

CASE REPORT

Acute panuveitis and Takayasu's arteritis

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A 12 year old boy presented with severe hypertension and was diagnosed with renal artery stenosis requiring balloon angioplasty. Takayasu's arteritis was subsequently diagnosed, but he also developed acute panuveitis, an entity not previously reported in a child with this condition.

Takayasu's arteritis (TA) is pathologically defined as a pan-arteritis involving intima, media, and adventitia of the larger vessels of the neck, thorax, and abdomen. Ischaemic complications are well recognised, including ischaemic optic neuropathy, but inflammatory eye disease is rarely reported. Mortality usually results from the complications of renovascular hypertension.¹

CASE REPORT

A 12 year old boy of Chinese origin presented with two days of low grade fever, occipital headache, nausea, and vomiting. Blood pressure was 180/110 mm Hg and tympanic temperature 39.9°C, but there were no focal signs of infection, meningism, cardiac murmurs, pulse discrepancy, or abdominal bruits. Fundoscopy, visual acuity, and urinalysis were normal and he had no dysmorphic features or cutaneous stigmata. Erythrocyte sedimentation rate (ESR) was 93 mm/h and creatinine 85 µmol/l.

Hypertension required nifedipine, captopril, atenolol, and prazosin for adequate control. Renal Doppler ultrasound and magnetic resonance angiography (MRA) of the neck, chest, and abdomen revealed moderate to severe, bilateral renal artery stenoses, but all other vessels were normal. Renal angiography (fig 1) identified no significant stenosis on the



Figure 1 Six weeks after initial presentation. Renal angiogram shows severe proximal left renal artery stenosis (arrow).

right, but a 95–99% stenosis of the proximal left renal artery, for which balloon angioplasty was successfully performed. The patient's hypertension resolved within two weeks.

He remained afebrile and normotensive, but complained of intermittent redness of the eyes and photophobia. Despite normal optic discs and visual acuity 6/6 in both eyes, he had evidence of bilateral anterior uveitis with marked ciliary flush, fine keratoprecipitates, and markedly increased cells in the anterior chambers bilaterally, but no posterior synechiae. He was successfully treated with topical steroid drops and cycloplegics for one month. ESR decreased to 23 mm/h. Table 1 lists all relevant investigations.

Table 1 Investigations

Investigation	Result	Normal value
White cell count	10×10 ⁹ /l	4–11×10 ⁹ /l
Urea	7.6 mmol/l	2.1–6.5 mmol/l
24 hour urine collection		
Creatinine clearance	80 ml/min/1.73 m ²	90–150 ml/min/1.73 m ²
Protein excretion	0.17 g/day	0.01–0.1 g/day
Catecholamines	Normal	
Complement levels		
C3	1.95	0.9–1.8
C4	0.36	0.1–0.4
Calcium	Normal	
Angiotensin converting enzyme	Normal	
Immunoglobulins	Normal	
Chest x ray	Normal	
CT head	Normal	
Echocardiogram	Normal	
Prothrombotic screen	Normal	
ANCA/ANA	Negative	
HLA-B27	Negative	
Rheumatoid factor	Negative	
Toxocara/Toxoplasma serology	Normal	



Figure 2 Four months after initial presentation. MRA shows origin of the left vertebral artery (arrowhead) and both subclavian arteries severely stenosed with flow gaps in mid-portions of the subclavian arteries bilaterally—left worse than right (arrows).

Two months later visual acuity deteriorated to 6/36 on the right and 6/60 on the left, and he had feeble upper limb and carotid pulses bilaterally. Slit lamp examination revealed bilateral anterior and intermediate uveitis extending to the posterior chamber, with markedly increased cells in the anterior chambers and vitreous bilaterally and posterior synechiae in the left eye. MRA (fig 2) showed severe stenosis of the origin of the left vertebral artery and both subclavian arteries. ESR was 9 mm/h.

Takayasu's arteritis was diagnosed and treatment commenced with steroid and cycloplegic eye drops and intravenous methylprednisolone. This was followed by an eight week course of oral prednisolone, cyclophosphamide, and aspirin, then maintenance methotrexate and low dose prednisolone. An MRA two months into systemic therapy showed the right subclavian artery was no longer stenosed. Visual acuity returned to normal within three months, but continued to flare intermittently, requiring brief doubling of prednisolone dose.

At 18 months the patient remained normotensive with palpable pulses in all limbs. There was chronic uveitis in the left eye, but the right was normal. Audiometry was normal, antineutrophil cytoplasmic antibody (ANCA) remained negative, and MRA showed a normal left vertebral artery and a patent left subclavian artery, although of smaller calibre than the right.

DISCUSSION

Takayasu's arteritis (TA) is found most commonly in women and in southeast Asia. TA is rare in children and Anglo-Celtic ethnic groups, in whom excluding the syndromes of neurofibromatosis, mucopolysaccharidosis, and Williams syndrome, leaves a differential diagnosis of TA and fibromuscular dysplasia (FMD). Stenoses in FMD are usually distal, while in TA are proximal. Despite extensive imaging, our patient presented with an isolated left renal artery stenosis, but three months later proceeded to fulfil the diagnostic criteria of TA.²

Identification of TA in the inflammatory phase has important implications and was a key dilemma in our patient. The subsequent development of widespread stenoses may have been averted by the early institution of immunosuppression. In favour of TA/vasculitis, he was of southeast Asian descent and had evidence of acute inflammatory disease (uveitis and raised acute phase reactants). However, uveitis is not typical of TA, he remained afebrile after presentation, and the localised single vessel stenosis favoured FMD. For this reason, initial treatment with angioplasty was chosen rather than immunosuppression.

Eye disease in TA is an infrequent, late complication. Marked hypoperfusion of the posterior ciliary artery leads to ischaemia of the retina, choroid, and anterior segment. This results in ischaemic glaucomatous neuropathy, and later, optic atrophy without local arteritis.³

Our case is unique as he had extensive panuveitis without glaucomatous ischaemic changes, negative ANCA levels on four separate occasions, and no evidence of sarcoidosis. Previously reported cases of TA and scleritis or uveitis have been either ANCA positive or had granulomas, and were subsequently co-diagnosed with Wegener's granulomatosis⁴ or sarcoidosis.⁵ There has been one adult patient reported with scleritis and TA, who was ANCA negative, but she did not develop uveitis.⁶ Finally, Cogan's syndrome was excluded by normal audiometry. Cogan's syndrome, which occurs mainly in young adults, involves inflammatory eye disease, vestibulo-auditory abnormalities, and up to 15% having an associated vasculitis.⁷

This patient is the first documented case of TA with acute panuveitis. Its presence may be a guide to the need for systemic immunosuppression, especially in children, where TA is most aggressive. Cyclophosphamide was employed in the initial treatment in our case in order to control the heightened state of systemic inflammation and prevent permanent total blindness. His case also emphasises the need to be alert to the diagnosis of TA as many low incident communities, such as Australia and the UK, now include significant populations from southeast Asia, where TA is the commonest cause of renovascular hypertension.

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