Coarctation of the aorta, hypertension and associated features

Introduction

Coarctation of the aorta represents between 5 and 8% of all congenital heart disease. While usually discovered in infancy, it may present in asymptomatic young adults found to be hypertensive. Major complications include endocarditis, aortic dissection, intracranial haemorrhage, heart failure and recognized associated heart defects.

Discussion

Aortic coarctation that is an isolated congenital defect and with discrete architecture will usually be managed with angioplasty and stenting. More complex lesions require surgical excision and repair. In up to 85% of cases of coarctation of the aorta the patient will have a bicuspid aortic valve, and an association with aneurysmal dilatation of the ascending aorta. Less common but associated features of cardiac anatomy include ventricular septal defects, mitral valve abnormalities and an association with Turner's syndrome (Kaemmaren, 2006).

Despite treatment blood pressure is unlikely to return to normal in an individual discovered to have this condition at this age. Lifelong antihypertensive medication is likely to be needed to prevent hypertensive end-organ damage and to protect the aorta. Long-term survival following intervention of any kind tends to be lower than in the general population because of arterial hypertension (Cohen et al, 1989) and associated cardiovascular features.

An associated feature to exclude is intracranial berry aneurysms, frequently located on the circle of Willis – having been reported in up to 5% of patients with aortic coarctation. Computed tomographic cerebral angiography was performed in this case to exclude this, in view of the physically active nature of the patient and the risk of cerebral haemorrhage from aneurysm rupture.

This patient was successfully treated by angioplasty and stenting. She will require annual surveillance and has been advised about antibiotic prophylaxis.

Conclusions

A general physician is unlikely to see a handful of cases of coarctation of the aorta in his/her whole career. Hypertension resulting from coarctation, as from many other causes, may be silent for many years. Early identification can cure the problem and should direct the search for associated cardiac and cerebrovascular complications. BJHM

Case Report

A 17-year-old female athlete who had a black belt in Tai-Kwondo was discovered to be hypertensive (blood pressure recorded as 150/90 mmHg in both arms) when she was screened to join a new gym. Although able to perform physical contact sports to a competitive level without apparent symptoms, direct enquiry revealed that she had calf pain on jogging.

On examination a grade 4/6 systolic murmur was audible throughout the precordium. Femoral pulses were impalpable bilaterally. Routine blood tests including renal function were all normal as was a renal ultrasound and urine analysis for protein and catecholamines. The electrocardiogram showed sinus rhythm, a normal axis and satisfied voltage criteria for left ventricular hypertrophy. A transthoracic echocardiogram was normal, with no evidence of left ventricular hypertrophy and good biventricular systolic function. However, on examination of flow in the abdominal aorta there was continuous forward flow throughout the cardiac cycle.

The chest X-ray (*Figure 1*) showed rib notching as a result of erosive pressure on the ribs from collateral blood flow in large vessels derived from the subclavian, axillary, internal thoracic, scapular and intercostal arteries.

Cardiac magnetic resonance imaging confirmed the diagnosis of coarctation of the aorta (*Figure 2*), explaining the continuous abdominal aortic blood flow. The murmur was the result of turbulent blood flow through an isolated, typical post-ductus location, coarctation of the thoracic aorta. The diameter at its narrowest was 0.6 cm. The femoral pulses were impalpable as the flow at the femoral arteries was reduced and continuous in character (as demonstrated by the echo study) rather than pulsatile and palpable.

Cohen M, Foster V, Steele PM, Driscoll D, McGoon DC (1989) Coarctation of the aorta: long-term follow up and prediction of outcome after surgical correction. *Circulation* **80:** 840–5

Kaemmaren H (2006) Aortic co-arctation and interrupted aortic arch. In: Gatzoulis MA, Webb GD, Daubeney PEF, eds. *Diagnosis and Management of Adult Congenital Heart Disease.* Churchill Livingstone, Philadelphia: 253–62

Figure 1. Chest radiograph showing rib notching (most obvious notches are enclosed by the white rectangles) caused by collateral blood flow.

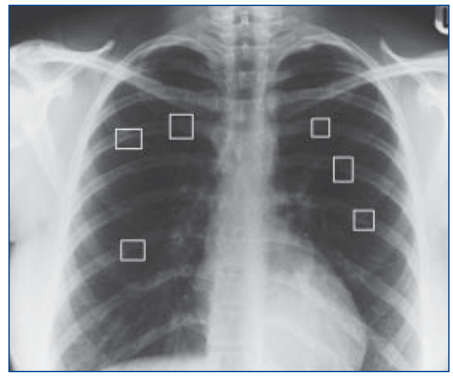
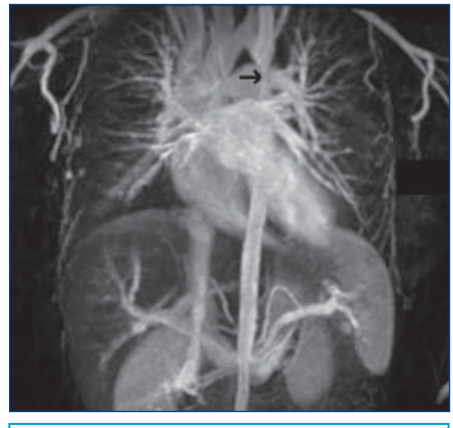


Figure 2. Cardiac magnetic resonance imaging scan performed as part of the investigation of this patient. The black arrow shows the site of the coarctation. They are often shelf-like in anatomy and frequently lie opposite the ductus arteriosus.



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