When undiagnosed coarctation leads to massive upper GI bleed

Michael Krieves MD1, Chris Nichols MD1, Mark Thompson MD2, Rich Ing MD1, Glenn Merritt MD1
Children’s Hospital of Colorado1, Covenant Children’s Hospital2

Introduction:
Coarctation of the aorta is one of the most common cardiac defects with an estimated incidence of 1 in 2,500 births (1). Most cases are diagnosed in infancy, however patients may present years later with severe complications including heart failure, aortic rupture, endocarditis, and intracranial hemorrhage.

Case Report:
15yo healthy male presented after 2 episodes of hematemesis. He reported two-week period of nausea, non-bloody emesis, weight loss, and back and shoulder pain.

Urgent evolving investigation included upper GI endoscopy, ENT consultation, and gastric arteriogram, all non-diagnostic. During this 8-hour period there were 3 episodes of massive hematemesis. After the third episode, a thoracic angiogram revealed a thoracic coarctation and aortoesophageal fistula. Endovascular stents were placed. After a fourth hemorrhagic episode later that evening, the patient underwent emergent thoracotomy where a mycotic aneurysm was resected, along with the coarctation. The esophageal fistula was isolated from the aortic repair with latisimus dorsi flap. The patient was resuscitated with 14 L of PRBC’s, 7 L of FFP, 3 L of platelets, and 221 ml of cryoprecipitate between arrival and thoracotomy (2 days).

A week later, suspicion for mediastinal infection prompted cervical esophagostomy, stapled closure of GE junction, and Stamm gastrostomy. He recovered after a 3-month course of antibiotics and returned home with persistent gastroparesis, but neurologically intact, and with good function.

Discussion:
Aortoesophageal fistula (AEF) was first reported in 1818 and Chiari’s triad of midthoracic back pain, sentinel arterial hemorrhage, hematemesis and finally exsanguination after a symptom free period was first described in 1914 (5). The diagnosis of AEF is rarely made before death and before 1983 no one was reported to survive surgery(3). Undiagnosed coarctation of the aorta leading to mycotic aortic aneurysm is quite rare and in the case of rupture usually lethal (2,3). Although currently rare, mycotic aneurysm secondary to aortic enteritis was responsible for approximately 20% of deaths in the presurgical era (3).

In this patient the initial work up unsuccessfully evaluated for typical upper GI sources of bleeding and aortoesophageal fistula and coarctation of the aorta were low on the differential diagnosis (4). Coarctation may have been considered earlier in this patient’s course had comparative blood pressure measurement been performed in the lower extremity. During the arteriogram, when no bleeding source was identified performing an aortogram to look for more proximal sources of bleeding may have helped identify the source.

Conclusion:
This case illustrates the importance of early detection and treatment of coarctation of the aorta, as the late consequences of undiagnosed coarctation have a high morbidity and mortality.

References:
Kenny, Coarctation of the aorta: From fetal life to adulthood. Cardiology Journal 2011
Wilkins, Diagnosis and Management of Upper Gastrointestinal Bleeding. Ann Fam Physician 2012