Takayasu’s Arteritis with Bilateral Anterior Ischaemic Optic Atrophy

A 22 years old male patient presented with sudden loss of vision in both eyes 5 days prior to admission. There were no other complaints or neurological deficit. On asking leading questions he admitted to having unilateral headache on left side and pain in the arms after heavy exertion for last 2 months which he neglected. There was no past history of diabetes, hypertension and pulmonary tuberculosis. He was a non-smoker and a non-alcoholic.

On clinical examination, he had Marfanoid habitus. He was afebrile. Pulse examination revealed that radial, brachial and axillary artery pulsations were not palpable in both upper limbs. Both the carotids were weakly palpable and there was bruit over both the carotids. Bruit was also heard over the abdominal aorta and right renal artery. Femoral, popliteal, posterior tibial and dorsalis pedis artery pulsations were well felt in both lower limbs. Blood pressure was unrecordable in both upper limbs and was 220/110 in both lower limbs. Systemic examination did not reveal any abnormality.

Eye examination revealed complete loss of vision in both eyes with absence of PR and PL. Fundoscopy showed bilateral anterior ischaemic optic atrophy (Fig. 1). In literature chronic retinal hypoxic changes and ischemic ocular inflammation resulting from cervical vasculart occlusion have been extensively described. The eye signs originally described by Takayasu i.e. new vessel formation in the retina giving a wreath-like appearance are seen in long standing and chronic illness. Anterior ischemic optic neuropathy has rarely been reported in this condition, and only in conjunction with other ophthalmic findings. Marked hypoperfusion of the posterior ciliary artery presumably caused acute onset anterior ischemic neuropathy in this patient. However, presentation with bilateral anterior ischaemic optic atrophy is rare.

Arch aortogram showed involvement of the right brachiocephalic artery (60% stenosis), right common carotid artery (80% stenosis involving short segment of 3 cm from origin), bilateral subclavian artery (proximal >90% stenosis with good collaterals), left common carotid artery (90-95% stenosis in a long segment from its origin with good collaterals) and normal vertebral arteries (Fig. 2). Aortogram further showed narrowing of the distal portion of the thoracic aorta, right renal artery (60% stenosis) and occlusion of proximal portion of superior mesenteric artery. The findings confirmed the diagnosis of Takayasu’s arteritis.

His ESR was raised and all the other investigations for secondary causes of large vessel vasculitis i.e. syphilitic arteritis, SLE, rheumatoid arthritis, other spondyloarthopathies, Behcet’s disease, Kawasaki’s disease were done which included VDRL, ds DNA, ANA, LE cell phenomenon and rheumatoid factor were negative. IgM and IgG antibody against Mycobacterium tuberculosis was also negative. The closest differential diagnosis of Takayasu’s arteritis remains giant cell arteritis which can be differentiated by tissue biopsy. However in absence of tissue biopsy, age at onset (< 40 years), clinical signs of vascular insufficiency in upper limbs, shoulder stiffness and scalp tenderness are the clinical variables that lead to correct diagnosis in 95% cases. A conclusive diagnosis of Takayasu’s arteritis was made in our patient on the basis of 4 out of 6 criteria laid down by the American College of Rheumatology.

This case was unique for it was a male patient who presented earlier i.e. 3rd decade with unusual presentation of bilateral anterior ischaemic optic atrophy. The patient was treated with oral prednisone for 6 weeks and amlodipine 5 mg bd. There was no improvement in his vision and he is still under follow up.

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