Case report

Double aortic arch in an adult undergoing coronary bypass surgery: a therapeutic dilemma?

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Abstract

Double aortic arch is a congenital anomaly that is rarely found in adults. Typically, this is treated in infancy secondary to symptoms. We describe the treatment of an asymptomatic adult male that was diagnosed with double aortic arch during cardiac catheterization.

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1. Introduction

Double aortic arch (DAA) is a rare congenital anomaly of the aortic arch system where a complete vascular ring is formed around the trachea and esophagus. Usually, this causes significant symptoms and patients are diagnosed and treated early in life. However, sporadic cases [1—5] have been reported in adults. We describe the treatment of an asymptomatic adult who was found to have DAA during his workup for coronary artery disease.

2. Clinical summary

A 63-year-old male with a past medical history of hypertension and diabetes mellitus presented with unstable angina and exertional dyspnea. He denied shortness of breath at rest and difficulty swallowing. Cardiac catheterization revealed three-vessel coronary artery disease, preserved left ventricular function and DAA (Fig. 1). Chest computed tomography confirmed DAA, with comparably sized anterior and posterior limbs. Esophagram demonstrated moderate extrinsic compression of the proximal one-third of the esophagus (Fig. 2). Bronchoscopy was not performed due to the lack of significant respiratory symptoms.

Following median sternotomy, cardiopulmonary bypass was initiated via the right femoral artery and right atrium. The anterior arch was divided proximal to the origin of the left carotid artery. Mediastinal fat was placed between the two ends of the divided arch and the trachea. Coronary bypass grafting proceeded without incident. The patient was discharged on day six with no neurologic sequelae.

3. Discussion

DAA is a congenital anomaly in which the right dorsal aorta, present during fetal development, fails to regress. It can present in multiple anatomical variations, depending on the size of the arches and the presence or absence of an atretic segment. DAA typically presents in infancy with symptoms of stridor, dyspnea, or poor feeding. The severity of symptoms depends on the severity of compression of the structures (trachea, esophagus) within the vascular ring. However, when compression is minimal, DAA can go undiagnosed into adulthood [1—4].

Surgical intervention with division of the minor arch is indicated for those adult patients who are symptomatic and can be approached via either left posterolateral thoracotomy or median sternotomy [5]. However, a therapeutic dilemma exists when asymptomatic patients undergo surgery for another cardiac condition. To date, there are only a handful of case reports that describe asymptomatic DAA diagnosed in the 6th and 7th decades of life [1—4]. All four patients were treated conservatively, even in one undergoing coronary bypass surgery [1]. Nevertheless, due to the sporadic nature of this anomaly, there are no good inferences on the long-term prognosis of untreated DAA in this age group. Long-term sequelae of untreated DAA in patients undergoing cardiac
surgery is unknown and certainly re-operation for DAA should be a significant concern.

Although our patient did not have any definitive symptoms at the time of presentation, some component of his exertional dyspnea may have been attributable to the vascular ring (tracheal compression with high cardiac output). More importantly, we elected to proceed with division of the vascular ring at the time of coronary revascularization because of the moderate esophageal compression documented with barium swallow. We elected to divide the anterior arch, as this was easiest through the sternotomy approach, given the two arches were equivalent in size. Cardiopulmonary bypass was utilized to control arterial blood pressure during manipulation, division and ligation of the anterior arch. Potential operative pitfalls of repairing DAA include hemorrhage and inadvertent damage or division of adjacent structures such as the trachea, recurrent laryngeal nerve, or esophagus (if dividing the posterior arch).

In conclusion, we present this case of an asymptomatic DAA in an adult diagnosed during workup for coronary artery disease. The standard of care is unclear due to the rarity of this finding in adults. Recent reports document non-operative management [2–4] even in a patient undergoing coronary bypass surgery [1] with good results. Most probably, had we elected not to proceed with division of the DAA, the patient would have recovered uneventfully. Furthermore, without evidence of esophageal compression on barium swallow, we would not have elected to proceed with division of the DAA. Nevertheless, we feel that concerns about the development of symptoms in the future requiring subsequent surgical intervention are valid and therefore proceeded with DAA division at the time of coronary revascularization. The addition of a vascular ring procedure to an already planned cardiac operation should add minimal operative mortality and therefore we support concomitant surgical intervention.

References