VIEWPOINT

Congenital coronary artery dilatation

S Chakrabarti, E Thomas, J G C Wright, J J Vettukattil

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Although dilated coronary arteries are uncommon in children, three cases are presented which, even more unusually, were not associated with Kawasaki's disease

Dilated coronary arteries are uncommon as a clinical problem in the paediatric population. When found, they are usually associated with the sequelae of Kawasaki disease.¹ We present a series of three children who have dilated coronary arteries, not associated with Kawasaki disease. There was no history suggestive of a possible previous Kawasaki disease in any patient. All of the children had bicuspid aortic valves. On angiography significant dilatation of the origins of the affected vessels were seen, followed by a conical and smooth tapering over the next 30 mm segments. The anatomy of the other segments of the coronary vessels was essentially within normal limits.

CASE 1

A newborn infant with antenatal diagnosis of dysplastic aortic valve and aortic regurgitation had evidence of a bicuspid aortic valve, aortic stenosis, and dilated coronary arteries on echocardiography. An angiogram at 9 years of age (fig 1) confirmed the dilatation of the coronary arteries. The child has been asymptomatic to date, with mild aortic stenosis and regurgitation.

CASE 2

A newborn infant with antenatal diagnosis of small left ventricle had additional diagnoses of a perimembranous ventricular septal defect (VSD), subaortic stenosis, bicuspid aortic valve, and persistent ductus arteriosus (PDA) in the postnatal

period. Ectasia of coronary artery origins were also noted in the postnatal scans. After undergoing resection of the subaortic stenosis and closure of the VSD and PDA, he is haemodynamically fine with mild residual subaortic stenosis and aortic regurgitation. The angiogram (fig 2) shows the ectasia of the coronary arteries.

CASE 3

A newborn infant was diagnosed as having coarctation of the aorta along with bicuspid aortic valve and subaortic stenosis. The baby had a subclavian flap repair. Over years of follow up evolving aortic stenosis and regurgitation were observed. Coronary dilatation was also suspected and confirmed on angiography (fig 3). There was no history of a possible Kawasaki-like illness during the entire period in follow up.

DISCUSSION

Coronary ectasia has been defined as distension of a part of the artery which is more than 150% of the diameter of an adjacent unaffected segment.² Some authors have suggested that involvement of the whole vessel should be termed "ectasia", while partial involvement is better called "aneurysm".³ Apart from association with Kawasaki disease and coronary fistulae, the aetiology of coronary dilatation in the paediatric age group is not very clear. It has been associated with coronary atherosclerosis, vasculitides⁴ (polyarteritis nodosa and Takayasu's disease), trauma,⁵ and hyperlipidaemia⁶ in the adult population.

Abbreviations: PDA, persistent ductus arteriosus; VEGF, vascular endothelial growth factor; VSD, ventricular septal defect

See end of article for authors' affiliations

Correspondence to:
Dr Joseph J Vettukattil,
Wessex Cardiac Unit,
Tremona Road,
Southampton SO16 7YD,
UK; joseph.vettukattil@
suht.swest.nhs.uk

Figure 1 Angiogram of case 1.

Figure 2 Angiogram of case 2.

Figure 3 Angiogram of case 3.

Interestingly, only the right coronary artery is involved in all our cases. All of the lesions were conical with a wide opening at the aortic end and smoothly tapering to the normal direction of the distal segment of the coronary artery.

Dilated segments are thought to change the rheology of blood in the affected vessel. Sluggish or turbulent blood flow may then predispose to myocardial ischaemia and its sequelae, including sudden death.⁷ The risk of ischaemic events is increased, regardless of the status of coexisting coronary lesions.⁸

It is unclear why patients develop dilatation of the coronary arteries. It has been postulated that vascular endothelial growth factor (VEGF), which increases in response to an inflammatory state,⁹ may play a role in the pathogenesis of coronary artery lesions in Kawasaki disease. This hypothesis would not be tenable in our series. It is possible that inherent weakness of the aortic wall predisposes to the pathogenesis of these lesions. Bicuspid aortic valve has been associated with serious abnormalities of the ascending aorta, including aortic dilatation, aneurysm, dissection, and coarctation. A common underlying developmental defect of the aortic valve and the ascending aorta has also been suggested.¹⁰

We postulate that an eccentric jet of blood from the bicuspid aortic valve into the coronary ostia leads to the initiation and then progression of these lesions. None of these patients had coexistent aortic root dilatation to explain the extension of the

pathology into the coronary origins. As our patients are young, it is likely that they may have the substrate for aortic dilatation, and accelerated juxta-ostial dilatation is still a possible pathologic process. Even an undiagnosed Kawasaki event in the past is unlikely to be a plausible explanation, with the compelling association of the bicuspid aortic valves in all the three patients. Moreover, in two out of our three patients, the lesion was detected soon after birth.

The natural history of these lesions is not fully known. It is very likely that these patients have an increased risk of ischaemic coronary disease. On the other hand, bicuspid aortic valve is one of the most common congenital heart anomalies in the general population. Such coronary dilatations, though rare, may actually cause significant morbidity and mortality by the virtue of the number of patients having bicuspid aortic valves. We suggest that patients with bicuspid aortic valves (especially those with an eccentric jet) need to be on periodic follow up for early detection and management of these lesions.

Authors' affiliations

S Chakrabarti, E Thomas, J J Vettukattil, Southampton University Hospital, Southampton, UK
J G C Wright, Birmingham Children's Hospital, Birmingham, UK

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