Dieulafoy's Lesion: A Rare Cause of Gastrointestinal Bleeding after Interrupted Aortic Arch Repair in an Adult

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Abstract

Survival into adulthood in patients with an interrupted aortic arch (IAA) is exceedingly rare. A recent literature review found 25 reported cases of IAA in adults. We describe the first case of prolonged, occult, postoperative gastrointestinal bleeding as a major complication following IAA surgery. We discuss the management of a 51-year-old man who underwent repair of an IAA, aortic valve replacement, and replacement of the ascending aorta for a known aortic aneurysm.

Keywords

- interrupted aortic arch
- gastrointestinal
- bicuspid aortic valve

Introduction

Interrupted aortic arch (IAA) is a neonatal surgical emergency representing 1% of all congenital cardiovascular diseases. Without correction, mortality approaches 90% in infants.[1] Patients survive into adulthood by developing an extensive arterial collateral circulation, enabling perfusion of the distal aorta beyond the interruption. Type A IAA is more common in adults. In infants, the classical presentation is with severe congestive heart failure. In adults, the symptoms can vary significantly from no symptoms to refractory hypertension, malaise, and claudication. Life-threatening complications have been reported at 13%, to include coronary artery disease, biventricular failure, and subarachnoid hemorrhage. Patients with Type B and C IAA are more likely to have central nervous system symptoms thought to be secondary to vertebral steal phenomenon.[2]

Case Presentation

We report a case of significant and prolonged gastrointestinal (GI) hemorrhage in a 51-year-old man as a challenging and initially treatment-resistant complication of an IAA repair.

The patient initially presented with persistent abdominal discomfort, iron deficiency anemia, and one episode of per rectal (PR) bleeding; both upper and lower GI tract endoscopies revealed mild gastritis and diverticulosis. He also experienced angina on exertion and dizzy spells. A computed tomography (CT) of the chest, abdomen, and pelvis demonstrated an incidental finding of a Type A IAA and aneurysm of the aortic root and ascending aorta (4.4 cm) (►**Fig. 1**). The patient denied symptoms of claudication or postprandial discomfort. No cause was identified for anemia. Medical history included severe hypertension (170/90) refractory to antihypertensive treatment and type 2 diabetes. Echocardiography identified severe bicuspid aortic stenosis (peak

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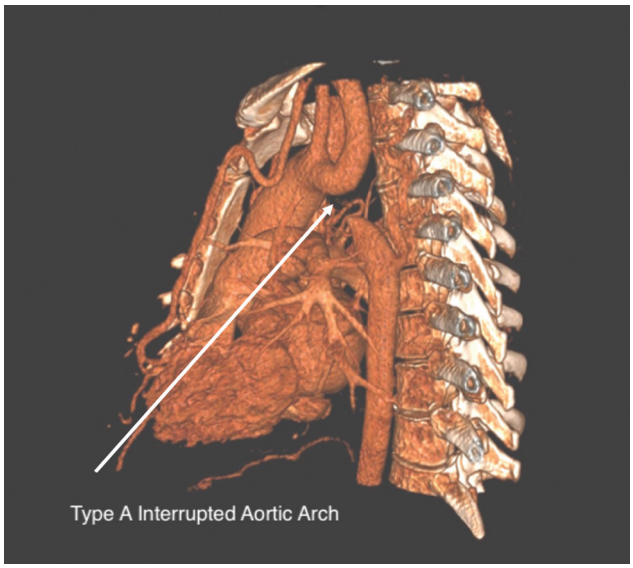

Fig. 1 Preoperative computed tomography reconstruction of Type A interrupted aortic arch.

gradient 58 mm Hg, mean 38, valve area 1 cm^2), and a small restrictive, perimembranous ventricular septal defect (VSD). Coronary angiogram showed no significant coronary lesions. CT angiography demonstrated numerous collateral vessels in the chest wall; the internal thoracic arteries measured over 1 cm in diameter. Following multidisciplinary discussion (MDT), the patient was scheduled for surgery.

