Anaesthetic management for nephrectomy in a child with Takayasu’s arteritis and severe renovascular hypertension

A case report

M. A. ROZWADOWSKI, J. W. DOWNING

Summary

The case of a 3-year-old boy with Takayasu’s arteritis and severe renovascular hypertension presenting for nephrectomy is described. Emphasis is laid on control of the hypertension during anaesthesia and careful monitoring of cardiovascular parameters, which may be difficult in the absence of peripheral pulses.


Takayasu’s arteritis or ‘pulseless disease’ is an uncommon condition, which occurs classically in young Asian females, although it has been described in other population groups. Characteristic of the disease is an arteritis of unknown aetiology which can affect the aorta, the proximal portions of its main branches and the pulmonary arteries. Ischaemia and renovascular hypertension follow involvement of the renal arteries.

Among the causes of severe persistent hypertension in childhood, renovascular lesions are second only to renal parenchymal disease, according to Wiggelinkhuizen and Cremin. These same authors found Takayasu’s arteritis to be the single most important cause of renovascular hypertension in non-white children. A few reports of the anaesthetic management of patients with Takayasu’s arteritis exist in the literature but none, so far as can be ascertained, have involved children.

A few reports of the anaesthetic management of patients with Takayasu’s arteritis exist in the literature but none, so far as can be ascertained, have involved children. The anaesthetic management of a child with Takayasu’s arteritis and severe hypertension, refractory to medical treatment and requiring nephrectomy, is described.

Case report

A 3-year-old Zulu boy presented at a rural hospital in a semicconscious state, with generalized convulsions, pyrexia (39°C), a pustular rash and evidence of undernourishment. Physical examination did not help to determine the nature of his illness. A chest radiograph, haematological and electrolyte studies and lumbar puncture were also non-contributory.

Immediate antibiotic (penicillin and chloramphenicol) therapy was started. After a further convulsion the following day, tuberculous meningitis was suspected and antituberculosis treatment was commenced (intramuscular streptomycin 250 mg daily and rifampicin 300 mg, isoniazid 200 mg and ethambutol 300 mg daily by mouth). After 5 days of treatment, the patient’s condition had improved dramatically. However, 4 days later, he suffered a relapse, became mentally dull, had a further convulsion and appeared to be blind. He was then transferred to King Edward VIII Hospital, Durban.

On admission the patient was pyrexial, malnourished, had healed rash marks, was mentally dull but responsive to questions, and blind. Tone, reflexes, and funduscopy were normal. All other systems were reported to be normal. Biochemical, haematological and cerebrospinal fluid investigations were normal. A chest radiograph revealed hilar shadows. A Mantoux test was positive.

At a subsequent examination, the blood pressure was 220/170 mmHg and the left radial and brachial pulses were feeble. Other pulses were present and normal. The findings were confirmed with a Doppler ultrasound probe. A cortical infarct in the occipital region was demonstrated on computed axial tomography. An aortogram and abdominal angiogram revealed obstruction of the left subclavian artery distal to the origin of the left vertebral artery. The right vertebral artery was relatively hypoplastic. There was occlusion of the proximal 2.5 cm of the right renal artery with collaterals originating from the adrenal and ureteric arteries. The left main renal artery was normal, but intrarenal branches showed changes indicative of nephrosclerosis. Intravenous pyelography performed after angiography, showed good dye excretion by the left kidney. Renal perfusion scans (technetium-99m diethylene triamine penta-acetic acid) indicated minimal perfusion in the right kidney and a normal pattern in the left kidney.

After 1 month the boy’s eyesight had improved substantially. Funduscopy still revealed no abnormality. Antituberculosis treatment was continued because of the diagnosis of Takayasu’s arteritis and its known frequent association with tuberculosis.

Hypertension was treated with hydralazine 10 mg 6-hourly, mist. pot. chlor. 5 ml 3 times a day, methyl dopa 250 mg twice a day and sotalol 40 mg daily, which maintained the blood pressure at 170/110 mmHg. Corrective vascular surgery to the right renal artery was contemplated but rejected on technical grounds. Instead it was decided to perform a right nephrectomy, in the hope of removing the cause of the hypertension, namely elevated renin/angiotensin levels secondary to kidney ischaemia caused by the renal arteritis.

