Successful Management of Exsanguinating Hematemesis in a Child with an Undiagnosed Vascular Ring, Esophageal Foreign Body and Aortoesophageal Fistula: A Case Report


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Introduction:

Aortoesophageal fistula (AEF) in the pediatric population is rare and life-threatening. To our knowledge, documented cases of AEF are associated with either congenital vascular ring or esophageal foreign body (EFB) independently. We report a case of vascular ring presenting as exsanguinating hemorrhage in the setting of AEF following foreign body ingestion.

Case Report:

A 6 year old 17 kg male presented to our satellite emergency department in hypovolemic shock secondary to exsanguinating hematemesis. He was hypotensive (BP 46/35), tachycardic (HR 168) and anemic (Hgb 5.4). Chest x-ray showed EFB. He was emergently transferred directly to the operating room on our main campus for surgical management of presumed AEF from battery ingestion.

Upon arrival, RSI was performed with ketamine and rocuronium. A right radial arterial line was placed. Of note, left upper extremity pulses were not palpable. Flexible bronchoscopy revealed normal mucosa, thrombi from presumed aspiration and distal tracheal compression. Intraoperative angiogram yielded no obvious signs of bleeding near the foreign body.

Patient developed recurrent massive hematemesis in the operating room and became acutely unstable. Emergent sternotomy was performed revealing abnormal aortic branching suspicious for right aortic arch as well as dense inflammatory phegmon, however, AEF was not visualized.

Given EFB location and patient’s aortic arch anatomy, a left thoracotomy was performed, with confirmation of a vascular ring consisting of right aortic arch, aberrant left subclavian artery and Kommerell’s diverticulum. The vascular ring was divided and esophagus transected revealing pulsatile blood along its posterior wall, corresponding to an area likely on the anterior aspect of the distal right aortic arch within the phegmon.

Consensus was made that the AEF would be best addressed via median sternotomy with the use of cardiopulmonary bypass, selective cerebral perfusion and circulatory arrest. Esophagoscopy was performed once supine, but only brisk bleeding was visualized requiring esophageal balloon tamponade. A heavily corroded quarter was retrieved and AEF was repaired by autologous pericardial patch aortoplasty. The esophagus was left in discontinuity and a gastrostomy tube was placed. The EBL was 4L. The patient received 12U of pRBCs, 6U of FFP, 5U of platelets and 4U of cryoprecipitate.

He was transferred to the ICU and extubated on postoperative day 6. Postoperative course was complicated by dehiscence of the thoracotomy incision, pleuro-esophageal fistula, mediastinitis and lung abscess. He was discharged home and returned to the operating room 4 months later for successful primary esophageal reconstruction.

Discussion:

EFB ingestion is a common problem in the pediatric population. Fortunately, the majority of cases are acutely identified and easily retrieved. A minority can go on unidentified for weeks to years. Chronically retained EFB can result in pressure necrosis of the esophageal wall causing abscess formation, perforation, mediastinitis, and AEF.

In our case, the combination of congenital vascular ring anomaly and a chronically retained EFB resulted in the development of an AEF. Although vascular ring anomalies usually present with aero-digestive tract symptoms (stridor, paroxysmal cough, dysphagia, and cyanotic episodes), cases presenting with retained EFB have been described. In two such cases, retention of the ingested foreign objects resulted in local esophageal erosion, but not AEF formation. Iatrogenic foreign bodies in the form of NG tubes have been reported to cause AEF in individuals with aortic arch anomalies. To our knowledge, this is the first case of AEF resulting from a chronically retained EFB in the setting of an undiagnosed aortic arch anomaly.

This case represents a successful team approach utilizing experienced pediatric cardiac anesthesiologists, cardiothoracic surgeons, pediatric surgeons, cardiologists, otolaryngologists, radiologists, and emergency medicine physicians to prevent what usually results in mortality. We attribute this patient’s survival to prompt aggressive volume resuscitation, intraoperative multidisciplinary diagnostic approach and immediate surgical management.

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