a cumulative effect leading to extrapyramidal manifestations although it is known that the plasma half-life of metoclopramide is short. From our observation and that of others, metoclopramide is sometimes prescribed in the treatment of trivial illness. As dystonic reactions are distressing both to the patients and their parents, greater care should be exercised by doctors when prescribing this drug for children. The dose should not exceed 0.5 mg/kg per day and prolonged use of the drug is not recommended. Extrapyramidal side effects after metoclopramide are rare and early diagnosis will depend on doctors being alert to the possibility so that unnecessary investigations can be avoided. Such reactions can be effectively stopped by administration of benztropine 1 mg intravenously.

We thank our colleagues for permission to study patients under their care.

Q fever endocarditis in a 6-year-old child

ROBERT W A JONES AND DAVID W PITCHER

Department of Paediatrics and Department of Cardiology, Guy's Hospital, London

SUMMARY A 6-year-old boy with a congenital bicuspid aortic valve presented with finger clubbing and hypertrophic osteoarthropathy, and subsequently he developed severe hypertension. The hypertension was successfully treated by nephrectomy, at which a thrombosed mycotic aneurysm of the renal artery was found. Echocardiography showed the presence of aortic valve vegetations. Blood cultures were sterile, but high antibody titres to the phase 1 and 2 antigens of Coxiella burnetii strongly suggested Q fever infection. We believe this is the first reported case of Q fever endocarditis in early childhood.

Case report

A 6-year-old boy from a rural area in Libya presented with a one-month history of polyarthritis and swelling of the fingers and toes. He had been in frequent contact with farm animals. After admission to hospital in Libya he was noted to have a heart murmur. Shortly afterwards he developed severe hypertension complicated by recurrent grand mal seizures. He was treated with a low salt diet and, after the demonstration of only one functioning kidney by intravenous urography, was transferred to Guy's Hospital for further investigation.

Examination showed pronounced finger and toe clubbing. There was also soft tissue swelling of the fingers which felt warm and podgy. A polyarthritis was present affecting the elbows, wrists, knees, and ankles; there was pronounced synovial thickening and local warmth. Systemic fever was absent. Blood pressure was 180/130 mmHg. The pulse was normal. Auscultation showed a loud ejection systolic murmur preceded by an ejection click heard maximally over the aorta. A soft early diastolic murmur was present. The liver and spleen were palpable 6 and 4 cm respectively below the costal margins. No renal bruit was heard. Neurological examination was normal.

Investigations showed: Hb 10.5 g/dl, WBC 10.3 × 10^9/l (normal differential), ESR 78 mm/1st hour, blood cultures (multiple) sterile, immunoglobulins: IgG 17 g/l (normal 8–17), IgA 1.4 g/l (normal 1.5–4.5), IgM 4.6 g/l (normal 0.45–1.45), urine microscopical examination: RBC 0.020–0.300 × 10^9/l. Electrocardiography showed left ventricular hypertrophy, chest x-ray showed slight cardiomegaly (cardiothoracic ratio 1:1.8), and on fluoroscopy aortic valve calcification was seen. X-ray of the limbs

References


Correspondence to Dr K M Goel, Royal Hospital for Sick Children, Yorkhill, Glasgow G3 8SJ.
showed hypertrophic osteoarthopathy affecting the entire length of the radius and ulna, and the lower ends of both tibae. Intravenous urography showed a large but otherwise normal right kidney; there was no evidence of a functioning left kidney. Renal scintillography (technetium $^{99m}$DTPA and DMSA) indicated the presence of a left renal artery, but showed no evidence of left-sided renal function. Renal arteriography showed two renal arteries supplying a normal right kidney; the appearances on the left side are shown in Fig. 1. Peripheral renin activity (Dr M J Dillon, Institute of Child Health, London) was 2660 ng Al/l per hour (normal mean for age 417, range 131–834), plasma aldosterone 990 pmol/l; 35·6 ng/100 ml (normal mean for age 147, range 28–616 pmol/l, 5·3, range 1–22 ng/100 ml), upper right renal vein renin 4592 ng Al/l per hour, left renal vein renin 20 600 ng Al/l per hour. In view of the suspected diagnosis of infective endocarditis and the failure to identify any organisms on repeated blood cultures, antibody titres to C. burneti were measured. Results are shown in the Table. The echocardiographic findings are shown in Fig. 2.

