Blinding vasculitis: bilateral severe vision loss as a presenting feature of undiagnosed Takayasu arteritis

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DESCRIPTION

A middle-aged man in his 50s presented with poor vision in both eyes, left eye (OS) more than the right eye (OD) since 1 week. He gave a history of headache, jaw claudication and easy fatigability for the last few weeks. He was a chronic smoker and alcoholic for the past 20 years and denied a history of any significant systemic illness. Best-corrected visual acuity of OD was counting fingers at 4 m and no light perception in OS. A relative pupillary defect in OS, the anterior segment, was unremarkable in both eyes.

Fundus examination revealed scattered cotton wool spots in the posterior pole with patchy retinal whitening in OD (figure 1A) and pallid disc oedema with central retinal artery occlusion (CRAO) in OS (figure 1B). Optical coherence tomography (OCT) confirmed patchy inner and middle layer hyperreflectivity in OD and diffuse inner layer hyperreflectivity in OS. The findings were consistent with the diagnosis of ischaemic retinopathy in OD and combined arteritic anterior ischaemic optic neuropathy (A-AION) with CRAO in OS.

In view of bilateral ocular ischaemia, he was advised thorough systemic evaluation. Examination revealed absent upper limb pulses with unrecordable blood pressure (BP). In the lower limbs, BP was 140/80 mm Hg. CT angiography of the thorax revealed absent flow in the left subclavian artery with normal flow in the aorta and right subclavian artery. Carotid Doppler studies revealed thrombotic plaques with stenosis. Magnetic resonance angiography of the brain revealed an absence of flow across posterior cerebral arteries with ischaemic changes in occipital and frontoparietal lobes. His C reactive protein titres were 77.6 mg/L. Flourescein angiography studies could not be done as the physician ruled out his fitness for the same. A diagnosis of Takayasu arteritis (TAK) was made and the patient was started on systemic steroids.

At 8-week follow-up, OD vision improved to 6/36 with the clearing of cotton wool spots (figure 2A). OS showed disc pallor, resolution of retinal oedema and cotton wool spots (figure 2B). OCT of the macula OD showed normal foveal contour and OS revealed thinning of inner retinal layers (figure 2C,D).

TAK is a rare chronic granulomatous inflammation-causing intimal fibrosis and vascular narrowing of the aorta, its branches and pulmonary artery.1 The prevalence of eye morbidities in TAK has been noted to range from 8.1% to 68%.2 These manifestations are due to poor blood flow across the aorta, carotid and distal arteries, which may present as global or focal ocular ischaemia. Takayasu retinopathy, retinal findings in patients with TAK, may range from subtle retinal dilatation to severe ischaemia of the retina with neovascularisation.3 Apart from Takayasu retinopathy, retinal artery occlusion, choroidal ischaemia and non-A-AION have been described in patients with TAK.4 The present case represents a severe form of bilateral ischaemic insult secondary to TAK. Our case is unique in that it presented in an elderly man in contrast to a typical young woman, severe bilateral manifestation and total vision loss in one eye. The OS had no light perception owing to combined A-AION with CRAO, which is a very rare presentation. Other differentials like...
ocular ischaemic syndrome, retinal artery occlusion secondary to atherosclerotic disease and nutritional/toxic retinopathy secondary to smoking and alcohol should be considered during evaluation.

Management involves early recognition of the disease with high level of suspicion of large vessel vasculitis. Retinal angiography studies including Doppler studies of retrobulbar vessels may pick up early retinal changes before development of any symptoms. CT and MRI angiographic studies highlight the location and severity of aortic branch involvement which decides the management plan. Corticosteroids, immunosuppressives, and biologics may be needed to control systemic and ocular inflammation, and endovascular intervention may also be required in severe cases.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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REFERENCES

Learning points
► Takayasu arteritis can present with varied ocular manifestations leading to significant ocular morbidity if not for timely intervention.
► Atypical presentation of Takayasu arteritis should be borne in mind in cases with extensive bilateral ocular ischaemia.
► Ocular involvement may be the first presenting feature of many systemic vasculitic disorders including Takayasu arteritis, emphasising the role of the ophthalmologist in diagnosis.

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