Examination at the pre-operative visit showed the patient to be a healthy-looking, active 3-year-old boy, weighing 14 kg. The pulse rate was 110/min and regular, blood pressure, measured in the right arm, was 190/140 mmHg. No pulses were palpable in the left arm, but all other pulses were present and normal. Heart sounds were normal and no murmurs were heard. The rest of the physical examination was normal. There

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was no evidence of the previously described cortical blindness. As the child had not received his antihypertensive medication on the day of the pre-operative visit, dihydralazine and reserpine were given intravenously, lowering the blood pressure to 170/110 mmHg. The results of the pre-operative investigations performed are listed in Table I.

On the day of operation, all routine medication was given orally 4 hours pre-operatively. Trimeprazine 2 mg/kg was administered 3 hours pre-operatively, followed by droperidol 2.5 mg orally 2 hours later. An infusion of 0.01% sodium nitroprusside in 0.9% sodium chloride solution was prepared for use in case sudden marked elevation of blood pressure occurred. On arrival in theatre, the child was asleep. The blood pressure was 110/60 mmHg and pulse 95/min. At the time of investigation, the patient received the following intravenous fluids: Plasmalyte L 100 ml, and whole blood 200 ml, which replaced blood as it was lost.

The course of the anaesthetic of 130 minutes' duration was uneventful (Fig. 1). There was no significant rise in blood pressure after ligation of the right main renal vessels and removal of the right kidney. Intra-operatively, the blood pressure at no stage rose above 110/75 mmHg. Postoperatively the child was observed in the recovery room for 2 hours; the blood pressure fluctuated between 140/90 and 150/90 mmHg, and the pulse rate varied between 90 and 104/min. For analgesia, tilidine hydrochloride 12.5 mg (5 drops) 4-6-hourly was administered with good effect.

**Discussion**

Takayasu's arteritis is an uncommon syndrome, but occasionally patients present for surgery either incidentally, or more frequently for correction of the consequences of vascular occlusive disease. Monitoring cardiovascular status and function is extremely important in these cases and may be complicated if no peripheral pulses are present, particularly in children. In the case described here, only the left arm was pulseless, so haemodynamic monitoring did not present any special problems. For reasons mentioned earlier, direct arterial cannulation was not performed. Similarly, pulmonary artery catheterization was contemplated but was rejected in view of the potential risks exceeding the possible benefits in a small child. The combination of ECG, oesophageal stethoscope, automated oscillotonometer blood pressure monitoring, palpation of the peripheral pulse, 

### TABLE I. PRE-OPERATIVE INVESTIGATIONS (normal range of values in brackets)

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Results</th>
</tr>
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<tbody>
<tr>
<td><strong>Haematological</strong></td>
<td></td>
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<tr>
<td>Haemoglobin</td>
<td>10.7 g/dl</td>
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<tr>
<td>Haematocrit</td>
<td>32.9%</td>
</tr>
<tr>
<td>Platelets</td>
<td>527 x 10^9/l</td>
</tr>
<tr>
<td>Erythrocyte sedimentation rate</td>
<td>11 mm/h</td>
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<tr>
<td><strong>Biochemical</strong></td>
<td></td>
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<tr>
<td>Serum Na</td>
<td>128 mmol/l (135-145)</td>
</tr>
<tr>
<td>Serum K</td>
<td>3.6 mmol/l (3.5-5.5)</td>
</tr>
<tr>
<td>Serum Cl</td>
<td>93 mmol/l (94-106)</td>
</tr>
<tr>
<td>Serum urea</td>
<td>5.6 mmol/l (2.5-6.6)</td>
</tr>
<tr>
<td>Serum creatinine</td>
<td>51 μmol/l (71-133)</td>
</tr>
<tr>
<td>Serum osmolality</td>
<td>273 mOsm/kg (275-305)</td>
</tr>
<tr>
<td>Glomerular filtration rate</td>
<td>25 ml/min*</td>
</tr>
<tr>
<td>Plasma renin activity</td>
<td>55 ng/ml/h</td>
</tr>
<tr>
<td><strong>Immunological</strong></td>
<td></td>
</tr>
<tr>
<td>Lymphocyte transformation: normal</td>
<td>1:4 (normal)</td>
</tr>
<tr>
<td>Helper/suppressor T-cell ratio: normal</td>
<td>1:4 (normal)</td>
</tr>
<tr>
<td>No increase in level of circulating immune complexes</td>
<td></td>
</tr>
<tr>
<td>Mantoux positive</td>
<td>*Normal for mass 118 ml/min.</td>
</tr>
<tr>
<td>*Normal supine 0.15 - 2.33 ng/ml/h; erect 1.3 - 3.9 ng/ml/h.</td>
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was supplemented with intravenous fentanyl 10 μg. Two additional doses of alcuronium 1 mg were required. At the end of the operation neuromuscular blockade was antagonized by neostigmine 1.25 mg and atropine 0.6 mg. A nerve stimulator was used to assess that recovery from neuromuscular blockade was adequate.