As hypertension was unsatisfactorily controlled by drugs, left nephrectomy was performed. At operation a firm nodule was palpable at the hilum of the left kidney. Subsequent dissection showed that this was a thrombosed mycotic aneurysm of the renal artery (Fig. 1). Histology showed a florid granulomatous arteritis of the aneurysm wall with many large multinucleate histocytes. Three small calcific nodules, probably embolic in origin, were present in glomerular hila. The glomeruli were otherwise normal. No micro-organisms were identified, and immunofluorescent staining for C. burneti was negative. Material from the aneurysm was sterile on bacterial culture, and failed to produce any evidence of Q fever.

Table  
Reciprocal of complement-fixing antibody titres to phase 1 and phase 2 antigens of Coxiella burneti

<table>
<thead>
<tr>
<th>Duration of Illness (weeks)</th>
<th>Phase 1</th>
<th>Phase 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>&lt;8</td>
<td>128</td>
</tr>
<tr>
<td>14</td>
<td>&lt;8</td>
<td>128</td>
</tr>
<tr>
<td>18</td>
<td>128</td>
<td>64</td>
</tr>
<tr>
<td>22</td>
<td>64</td>
<td>16</td>
</tr>
</tbody>
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Fig. 1  Left: left renal arteriogram. Catheter tip lies at the origin of the left renal artery, which is narrow and ends abruptly 1·5 cm from its origin. Distal to the block are seen multiple leashes of collateral vessels. Right: left nephrectomy specimen (viewed from behind) showing the thrombosed mycotic aneurysm of the renal artery situated at the renal hilum (arrowed).
infection on guinea-pig inoculation (Dr A D Evans, Public Health Laboratory, Cardiff).

Postoperatively the blood pressure remained normal without antihypertensives. The phase 1 antibody titre to *C. burnetii* had now risen 4-fold (Table). Co-trimoxazole and doxycycline were started. One month later the polyarthritis had resolved, the clubbing was less pronounced, and the ESR had fallen to 34 mm/1st hour. X-ray of the forearms showed resolution of the hypertrophic osteoarthropathy. Auscultatory and echocardiographic findings were unchanged. Our patient subsequently returned to Libya where (one-year later) he is known to be well. He remains on treatment with antibiotics, but details of his clinical and serological status are not available.

**Discussion**

Chronic Q fever is a rare but increasingly recognised cause of infective endocarditis in adults. Diagnosis is based on the identification of *C. burnetii* in affected valve tissue obtained at operation or necropsy, or on the demonstration of a high antibody titre to the phase 1 antigen of this organism.\(^1\)\(^-\)\(^2\) In the present case, delay in diagnosis resulted from the initial observation of a low phase 1 titre, the high phase 2 titre suggesting acute rather than chronic infection. These findings, and the subsequent 4-fold rise in phase 1 titre, suggest that acquisition of infection was recent, contrasting with the long interval to diagnosis in many adult cases.\(^3\) Early clinical presentation was due to the rapid development of a severe hypertrophic osteoarthropathy, a previously described finding in Q fever endocarditis.\(^5\) A further complication in the present case was the development of renovascular hypertension in association with a thrombosed mycotic aneurysm of the renal artery. Despite the failure to identify rickettsiae in the aneurysm wall, embolism followed by arteritis is the likely pathological explanation, similar lesions of the popliteal and posterior tibial arteries having recently been reported.\(^1\)\(^4\)

