An unusual presentation of right coronary artery fistula

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SUMMARY A four year old girl with infective endocarditis had unexplained facial swelling. Cross sectional echocardiography showed that a large right coronary artery fistula to the right atrium was obstructing the distal superior vena cava. The diagnosis was confirmed by cardiac catheterisation and at operation. The child was symptom free one year after operation.

Coronary artery fistula is a rare congenital heart lesion which is diagnosed in most instances in young patients who present with atypical murmur. The definitive diagnosis relies on cardiac catheterisation although several cases diagnosed by cross sectional echocardiography have been reported recently.

The haemodynamic complications of a coronary artery fistula are usually related to an important left to right shunt, ischaemic heart disease (caused by coronary artery steal), or accelerated atherosclerosis (in the older patient). Infective endocarditis, thrombosis, and rupture of the fistula may occur. In rare instances the fistula may obstruct one of the cardiac vessels or cavities. We report a rare case of a right coronary artery fistula that caused obstruction of the superior vena cava.

Case report

A four year old girl with no previous cardiac symptoms was referred with fever, lethargy, arthralgia, and episodes of facial swelling that had started six months before admission to hospital. On examination she was well developed, blood pressure was 90/50 mm Hg, the pulse was normal with a rate of 100 beats per minute, and her face was swollen. There was finger clubbing. A diffuse cardiac apex was felt at the sixth intercostal space in the anterior axillary line. Heart sounds were normal. A 2/6 continuous murmur was heard over the right sternal border and a 3/6 early diastolic murmur was detected over the aortic valve area. The liver and spleen were enlarged (6 and 10 cm respectively below costal margin). The biochemical and haematological laboratory results were within normal limits; the erythrocyte sedimentation rate was 35 mm in the first hour. The electrocardiogram and chest x ray were normal. The clinical diagnosis of infective endocarditis with aortic valve regurgitation was supported by finding viridans streptococci in multiple blood cultures.

Cross sectional echocardiography (Diasonic CV 400, 3.5 MHz transducer) showed a 5 mm vegetation on the right coronary cusp and Doppler echocardiography showed mild aortic regurgitation. The unexpected finding of a very dilated proximal right coronary artery (14 mm in diameter) raised the suspicion of a coronary artery pathology. The origin of the left coronary artery from the aorta was well visualised. From the subcostal short and long axis views a large right sinus of Valsalva was seen and a dilated proximal right coronary artery could be followed. It started from a normally located ostium and ran conventionally in the right anterior atrioventricular groove. At Doppler examination turbulent flow was found above the aortic valve in the right sinus of Valsalva. From the second third of the right coronary artery, just distal to the origin of the acute marginal branch, a fistula was seen running posteriorly toward the superoposterior portion of the interatrial septum (sinus venosus area) and ending over the right aspect of the interatrial septum as a distal spherical aneurysm (fig 1).

Doppler examination around the aneurysm showed high velocity turbulent flow but the exact location of the communication with the right atrium could not be visualised. The entrance of the superior vena cava into the right atrium was very narrow (2

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mm) because it was compressed by the dilated fistula. There was no sign of right ventricular volume or pressure overload.

Cardiac catheterisation was undertaken to confirm the anatomical diagnosis. Special attention was paid to the distribution of the branches of the right coronary artery (fig 2). No left to right shunt was detected by measurements of oxygen saturation; none the less, contrast injection in the proximal right coronary artery showed mild opacification of the right atrium, ventricle, and pulmonary artery. The pathway of the fistula was confirmed by selective injection into the right coronary artery. Small marginal branches were visualised as they branched from the dilated right coronary artery. The superior vena cava could not be entered from the right atrium. A digital subtraction angiography was then performed by injecting 4 ml of contrast material in the right external jugular vein. A near total obstruction of the distal superior vena cava was shown; a very small amount of contrast material directly entered the right atrium from the superior vena cava while most of the contrast material drained into a dilatedazygos vein towards anastomoses with the inferior vena cava.

Operation confirmed the anatomical diagnosis and the patient had ligation of the fistula and patch augmentation of the distal superior vena cava. She was symptom free a year after operation.

Discussion

A congenital coronary artery fistula is a rare anomaly. It was found in 1 out of 50,000 children with congenital heart disease. Several groups have shown that cross sectional echocardiography shows the anatomy of the fistula, its pathway, and its correlation to adjacent structures. Moreover, the haemodynamic effects of the fistula can be studied by Doppler echocardiography, which can detect a fistula-to-cavity shunt and estimate the shunt ratio. In this child the detailed pathway of the fistula, its connection to adjacent cardiac structures, and a restrictive left to right shunt were shown. In fact, the main reason for undertaking cardiac catheterisation was to ascertain the branching of the right coronary artery—a feature that was not shown by cross sectional echocardiography. Infective endocarditis is not often associated with coronary artery fistulas, but there are several published reports of such cases. The dilatation of the right sinus of Valsava with the turbulent flow across the aortic valve that we saw in our patient might have enabled bacterial colonisation over the right coronary cusp. The compression of the superior vena cava by the aneurysm in the right atrium in our patient is unusual and to our knowledge has not been described before.

This case shows that a coronary artery fistula may present obstruction of the superior vena cava. Although the fistula and its pathway were easily diagnosed by cross sectional echocardiography, car-
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