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Single Stage Extra Anatomic Bypass of an Adult Interrupted Aortic Arch with Aortic Regurgitation

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ABSTRACT

BACKGROUND

Adult patients with interruption of the aorta are an extremely rare occurrence. It is commonly associated with additional cardiovascular anomalies. Surgery is the mainstay in the management of the patients. Interrupted aortic arch and associated cardiac disease is complex situation that requires correction either simultaneously or in stages.

CASE

A 32-year-old male planned for aortic valve replacement, incidentally diagnosed with interrupted aortic arch Type A. He was managed with single stage aortic valve replacement and extra anatomical bypass.

DISCUSSION

Various management options of adult interrupted aortic arch and associated anomalies are discussed.

KEYWORDS

Interrupted aortic arch; Bicuspid aortic valve; Aortic regurgitation; Aortic valve replacement; Single stage repair; Extra-anatomical repair

INTRODUCTION

Interrupted aortic arch, first described in 1778 by Steidele and classified by Celoria and Patton in 1959 is a rare congenital malformation reported in 3 per million live births in which there is loss of luminal continuity between the ascending and descending portions of the aorta. It is commonly associated with other intracardiac malformation like bicuspid aortic valve, left ventricular outflow tract obstruction, ventricular septal defect, patent ductus arteriosus and so on [1]. As per our knowledge only around 35 cases are reported in adults till today making it an extremely rare occurrence [2].

CASE REPORT

A 32-year-young male patient presented to other hospital with dyspnea and palpitation since 4 years. He was diagnosed to have rheumatic heart disease, severe aortic

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regurgitation and planned for aortic valve replacement. On midline sternotomy, the dilated bilateral internal thoracic arteries with tortuous extensive collaterals in the retrosternal area seen. On aortic cannulation the increased pressure gradient between the ascending aorta and femoral artery was noticed and hence coarcatation of aorta suspected. The planned procedure was abandoned and computed tomography- aortogram done was suggestive of Type A interrupted aortic arch with tortuous extensive collaterals. He was referred to our centre for further management.

On examination, blood pressure in the right arm was 192/80 mmHg. Echocardiography done at our centre was suggestive of the bicuspid aortic valve, severe aortic regurgitation, mild aortic stenosis (Peak/Mean Gradient - 33/17 mm Hg) and mildly dilated proximal ascending aorta (36 mm). Aortic annulus of 31 mm. Global left ventricular hypokinesia, mild mitral regurgitation, mild tricuspid regurgitation, mild pulmonary arterial hypertension, concentric left ventricular hypertrophy, dilated left atrium and ventricle. Left ventricular ejection fraction 40% - 45%, Left ventricular systolic dysfunction. Diastolic spill seen in descending aorta suggestive of coarcatation of aorta.

A single-stage operation was performed. It consisted of a standard median sternotomy, aortic and right atrial cannulations, and establishment of extracorporeal circulation. After the establishment of cardiopulmonary bypass and aortic cross clamp, under cold blood cardioplegia the entire heart was retracted upward and right out of the pericardial sac, so the retrocardiac descending aorta could be easily seen using trans posterior pericardial approach. The aorta was then side clamped, an adequate aortotomy done. An 18-mm polytetrafluoroethylene graft was anastomosed end-to-side with 4-0 prolene continuous suture. The graft was clamped, and kept in pericardium. Aortic valve was replaced with a 23 mm SJM mechanical valve using pledgeted intermittent sutures. Aortotomy was closed with full precautions after de airing, and rewarming was begun. On releasing Aortic cross clamp, cardiac activity resumed.

Proximal anastomosis of graft to ascending aorta was done by side-biting clamp in end-to-side fashion, with a running 4-0 polypropylene suture. The patient was weaned from Cardiopulmonary bypass smoothly and the decannulation done.

The Cardiopulmonary bypass time was 320 minutes, Aortic cross clamp time 171 minutes. Patient was cooled to 26 degree centigrade. Ventilatory support was needed for 13 hours, Intensive care unit stay for 3 days.

Postoperative Echocardiography showed prosthetic valve in situ with peak and mean gradients of 19 mmHg and 11 mmHg. No valvular or paravalvular leak. Left ventricular ejection fraction 35%.