Intra-operatively, the patient received the following intravenous fluids: Plasmalyte L 100 ml, 5% dextrose/half-strength Darrow's solution 100 ml, and whole blood 200 ml, which replaced blood as it was lost.

Fig. 1. Cardiovascular and temperature changes and end-expired carbon dioxide levels during anaesthesia.
cation using trimethazine and droperidol (an effective combination in view of the α-adrenergic antagonist properties of the latter), a stress-free induction, and intubation under deep general anaesthesia and muscle relaxation were presumed to be essential. Remarkably, the child’s previously intractable hypertension settled following premedication, and remained stable during the anaesthetic. This experience parallels that of Cobb and Vaughan, who described a case of nephrectomy for a 27-month-old patient with Wilms’s tumour and severe renovascular hypertension. In the event of a sudden rise in blood pressure, a sodium nitroprusside infusion would have been started. Extubation with the patient asleep but breathing spontaneously, is recommended.

Hypotension also needs to be avoided because of abnormal regional blood flows with Takayasu’s arteritis. In this case, after intubation the blood pressure did decrease to 60/25 mmHg, but after reduction of the inspired halothane concentration it increased to 80/42 mmHg within 10 minutes. The child did not appear to suffer any ill-effects from this episode of hypotension. Other procedures which may diminish carotid artery blood flow, for example extension of the head on the atlanto-occipital joint, should be avoided. In the event of pulmonary artery involvement, abnormalities in the ventilation-perfusion relationship may be expected.

A curious feature of this case was the much greater ease of blood pressure control after removal of an apparently unperfused, non-functioning right kidney. Pre-operative plasma renin activity (PRA), sampled from a peripheral vein, was 55 ng/ml/h. Surprisingly, intra-operative PRA was higher when measured in samples drawn from the inferior vena cava (94,7 ng/ml/h) than from the right renal vein (82 ng/ml/h). This anomalous finding suggests the possibility that the specimens were mislabelled at the time of collection or switched around during laboratory procedures. The laboratory acknowledges that the latter may have occurred. Six weeks postoperatively, PRA sampled from a peripheral vein was 2,29 ng/ml/h. Four weeks postoperatively, the erythrocyte sedimentation rate was 82,1 mm/2nd h (Westergren) and the glomerular filtration rate 31/min.

In conclusion, Takayasu’s arteritis is an unpredictable, progressive disorder or group of disorders with many clinical features indicative of vascular involvement, some of which may fluctuate in severity. Thus, alteration of symptoms and signs may be due to the effects of treatment, or to variation in the activity of the arteritis itself. In time the disease may burn itself out, and stenosed vessels may even recanalize. However, the prognosis should be guarded. On the basis of Ishikawa’s follow-up study, our patient (classified as Takayasu’s disease with a single severe complication) will be at greatest risk during the first 5 years after initial diagnosis (25,8% mortality). Between 5 and 10 years after diagnosis there appears to be little increase in mortality.

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REFERENCES