, CaCl₂, albumin and other resuscitative drugs. These measures resulted in return of central pulses and measurable end-tidal carbon dioxide within 10 minutes. Operation continued by surgery team consisting of pediatric, vascular and cardiothoracic surgeons. With presumption of AEF, approach to the aorta via left lateral thoracotomy was performed, no blood in the thoracic cavity was seen; however after exploration of the thorax

and great vessels, a 3-mm AEF was identified at 1 cm distal to the origin of the left subclavian artery. The aortic wall was repaired with interrupted monofilament (5-0 prolene) sutures without cardiopulmonary bypass. No granulation tissue or inflammation was observed around the fistula; the esophageal defect was closed with continuous vicryl sutures. The thorax was closed and after repairing of stomach and performing gastro-jejunostomy; the patient was transferred to PICU. Although the infusion of vasopressors, resuscitative drugs and blood products (packed RBC, FFP and platelet) were continued during surgery and after transmitting to PICU; the infant experienced severe metabolic acidosis (PH=6.7) and cardiogenic shock postoperatively; followed by cardiac arrest and unfortunately passed away, 4 hours after surgery.

Discussion

In this article, we report a previously healthy infant with no history of liver disease or ingestion of foreign bodies which presented with bloody sputum and episode of hematemesis. Primarily due to insignificant amount of bleeding, AEF was not part of our differential diagnosis; however, after massive GI bleeding, the possibility of a bleeding with vascular origin was more obvious.

Massive UGIB causing hemodynamic instability is rare in children; suggesting the possibility of variceal bleeding, peptic ulcer disease or AEF. Albeit the primary AEF is very rare and always almost fatal in children [6], it should be considered in the absence of any history of foreign body ingestion, surgery or trauma.

There are a few case reports that have suggested various other causes for AEF such as: continuous nasogastric tube insertion, vascular malformation and esophagitis due to tuberculosis [10].

A thorough examination and history taking should always be performed in any child with a massive UGIB to perceive the foreign body ingestion (disk battery of toys), liver disease, trauma and former cardio-esophageal surgery [1].

Some diagnostic modalities including esophagoscopy, real-time computed tomography with intravenous contrast, arteriography, and upper gastrointestinal contrast imaging have been suggested to diagnosis AEF [6,8]. Although early diagnosis leads to a better outcome, primary AEF is frequently misdiagnosed. Even though esophagogastroduodenoscopy (EGD) is the most sensitive and specific modality for diagnosis of AEF (9), in some clinical studies, EGD fails to detect most cases and high clinical suspicion is required [11]. The sensitivity of EGD to identify the source of bleeding in other causes of UGIB is 90%, but for AEF it is less than 10 % [12-13]. In many studies this failure of diagnosis was due to active pulsatile bleeding or a lesion covered by a clot or massive bleeding [14-18].

The hemodynamic stability of the patient is a major factor for following proper investigation, so we could not proceed a far examination in our case, which was unstable. Insertion of a Sengstaken-Blakemore tube is one of the options discussed in some studies to control the bleeding with esophageal origin [1], unfortunately due to the patient's condition, we did not have the opportunity to perform an endoscopy to locate the bleeding source and perform this procedure.

Several types of treatment have been clarified in the literature for AEF. The thoracic aortic wall defect could be managed with extra-anatomic bypass or in situ reconstruction using grafts. The esophageal defect could be repaired with direct suture or subtotal esophageal resection followed by gastro-esophageal reconstruction immediately or later [9]. Urgent resection of the esophagus and reconstructing the gastrointestinal continuity as an elective case after the inflammatory processes cleared up; was the choice option in few studies [7-11]. Consistent to some of the mentioned studies, in our case; rapid surgical treatment was performed and the very small defect in aortic wall (3mm) and esophageal defect were closed in situ. Treatments such as endoscopic heater coagulation probe and application of hemovascular clip are suggested in the hemodynamically stable patients [16-17]. Akashi et al, concluded that aortic endovascular interventions alone were not a definitive method, and open surgical interventions such as esophagectomy and repair with omentum, aortic replacement through prosthesis or homograft, were more effective and increased the patients survival too [19].

In our case, the autopsy was not performed because the consent was not allowed by his father. This is the limitation of the case report.

Conclusion

AEF as a primary cause of upper GI bleeding is rare in pediatrics and infants, but based on the high rate of mortality; clinicians should have a high index of suspicion in any patient presenting with hematemesis with or without a prior history of foreign body ingestion or even though negative endoscopic findings.

In conclusion, early diagnosis and aggressive surgical approach without delay and elimination of the source of bleeding is the only option for effective treatment. The management of these patients should be performed with a multidisciplinary approach involving radiologists, pediatric gastroenterologist, pediatric surgeon and cardiothoracic surgeon to obtain the most favorable results in the management of the patients with AEF.

We should be alert about the need to massive volume transfusion and patient resuscitation.

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Patient consent: "Consent to publish the case report was not obtained because this report does not contain any personal information that could lead to the identification of the patient."

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