Unilateral hemorrhagic keratouveitis as the initial presentation of Takayasu's arteritis

Dear Editor,

A 35-year-old female presented with pain, redness and decreased vision of right eye of one-month duration. Examination showed

corneal edema with macular opacity circumferentially along the peripheral cornea, dispersed hyphema and fine keratitic precipitates. Corneal sensation was normal. Vision was hand movements. The fellow eye was normal with vision of 20/20. She had a similar episode two years back and was treated as herpetic uveitis elsewhere.

The blood pressure was 100/60 and 160/130 mmHg in the right and left arm respectively. The right carotid, brachial and radial pulses were absent. Investigations on the lines of uveitis like serology for rheumatoid factor, anti-nuclear antibodies, anti-neutrophilic cytoplasmic antibodies, angiotensin converting enzyme levels, HLA-B27 typing and 24-h urinary calcium (for sarcoidosis) were inconclusive. Also, infective etiology like syphilis, pulmonary tuberculosis (chest X-ray and Mantoux test) and viral (serology for Varicella zoster and Herpes simplex virus-1 DNA) were ruled out. However, ESR was raised which was 40 mm at one hour.

Ultrasonography of the abdomen revealed small left kidney. Echocardiography was normal. Doppler ultrasound and whole body angiography revealed narrowing of large vessels, mainly right vertebral, right common carotid, right subclavian artery, abdominal aorta and left renal artery [Fig. 1]. Thus, diagnosis of Takayasu's arteritis (TA) was made.

She was started on oral prednisolone (1.5 mg/kg body weight) and topical 1% prednisolone eye drops one-hourly. Hypertension was managed by amlodipine and atenolol. Oral prednisolone was tapered slowly over the next four months. Fundus examination showed few capillary microaneurysms and dilated and tortuous vessels. Fluorescein angiography did not show any evidence of retinal or iris neovascularization. Visual acuity returned to 20/30. It is now two years since her presentation and she had mild flare-up of uveitis twice, which was managed by topical 1% prednisolone eye-drops alone four-hourly, tapered over a month. However, she is still continuing amlodipine and atenolol to control hypertension and on her last follow-up the blood pressure was 90/60 and 130/86 mm Hg in the right and left arm respectively.

Takayasu's arteritis is an occlusive vasculitis of unknown etiology, predominantly affecting young women. It involves the aorta and its branches resulting in end organ hypoperfusion. Patients can present initially with obscure systemic symptoms such as fever of unknown origin or more commonly with signs of large vessel vasculitis such as hypertension from renal artery stenosis or aortitis.[1]

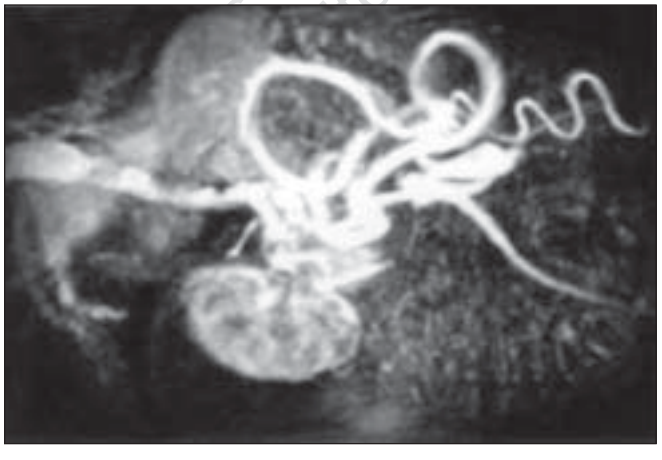

Figure 1: Angiography showing non-visualization of left kidney due to narrowed left renal artery

Ocular complications include ischemia of the retina, choroid and anterior segment.[2] Panuveitis,[3] scleritis'[4] central retinal artery occlusion and recurrent uveitis[5] are rare complications of the disease. However, recurrent unilateral hemorrhagic keratouveitis as its presenting feature has not been reported.

Takayasu retinopathy includes small vessels' dilatation, capillary microaneurysm or arterio-venous anastomosis. The variability of ischemic changes depends upon the duration and rate of ocular vascular insufficiency.[6] Hypertensive retinopathy may also be seen.

The reason for uveitis in TA is not known but may be related to chronic ischemia or the inflammatory process of the disease itself.[7] She had recurrent unilateral manifestation only, which might be related to the narrowing of major blood vessels predominantly on the right side. Though this patient presented with dispersed hyphema neither the retina nor iris had any sign of neovascularization. The source of bleed might be dilated iris vessels, "cobs buds" as TA is known to be associated with small vessel dilatation.

In the absence of clinical signs of ocular ischemia, the uveitis and hyphema in this patient might be related to the inflammatory process of the disease itself. This case underscores the importance of systemic workup in an ocular case and the varied manifestation of a rare disease.

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