CASE REPORT

'Eye is a window to the pulse': bilateral ocular ischaemic syndrome as a presenting manifestation of Takayasu arteritis

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SUMMARY

A 22-year-old lady presented with gradually progressive visual loss and chronic pain in both the eyes of 3 months duration. The clinical examination and fundus fluorescein angiography were suggestive of bilateral ocular ischaemic syndrome. Her upper limb pulses were feeble and blood pressure was not recordable in the same. Carotid Doppler revealed a bilateral carotid artery occlusion. Systemic markers for inflammation were elevated. Diagnosis of Takayasu arteritis was made on the basis of clinical and laboratory findings.

Takayasu arteritis should be considered in the differential diagnosis of ocular ischaemic syndrome, particularly in young Asian women.

BACKGROUND

Bilateral ocular ischaemic syndrome has rarely been reported as the initial presenting symptom of Takayasu arteritis. Demir *et al*¹ reported the case of a 14-year-old girl who presented with bilateral ocular ischaemia. A case of bilateral ocular ischaemia with unilateral ischaemic optic neuropathy secondary to Takayasu disease in a 42-year-old Pakistani man who was treated with pan-retinal photocoagulation and intravitreal injection of bevacizumab has also been reported.² This rare manifestation of Takayasu arteritis highlights the role of ophthalmologists in diagnosing a life-threatening systemic vascular disease.

CASE PRESENTATION

A 22-year-old lady with no known systemic illness presented to the ophthalmology department of a tertiary care hospital with the complaints of an

insidious onset of gradually progressive visual loss and chronic pain in both eyes since 3 months. She also complained of headache, fatigue and giddiness. After examination her best corrected visual acuity in both eyes was 3/60 and near vision less than N36. The anterior segment examination showed mild ciliary congestion, pigmented keratic precipitates and grade 1 flare. There was no iris neovascularisation. Intraocular pressure was 4 mm Hg in the right eye and 5 mm Hg in the left eye. Gonioscopy showed open angles in both eyes without any angle neovascularisation. A dilated fundus examination showed a normal optic disc, arteriolar narrowing and venous dilatation without much tortuosity. Multiple microaneurysms and punctate haemorrhages were present in the posterior pole extending up to the mid periphery along with few cotton wool spots in both eyes.

A general physical examination revealed feeble upper limb pulses and normal pulses in the lower limbs. There was no audible murmur. Other systems were normal.

INVESTIGATIONS

Fundus fluorescein angiogram showed patchy choroidal filling, delayed arm to retina circulation time (right eye 1 min 5 s and left eye 48 s), and delayed arteriovenous transit time, advancing dye front in the arteries, multiple microaneurysms and vascular staining. There was no neovascularisation (figures 1–4).

Her erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) were elevated, 73 mm/h and 18 mg/l, respectively. Echocardiogram was normal with no evidence of coarctation of the aorta.

Figure 1 A colour fundus photograph of the right eye (A) and left eye (B) showing arteriolar narrowing (horizontal arrows), venous dilation without much tortuosity (vertical arrows). Multiple microaneurysms and punctate haemorrhages (curved arrows) were present in the posterior pole extending up to the mid periphery along with few cotton wool spots in both the eyes.

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Figure 2 Fundus fluorescein angiogram of the right eye showing patchy choroidal filling and delayed arm to retina circulation time (1 min and 5 s).

The carotid and vertebral arteries' Doppler showed bilateral carotid artery occlusion with normal vertebral arteries. A CT of the aorta showed type II Takayasu arteritis (mixed type) involving the arch of the aorta with its branches and descending thoracic aorta causing significant stenosis of the right brachiocephalic, the left common carotid and the left subclavian arteries (figure 5).

DIFFERENTIAL DIAGNOSIS

On the basis of the clinical picture and fundus fluorescein angiogram a diagnosis of bilateral ocular ischaemia was made. The bilateral ocular ischaemic syndrome in young patients can occur owing to a compromise of the vascular lumen secondary to inflammation or infection. The differential diagnosis under consideration was vasculitic conditions like polyarteritis nodosa and Wegener's granulomatosis, Kawasaki's disease and Takayasu arteritis. The age of onset in a young woman with clinical findings of pulselessness, elevated systemic inflammatory markers along with the radiological features of vascular narrowing of the proximal aortic branches suggested the diagnosis of Takayasu disease.

TREATMENT

The patient was started on systemic steroids with a dose of 1 mg/kg body weight.

Figure 4 Late phase fundus fluorescein angiogram of the left eye showing staining of the vessel walls capillary non-perfusion areas (vertical arrows) and microaneurysms in the posterior pole.

She was advised stenting of neck vessels once the active disease the left subsides.

Figure 5 CT angiography of the aorta shows critical stenosis of the right brachiocephalic (BCA), the left common carotid (L CCA) and the left subclavian arteries (L SCA) with a trickle of contrast flow in the residual lumen.

Figure 3 Fundus fluorescein angiogram of the left eye (A) and right eye (B) showing staining of the vessels (horizontal arrows), capillary non-perfusion areas (vertical arrows) and microaneurysms in the posterior pole (curved arrows).

OUTCOME AND FOLLOW-UP

The patient was followed up after 1 month. Her symptoms were slightly improved with a decrease in ESR and CRP. However, she was lost for follow-up for the next few months. A telephonic review at the end of 3 months revealed that she had a sudden cardiac death at home. No medical records were available.

DISCUSSION

Takayasu arteritis is a granulomatous vasculitis of an unknown aetiology affecting the aorta and its major branches and occurring most commonly in women of childbearing age.³ Ocular manifestations of Takayasu disease include Takayasu retinopathy, hypertensive retinopathy and rarely ocular ischaemia. Ocular ischaemia is due to the hypoperfusion by the ophthalmic artery which leads to chronic anterior and posterior segments ischaemia.⁴ Generally ocular manifestations appear late in Takayasu arteritis. Ocular ischaemia as a presenting feature of Takayasu arteritis has been reported very rarely.^{1 2} Though this patient had features of ocular ischaemia in both eyes as found after fundus fluorescein angiography as there was no neovascularisation, we did not perform pan-retinal photocoagulation at the initial visit. Ocular treatment in Takayasu arteritis is aimed at improving the perfusion and preventing neovascularisation.

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Competing interests None.

Patient consent Obtained.

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Learning points

- The patients presenting with ocular ischaemic syndromes should be subjected to a careful general physical examination to rule out ocular manifestations of a more serious systemic disease.
- The possibility of Takayasu arteritis should be considered in ocular ischaemic syndrome when presenting in a young woman.
- Fundus fluorescein angiogram plays an important role in diagnosing ocular ischaemic syndrome and guiding therapy.

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