There were no perioperative complications. The patient recovered uneventfully and discharged on postoperative day 6.

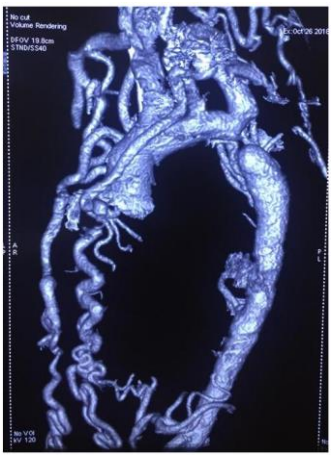

Figure 1: Preoperative, contrast-enhanced, 3-dimensional, computed tomography aortogram reveals interruption of the aortic arch beyond left subclavian artery (interrupted aortic arch, type A). Extensive collateral vessels to descending aorta are evident from the subclavian and intercostal arteries.

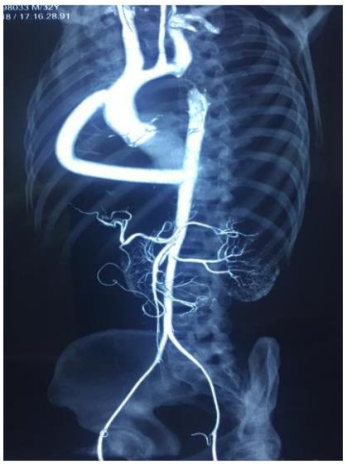

Figure 2: Postoperative, contrast-enhanced, 2-dimensional, computed tomography maximum intensity projection images shows patent flow in the 18 mm PTFE graft anastomosed end-to-side to the mid-portion of the ascending aorta and to the distal portion of the descending thoracic aorta. There is no luminal narrowing or filling defect. The collaterals disappeared within a month postoperatively.

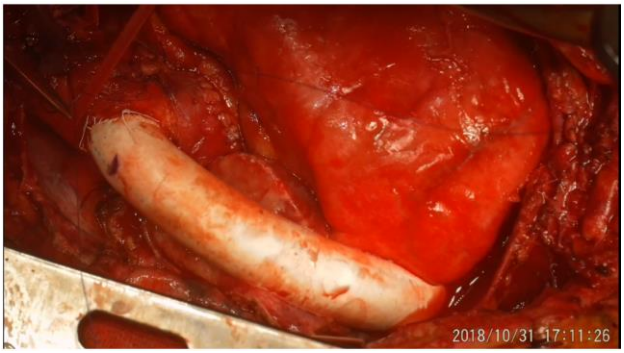

Figure 3: Intraoperative photograph showing the proximal and distal end-to-side anastomosis of the 18 mm graft to ascending and descending aorta respectively.

Computed tomography-contrast aortogram postoperatively showed well homogenously opacified patent graft with no obvious filling defect or luminal narrowing. Collaterals disappeared (Figure 1 - Figure 3).

DISCUSSION

Interrupted aortic arch commonly manifests in infants, as congestive heart failure. In rare cases, its presentation in adults ranges from asymptomatic or incidental finding in association with other cardiac anomalies [3,4]. The obligatory extensive collateral circulation maintain distal flow. Most reported cases in adults have been repaired in single stage using an extra-anatomic approach [5]. Single or two stage repair has been debatable. However single-stage repair is increasing in infants [6].

Operating on the coexistent cardiac lesions without correcting the aortic interruption can result in significant under perfusion of distal organs, severe pressure load on left ventricle because of hypertension and congestive heart failure. Hence simultaneous repair of both interruption and associated cardiac lesion is preferred. Single stage repair with combined thoracotomy and sternotomy increases postoperative pain and complications. Thus only median sternotomy is preferable.

The extra anatomic ascending to descending aorta graft can be routed along left or right margin of the heart and also in front of or behind inferior vena cava [7,8]. The latter avoids damage to higher vessels and phrenic nerve. The antecaval approach avoids the compression of the pulmonary veins and inferior vena cava. The retrocaval method makes bleeding control difficult but easily dissected during reoperation.

CONCLUSION

The interruption of aorta with concomitant cardiac lesions in adults can be safely and efficaciously be repaired in the single-stage procedure using median sternotomy.

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