Case report

Left cervical aortic arch associated with pseudocoarctation and aortic and mitral regurgitation: one-stage surgical repair

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Abstract

Cervical aortic arch is rare with a prevalence of less than 1/10 000 live births [1]; but may occur with other cardiovascular congenital abnormalities. We report pseudocoarctation of a left-sided cervical aortic arch with an aberrant right subclavian artery and aortic and mitral regurgitation, corrected by an anterior mediastinal shunt from ascending to descending aorta and aortic and mitral valve replacements, as a one-stage procedure. © 2001 Elsevier Science B.V. All rights reserved.

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1. Case history

Cervical aortic arch is rare with a prevalence of less than 1/10 000 live births [1]; but may occur with other cardiovascular congenital abnormalities. Our patient presented in childhood, when an ejection systolic murmur was detected at the base of the heart, radiating to the back, with systolic murmurs over both clavicles. Blood pressure measurement was not possible because the upper limb pulses were palpable and the femoral pulses were weak. At cardiac catheterisation the pressure in the ascending aorta was 237/120 mmHg. An exploratory left lateral thoracotomy was performed at the age of 18 years, with the intention of repairing the suspected aortic coarctation. This revealed calcification of the right subclavian artery, the origins of the carotid arteries and the descending aorta to the level of T5. The operating surgeon felt that the only possible operation was a long graft from the ascending aorta around the heart and hilum of the lung, to the descending thoracic aorta. It was decided then not to carry out the procedure in view of the uncertainty of the fate of such a graft and so the chest was closed.

She was lost to cardiological review; but represented at age 28 when transthoracic echocardiography revealed mild mitral and aortic regurgitation. Empirical treatment for hypertension with metoprolol was commenced.

In 1993, at the age of 36, a chest X-ray showed left atrial enlargement and the aortic arch high in the left side of the neck. A CT scan showed the aortic arch at the level of T2 and marked narrowing of the arch at this point. A magnetic resonance (MR) scan showed that the arch was left-sided, markedly ectatic and that the top was high in the base of the neck (Fig. 1a). The aortic root was moderately dilated at 26 mm and the highest point of the arch was narrowed to 10 mm. The right subclavian artery arose from the descending aorta, distal to the left subclavian artery origin.

Transthoracic echocardiography and cardiac catheterisation showed moderately severe aortic regurgitation, severe mitral regurgitation and a dilated left atrium. Arch angiography in 1998 showed that the great vessels consisted of (in order): the right and left common carotid arteries (proximal to the narrowing at the highest point of the arch), and the left and right subclavian arteries distal to the narrowing (Fig. 1b). The systolic pressure difference across the aortic narrowing (systolic pressure proximal to the narrowing minus the systolic pressure distal to the narrowing), was 100 mmHg.

In October 1998 surgery was performed through a median sternotomy. Bypass was commenced with cannulation of both the ascending aorta and femoral artery. After mobilization of the heart the left ventricle was noted to be very enlarged. In order to reach the descending aorta the heart was arrested, the ventricle displaced and the posterior peri-
cardium opened. An 18 mm Dacron graft was sewn to the descending aorta over a side-clamp, through a left-sided window in the posterior pericardium. The graft was directed to the diaphragmatic surface of the heart, around the right side of the right atrium anterior to the inferior vena cava (IVC) and anastamosed to the right side of the ascending aorta over a side-clamp (Fig. 2a,b) The aortic and mitral valves were replaced with mechanical prostheses whilst preserving the mitral subvalvar apparatus. The patient is well and symptom-free 2 years later.

2. Discussion

Cervical aortic arch and aberrant origin of the right subclavian artery arise secondary to abnormal involution or persistence of embryonic arches [2,3]. Cervical aortic arch may be associated with coarctation [4] and true coarctation usually occurs just above or below the ductus arteriosus. Pseudocoarctation is characterized by deformity and narrowing between the left subclavian artery and the ductus, with varying degrees of proximal dilatation and tortuosity [5].

In our case true coarctation was not present; rather than

Fig. 1. (a) Saggital plane T1-weighted spin echo MR scan showing dilated left atrium and the cervical aortic arch. (b) Line drawing illustrating the anatomy seen on the arch aortogram (saggital oblique view).

Fig. 2. (a) Composite image from two adjacent saggital post-operative MR slices. T1-weighted spin echo images show the graft arising from the ascending aorta, passing around the inferior surface of the heart, and anastamosis with the descending aorta just above the diaphragm. (b) Line drawing of MR scan in (a).
obstructive disease in the aortic wall, the MR scan showed marked ectasia of the aortic arch and its position in the neck caused kinking at its highest point, thereby causing a functional obstruction. A gradient of 100 mmHg was demonstrated, in contrast to a previous report in which small gradients were recorded [6].

Coarctation of the aorta was first surgically corrected in 1945 [7], with complete resection and end-to-end anastomosis in children. When coarctation presents or re-presents in adulthood, however, the direct approach to repair may present difficulties because of significant pulmonary disease (such as obstructive airway disease), a large collateral circulation or dense scarring and friability from previous repair.

In patients who have correction of coarctation in adulthood, the unfavourable haemodynamic effects of coexisting disease (particularly coronary artery or aortic valve disease) and the morbidity of a second operation for its correction, may be quite significant. It is therefore now not unusual for a one-stage procedure to be performed with ascending to descending aortic bypass graft in the posterior mediastinum: aortic valve replacement, coronary artery bypass grafting and cardiac transplant have all been successfully performed in this context [8].

In our case the graft was not taken around the left side of the heart as there was severe left ventricular hypertrophy with severe adhesions in the left chest from the previous thoracotomy. As the heart lay, the left-sided route would have been adverse and therefore difficult to allow the graft to lie well. As it lies, the graft will not irritate the coronary artery as it lies low on the right side of the right atrium. Furthermore, a reasonably large graft was used and so the possibility of constriction of the IVC was avoided by taking it anterior rather than posterior to the IVC.

It was elected to replace rather than repair the mitral valve because of the appearance of mixed pathology: there were features of congenital disease (a cleft posterior leaflet); but also secondary degenerative features (significant subcordal fusion reminiscent of a rheumatic process). Therefore a repair would have been complex and so the possibility of needing to revise this was avoided by valve replacement.

To our knowledge, there are no previous reports of pseudocoarctation associated with severe aortic and mitral regurgitation managed by ascending to descending aortic bypass through the anterior pericardium with aortic and mitral valve replacements, as a one-stage procedure.

References