Bilateral ischaemic optic neuropathy and retinopathy along with cortical infarct in a case of Takayasu disease

Nripen Gaur,[1] Pradeep Sharma,[1] Brijesh Takkar,[1] Jagjeet Singh[2]

[1]Ophthalmology, Dr Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, Delhi, India
[2]Radiology, All India Institute of Medical Sciences, New Delhi, Delhi, India

Correspondence to
Professor Pradeep Sharma, drpsharma57@yahoo.com

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DESCRIPTION

A 27-Year-old female presented with complaints of sudden onset visual loss along with right sided deviation of the angle of mouth since past 30 days. Vision loss had worsened in the last week. There was history of severe headache which was not associated with vomiting. There was no history of any other neurological deficit or prior systemic illness. Examination for cranial nerve function revealed a left sided upper motor neuron facial nerve palsy and rest of the neurological examination was within normal limits. On ocular examination, the patient had best-corrected visual acuity of light perception (PL) in right eye (RE) and 6/36 in left eye (LE). A grade four relative afferent pupillary defect was noted in RE, while slit lamp examination and tonometry were normal. On fundus examination, RE optic disc had yellowish-white pallor while LE optic disc had temporal pallor (figure 1A,B). Multiple retinal micro-aneurysms were present diffusely in both eyes.

Fundus fluorescein angiography revealed delayed filling of the retinal vessels. It confirmed the retinal findings and ruled out capillary non-perfusion and neo-vascularisation (figure 1C,D). Erythrocyte sedimentation rate and C reactive protein were abnormally raised. MRI scan was done which revealed an acute infarction of the right anterior centrum semiovale (figure 2A,B). Magnetic resonance arteriography (MRA) revealed uniformly thickened non enhancing wall of the major aortic arch branches, along with diffuse long segment narrowing of the arteries (figure 2C). Based on the retinal and MRA findings, the patient was diagnosed to be a case of bilateral ischaemic optic neuropathy with seventh nerve palsy secondary to Takayasu disease. The patient was started on oral steroids. At 3 months of follow-up visual acuity in LE had improved to 6/12, however, there was no improvement in the visual acuity of RE.

Takayasu arteritis is a systemic disease and T-cell mediated autoimmune reaction against the vessel wall components is accepted to be the cause.[1] The clinical features include absent or diminished peripheral pulses, hypertension, vascular bruits, retinopathy, dyspnoea and headache. Ocular findings include microaneurysms, ocular ischemia and neovascular glaucoma.[2]

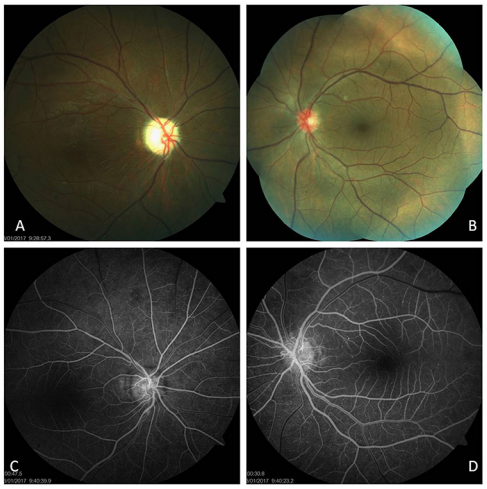

Figure 1 (A,B) Clinical photograph showing total disc pallor in RE along with temporal disc pallor in LE. (C,D) Early phase FFA images of both eyes revealing microaneurysms. These are seen to be scattered diffusely, giving the image a starry-sky appearance.

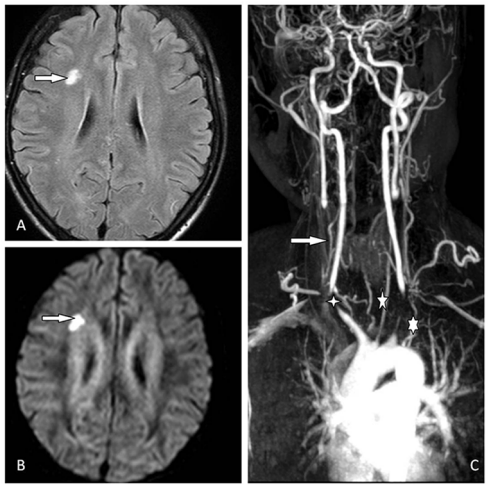

Figure 2 (A) MRI fluid-attenuated inversion recovery image shows a hyper-intense focus located in the right peri-ventricular region. (B) This focus showed diffusion restriction on diffusion weighted image image, suggesting an acute infarct. (C) Magnetic resonance angiography imaging showed narrowing of the brachiocephalic artery and narrowing of the proximal right subclavian artery involving the origin of right vertebral artery (four pointed star).There is long segment narrowing of right common carotid artery (arrow).There is also long segment narrowing of the left common carotid artery (five pointed star) with narrowing of proximal left subclavian artery involving origin of left veretebral artery (six pointed star).

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Retinopathy in Takayasu disease has been staged through milder stages of distension of veins and micro-aneurysms, to arterio-venous anastomoses and severe vision threatening ocular complications.[3] In our case, presence of retinal micro-aneurysms and facial palsy alerted us to the possibility of arteritis related ischemia.

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Competing interests None declared.

Patient consent Obtained.

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Learning points

- Ischaemic optic neuropathy is a rare complication of Takayasu disease.
- Physicians should have a high index of suspicion in young cases with bilateral involvement of the optic nerves and retinal microaneurysms.

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