While echocardiographic evidence of vegetations is sometimes lacking in patients with proved infective endocarditis,\(^5\) in the present case the echocardiographic appearances of aortic valve vegetations (Fig. 2) confirmed the clinical diagnosis. In a critical transducer position the shaggy echoes seen in diastole appeared to fill the aortic root. Slight alteration of transducer angulation showed that these echoes moved anteriorly during systole, when a normal posterior cusp echo was recorded, suggesting that the vegetations were attached to the anterior cusp. The recording of abnormal echoes from the left ventricular outflow tract could be artefactual, due to scatter of the ultrasound beam, but in other reported cases of infective endocarditis with this appearance operative findings indicated that these echoes arose either from bulky vegetations within the subaortic left ventricular outflow tract\(^6\) or from a flail aortic leaflet prolapsing into this region.\(^7\) In the absence of echocardiographic evidence of aortic cusp destruction the former explanation seems more likely in the present case. Additional echocardiographic findings in our patient included fluttering of the mitral valve in diastole indicating aortic regurgitation.\(^8\) The auscultatory findings suggested a congenitally bicuspid valve. Eccentrically placed diastolic echoes from the aortic valve, as seen in the present case, occur with bicuspid valves but may also be seen with oblique transducer angulation across an aortic valve with three cusps. Abnormal echoes arising from vegetations in our patient made clear confirmation of a bicuspid valve impossible, and this diagnosis remained a predominantly clinical one.

Although previously considered rare in patients
with congenital heart disease, the youngest previously reported case of Q fever endocarditis was in a 15-year-old boy with congenital subaortic stenosis, and 7 adults with involvement of congenital bicuspid aortic valves have also been described. The present case emphasises that chronic Q fever infection must be considered in the differential diagnosis of infective endocarditis in childhood, particularly if there is a history of contact with farm animals and if blood cultures are negative.

We thank Dr G A K Missen and Dr M J Tynan for help and encouragement.

References

Correspondence to Dr R W A Jones, Department of Paediatrics, Guy’s Hospital, London SE1 9RT.

Facial palsy in an infant with coarctation of the aorta and hypertension

P MOORE AND G I FIDDLER

Department of Paediatric Cardiology, Killingbeck Hospital, Leeds

SUMMARY We report the unusual association of facial palsy and severe hypertension in an infant with coarctation of the aorta. The facial palsy resolved before the hypertension was cured.

Case report

A 14-week-old boy was admitted because of failure to thrive and for recoarctation after a left subclavian flap aortoplasty which had been performed for a severe isolated coarctation at age 2 weeks. The femoral pulses which had been present after surgery were absent at 10 weeks, and repeat cardiac catheterisation and angiography at 11 weeks had demonstrated the site of recoarctation and shown that left ventricular function was poor and there was a thickened left ventricular wall. On admission the infant was in mild cardiac failure and his systolic blood pressure was 200 mmHg in the right arm and 80 mmHg in the legs, measured by the flush method. The next day a right lower motor neurone facial palsy was noted (this was particularly obvious when he cried), with facial asymmetry and inability to close the eye. No other neurological abnormality was found and fundoscopy was normal. Despite little success in controlling blood pressure with β-receptor blockade, the facial palsy improved after 5 days and had resolved after 3 weeks. The only other treatment given was methylcellulose eye drops and a pad to close the eye. Six weeks later the coarctation was successfully repaired by inserting a Dacron bypass graft at the site of the previous aortoplasty. The blood pressure fell to 140 mmHg (flush method) in the right arm in 4 days. Three weeks later there was a further fall to 120 mmHg. He remains well and is normotensive 6 months later.

Discussion

Lower motor neurone facial palsy associated with hypertension was first noted by Moxon in 1869. Since then the association has been described in adults and children. An incidence of between 3 and 17% has been reported, although it was not noted in a review of 100 patients. All the reported cases have been associated with hypertension of a renal or idiopathic aetiology and the overall prognosis has been poor. The present case appears to be the first to be caused by hypertension.