The vascular team exposed the abdominal aorta proximally to the celiac axis. A 14-mm Dacron graft (Vascutek-Terumo) was anastomosed end-to-side and tunneled through the diaphragm into the pericardial cavity (►**Fig. 2**). Following aortic cross-clamping and cardioplegia delivery the aortic root was approached. The aortic valve (Sievers type I R-N) was heavily calcified. The VSD was identified and closed. A modified Bentall procedure with a 23-mm CarboMedics valved-conduit (Sulzer-Carbomedics) was completed. Following ascending aorta replacement, the

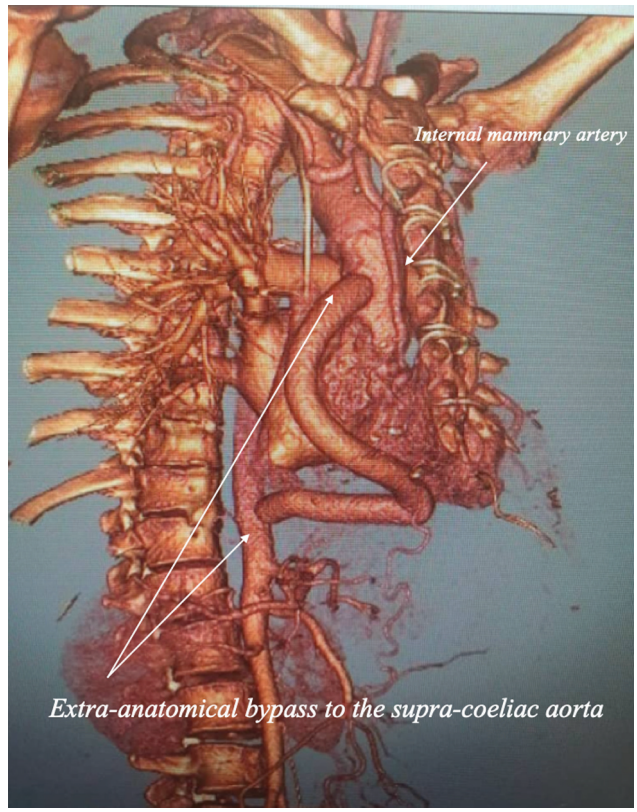

Fig. 3 Reconstructions showing extra-anatomical bypass to the supra-celiac aorta. Note also the enlarged internal mammary artery.

14-mm supraceliac graft was placed to lie along the right side of the heart and anastomosed end-to-side to the ascending aortic graft (►**Figs. 2A, B** and **3**). Cardiopulmonary bypass and cross-clamp times were 214 and 188 minutes, respectively, and the patient was cooled to 30°C.

On day 3 postoperatively, the patient developed a mild derangement of hepatic function, an international normalized ratio (INR) > 8, and melena. Warfarin was withheld and a lower dose heparinization commenced when the INR had normalized

