Operative challenges in management of concurrent interrupted aortic arch and descending thoracic aortic aneurysm

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Interrupted aortic arch is a rare finding in the adult patient. This condition in combination with a descending thoracic aortic aneurysm is an even more exceptional occurrence. Surgical management includes open, endovascular, and hybrid options. We present the case of a 57-year-old man with interrupted aortic arch and concomitant descending thoracic aortic aneurysm, review characterization of this entity, and discuss management options with consideration to associated risks. (J Vasc Surg 2013;57:1661-3.)

Interrupted aortic arch (IAA) is a rare vascular malformation constituting approximately 1.5% of congenital cardiovascular anomalies and is often associated with other cardiac anomalies.1 Excluding this report, only one case of IAA presenting with a descending thoracic aortic aneurysm (DTAA) has been reported in the literature to date.2 The resulting physiological and structural alterations of each separate condition pose challenges during open and endovascular repair when addressed simultaneously. This case report reviews characteristics of IAA, describes the successful management of IAA with DTAA in an adult, and unveils risks associated with various surgical approaches.

CASE REPORT

A 57-year-old man presented to his primary physician with respiratory complaints including shortness of breath and wheezing. His medical history included hypertension and no prior surgery. Members of his family have had cardiac procedures as infants; however, details were not known. Significant findings on physical examination included absent distal lower extremity pulses and weak femoral pulses bilaterally without a history of claudication. Chest X-ray demonstrated a widened mediastinum and bilateral rib notching. Evaluation with computed tomography angiography revealed a 7.8- × 7.7- × 8.2-cm DTAA and no discernible continuation of the aorta beyond the left subclavian artery (SCA) (Fig 1). The aneurysm appeared just beyond this interruption and exerted mass effect on adjacent structures including those of the left pulmonary hilum, the esophagus, and the left atrium. An extensive network of collateral vessels reconstituted the descending thoracic aorta. The aortic root was mildly dilated at 4.3 cm. An echocardiogram showed a bicuspid aortic valve with mild insufficiency as well as an ejection fraction of 45% to 55% with left ventricular hypertrophy and impaired relaxation without evidence of intracardiac defects.

Because of the size of the aneurysm, evidence of ventricular remodeling in the face of uncontrolled hypertension, and mild aortic insufficiency with a bicuspid aortic valve, the decision was made to surgically address both the aneurysm and coarctation. The abnormal aorta was incorrectly thought to represent progressive coarctation amenable to endovascular recanalization and repair using a thoracic endograft. We anticipated left SCA coverage and performed a left common carotid-subclavian bypass. The initial aortogram via left brachial access revealed complete occlusion of the aorta with a blind cul-de-sac beyond the left SCA, and this was confirmed via right brachial access. An aortogram obtained via femoral access showed a blind ending of the descending thoracic aorta (Fig 2). Despite several attempts from each access site, we were unable to pass a wire through the area of aortic discontinuity and abandoned this approach.

After discussion with the patient, we pursued an open surgical approach via a left posterolateral thoracotomy. We encountered massive collateral vessels on entering the chest. Intraoperatively, we concluded the diagnosis of congenital thoracotomy. We encountered massive collateral vessels entering the chest. Intraoperatively, we concluded the diagnosis of congenital IAA (type A) due to absence of the aorta (Fig 3). Because of the extensive calcifications, we were unable to clamp the aorta proximally and ultimately performed an extra-anatomic bypass from the 20-mm left SCA in an end-to-side fashion to the descending aorta end-to-end using a 20-mm Dacron graft with resection of the aneurysm. On opening the aneurysm sac, we encountered significant bleeding from the collateral vessels, necessitating emergent femoral-femoral cardiopulmonary bypass and massive transfusion of blood products. The patient stabilized following these measures and the repair was completed. At 3 months of follow-up, shortness of breath resolved and antihypertensive medication was discontinued.

DISCUSSION

Steidele first described IAA in 1778,3 and Samson and co-workers performed the first successful surgical
correction in 1957. The classification of IAA is based on the site of aortic interruption, with the majority of cases occurring between the left common carotid and left subclavian arteries (referred to as type B). Interestingly, 22q11 deletions have been seen in 50% to 80% of cases of this type of interruption. Our case involved a type A, or interruption distal to the left SCA, which accounts for 79% of cases in adults. Type C interruption occurs proximal to the carotid artery and constitutes 4% of IAA cases.

Accurate diagnosis of congenital IAA in the adult may be challenging, as it is difficult to distinguish from a progressively narrowed coarctation resulting in an acquired IAA. This has been proposed as a reason for the predominance of type A interruptions in adults. Diagnosis is suggested by imaging modalities (echocardiography, computed tomography angiography, magnetic resonance imaging) in a patient with hypertension and differential between upper and lower extremity pulse exams. Ascending and descending thoracic aortic angiograms showing a blind cul-de-sac confirm the diagnosis.

Refractory hypertension, claudication, heart failure, and cerebrovascular accident are the most common presentations in the adult and, generally, indications for surgical repair. Multiple options exist for treatment including open anatomic reconstruction and extra-anatomic bypass; the latter is the technique most commonly employed and has the advantage of avoiding numerous enlarged collaterals. Endovascular reconstruction with or without septal perforation and subsequent dilation is a third possibility for repair, although these cases generally involve acquired IAA resulting from severe progressive coarctation.

Our case posed a unique challenge in dealing with IAA and DTAA simultaneously. The optimal treatment for this exceedingly rare occurrence is unknown and case specific. We initially opted for an endovascular solution and assumed both would be amenable to treatment with an endograft for dilation and exclusion, respectively. We planned for several contingencies. Coverage of the left SCA would be necessary and a carotid-subclavian bypass was performed. Additionally, we thought support with stent grafts and bare metal stents and possible serial dilations would possibly be necessary for adequate expansion. However, this approach would avoid disturbing the massive collateral circulation seen on preoperative imaging.
We were surprised to discover a complete occlusion and were thwarted in our attempts to gain wire access across the occlusion. A stiff-ended wire, Brockenbrough transeptal needle, and radiofrequency probe are the instruments used for perforation of aortic occlusions at the level of the aortic isthmus, with subsequent balloon dilation or stent placement, that have been reported in the literature. As suggested by Joseph et al, a spectrum from coarctation to IAA exists and performing perforation of an aortic occlusion beyond focal aortic atresia could result in catastrophic consequences.

Open surgical bypass overcomes the endovascular challenges presented; however, it introduces a different set of difficulties. Proximal arch clamping was not permitted because of the calcification and as an alternative, the left SCA was used for an extra-anatomic bypass. The added task of aortic aneurysm exclusion necessitated direct incision into the aneurysm. This action led to significant hemorrhage from collaterals whose collective cross-sectional area was equal to that of an aorta with normal diameter. Preparation for this included exposing the femoral vessels for cardiopulmonary bypass, employing rapid autotransfusion, and using occlusive balloon catheters for large branches. Despite our preemptive measures, control of blood loss became a formidable task. In the future, we would consider performing the aneurysm repair using deep hypothermic circulatory arrest.

CONCLUSIONS

The presence of IAA and DTAA is an exceedingly rare combination of aortic anomalies, and simultaneous treatment poses a unique surgical challenge. Endovascular therapy may have a role but must be judiciously employed in the case of IAA when perforation of an occluded segment is being considered. An open approach may be necessary, and collateral bleeding should not be underestimated, with consideration of repair under hypothermic circulatory arrest.

REFERENCES
