Self-assessment corner

Stroke in a 24-year-old man

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A previously healthy 24-year-old right-handed man presented with a bitemporal throbbing headache. Within five hours he had developed sudden paraesthesia and weakness affecting his left arm and leg with evidence of a mild left hemiparesis and left hemibody sensory impairment affecting pinprick and light touch.

Cardiovascular examination revealed a heaving apical beat, an aortic ejection click and an ejection systolic murmur, loudest in the praecordium and periscapular region. There was marked radiofemoral delay and weak pulses distal to the femorals bilaterally.

Investigation initially consisted of normal head computed tomography (CT) and electroencephalogram with electrocardiogram showing left ventricular hypertrophy by voltage criteria. His chest X-ray is shown in figure 1.

His mild hemiplegia improved on aspirin within 48 h. He was further investigated with transthoracic and transoesophageal echocardiograms revealing a bicuspid aortic valve. Repeat CT head scan showed a low density non-haemorrhagic infarct in the periventricular white matter on the right. Aortography was performed one month after discharge (figure 2).

Figure 1 Chest X-ray.

Figure 2 Aortogram

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Questions

- 1 What is the most likely diagnosis?
- 2 What does the chest X-ray show and what are the differentials?
- 3 What does the aortogram show and what are the associated complications of the underlying diagnosis?

Answers

QUESTION 1
The diagnosis is coarctation of aorta.

QUESTION 2
The chest X-ray shows rib notching from the third to the ninth ribs and an unfolded aorta. The differentials of rib notching include enlarged intercostal arteries, vein and nerves (box 1).¹

QUESTION 3
The aortogram shows a hypoplastic aorta beyond the left subclavian artery with some antegrade flow across the coarctation. Complications of aortic coarctation include congestive cardiac failure, aortic dissection, infective endocarditis, and cerebral haemorrhage (box 2).

Clinical features

Aortic coarctation is one of the most common congenital heart diseases accounting for about 7% and is seen in 1 in 4000 children. It is found on routine examination in 1 in 10 000 otherwise healthy individuals and is three times as common in men compared to women.

Clinical manifestations depend on the site and extent of the obstruction and the presence of associated cardiac abnormalities such as bicuspid aortic valves occurring in 25 to 70% of cases. These valves may be functionally normal, stenotic or incompetent. Mitral valve abnormalities also occur with an incidence of between 25 and 55%.²

Minor symptoms include headache, epis-taxis and fatigue. Major symptoms are usually secondary to the main complications (box 2). Bacterial endocarditis and cerebrovascular accidents occurred in 2% and 6%, respectively, in one series.³ These complications increase with age and this includes the mor-bidity and mortality associated with hyperten-sion.

Prognosis

Significant symptoms occur in two stages, early infancy and between the ages of 20 and 30

Differential diagnosis of rib notching¹

- enlarged intercostal arteries: abdominal aortic thrombosis, subclavian artery obstruction, Blalock–Taussig operation
- enlarged intercostal veins: superior/inferior vena cava obstruction
- enlarged intercostal nerves: neurofibromatosis

Box 1

Complications of aortic coarctation

- congestive cardiac failure
- aortic dissection
- infective endocarditis
- cerebral haemorrhage

Box 2

years. Most of those surviving early hazards live to reach adulthood but 25% die by 20 years and 75% by 50 years.⁴

Management

Surgical correction of coarctation should preferably be performed at 3–5 years of age, to minimise recurrence of stenosis and post-operative hypertension.⁵ Most patients are hypertensive prior to operation but are nor-motensive within two years. However, on pro-longed follow-up the proportion of normoten-sives declines gradually.⁶ Therefore, surgery is preferable to the poor results of long-term medical care and vigorous treatment of even mild hypertension is an important feature of postoperative care.

Final diagnosis

Aortic coarctation and bicuspid aortic valve with cerebral infarction

Keywords: aortic coarctation, stroke

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