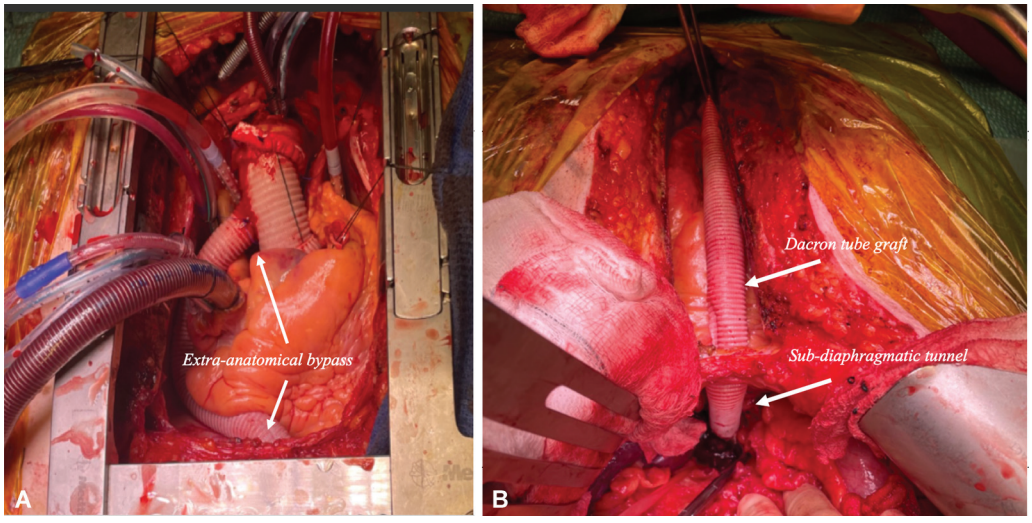

Fig. 2 (**A**) Intraoperative Image showing extra-anatomical bypass. (**B**) Dacron tube graft tunneled through the diaphragm into the pericardial cavity.

(activated partial thromboplastin time ratio target: 1.5–2). The melaena persisted, however, and all anticoagulation was withheld until the bleeding significantly reduced.

Extensive radiological and invasive GI investigations proceeded to help identify the source of GI bleeding. Over a 10-week period, 60 units of packed red cells were transfused. Multimodality assessment of the GI tract with endoscopies (including gastroscopy, colonoscopy, enteroscopy, and capsule endoscopy), and focused abdominal CT angiograms, failed to identify any source of bleeding. Following one enteroscopy, two arteriovenous malformations were identified in the proximal and mid-jejunum. Although not actively bleeding at the time, they were preventatively treated with argon plasma coagulation and clips. The patient continued to bleed and an exploratory laparotomy with repeat enteroscopy was performed. On this occasion, the small bowel could not be assessed in its entirety and interventional vascular radiology proceeded with angiography and mesenteric embolization, which proved futile. Following further MDT discussion, the decision was made to repeat a laparotomy and assess the entire small bowel endoscopically through an enterotomy. On this occasion, an angiodysplastic lesion in the form of an ileal Dieulafoy was identified and clipped. An ileal stoma was created to facilitate future direct endoscopic intubation of the small bowel. Consequently, bleeding with requirement for blood transfusions reduced significantly, but not entirely, as seen on further enteroscopic assessments via the ileostomy. To accomplish reduced splanchnic blood flow, octreotide (100 µg subcutaneously twice daily) was commenced. After 10 weeks, the patient was discharged home on octreotide and did not experience any further melaena. The patient is scheduled for elective reversal of the ileostomy.

Discussion

IAA is associated with other cardiac abnormalities: 90% of individuals will have a VSD and a third of patients will have an associated bicuspid aortic valve (BAV).[3] Repair of IAA, BAV with severe Aortic stenosis (AS), VSD, and aortic root aneurysm in an adult has been documented only once previously to our knowledge.[4] A review from Jiang et al propose that adults with asymptomatic IAA and hypertension could be managed nonoperatively with antihypertensive therapy and kept under close review.[1] In this case example, however, the patient had symptomatic refractory hypertension in addition to aortic stenosis and a dilated aortic root aneurysm.

Persistent, occult, GI bleeding was a challenging and unexpected postoperative complication despite utilization of highly sensitive endoscopic as well as angiographic imaging modalities. These diagnostic methods are normally successful at identifying the challenging 5% of upper GI bleeds.[5] The patient received multiple blood transfusions and invasive GI investigations, and he had a prolonged postoperative stay, which impacted his mental health.

The GI bleeding had significantly improved following Dieulafoy's clipping; however, it did not cease completely until octreotide treatment had commenced. Dieulafoy's lesions are a rare and specific type of angiodysplasia where a large, tortuous, submucosal end artery penetrates through the mucosa over time.[6] Mortality rates for patients with Dieulafoy's lesions can be as high as 80%. We speculate that the persistent postoperative bleeding could be due the splanchnic hypertension and impaired autonomic autoregulation within the abdominal aorta and its branches following the newly performed extra-anatomical bypass, exacerbated by the requirement for anticoagulation for mechanical aortic valve replacement.

We present lessons learned from protracted GI bleeding following a complex composite aortic root and ascending aortic replacement with extra-anatomical bypass to the supraceliac abdominal aorta.

Conflict of Interest
None declared.

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