Bilateral visual loss as a presenting manifestation Takayasu arteritis

SHAM S SANTHANAM
Department of Rheumatology,
MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract:
Takayasus arteritis is a large vessel vasculitis with involvement of aorta and its major branches. Ophthalmic manifestations are not uncommon and can be found in around 45 of cases. The commonest manifestation are Takayasus retinopathy followed by other ischemic manifestations like anterior ischemic optic neuropathy, central retinal artery occlusion and ocular ischemicsyndrome. We had a 19 year old female who was referred from Ophthalmology with acute onset of bilateral visual loss. She complained of claudication pain of both upper limbs. On examination she had absent pulses over both upper limbs bruit over her neck had severe systemic hypertension. Her fundus examination showed bilateral secondary optic atrophy. Her CT angiogram showed diffuse thickening of aortic wall, its branches and bilateral renal arteries with gross luminal narrowing of both common carotid arteries. So a diagnosis of Takayasus arteritis was made. Bilateral ischemic oculararsyndrome anterior ischemic optic neuropathy due to ocular hypoperfusion has been rarely reported and that too as a presenting manifestation is a rare entity in Takayasu’s arteritis.

Keyword: Takayasus arteritis, Takayasu retinopathy, Ischemic ocular syndrome, Anterior ischemic optic neuropathy

Case Report:
19 year old female presented with sudden onset visual loss of left eye and within 4 days she had loss of vision of right eye. She was able to perceive only light. She also gave history of claudication pain of both upper limbs, episodes of pre syncope a couple of times. She also complained of excessive fatigue and decreased appetite. On examination her bilateral axillary, brachial & radial artery were not palpable. Her lower limb pulses were palpable and BP recorded in her lower limb was 200/90 mm of Hg. She had bilateral carotid and subclavian bruit and also bilateral renal bruit. She had loud A2 on auscultation and had benign hypermobility on musculoskeletal examination.
Her blood investigations revealed an elevated ESR of 80 mm/hr and CRP > 6 mg/L. Her renal function and liver function tests were normal and urine routine didn’t reveal any abnormalities. ECG was suggestive of gross left ventricular hypertrophy. Echocardiogram showed features of concentric LVH & arterial doppler showed absent flow in bilateral upper limb arteries and severe stenosis of bilateral common carotid arteries and right subclavian artery. She was subjected to CT Angiogram and it showed diffuse thickening of aortic wall involving the arch and its major branches & diffuse wall thickening of both common carotid arteries with gross luminal narrowing (left > right). She also had abdominal aortic wall thickening upto 2.2 cms below origin of renal arteries and B/L renal artery wall thickening. CT brain was also done. It showed left frontal infarct though she didn’t give any past history of neurological deficits.

Ophthalmologists opinion was obtained & on fundus examination her right eye showed pale disc with exudates and absent macular foveal reflex. Her left eye also showed a pale disc with retinal folds, exudates and macular oedema and findings were suggestive of secondary optic atrophy of both eyes. Fundus Fluorescein Angiography was done. Both eyes showed dilated disc vessels with dilated long tardy venous columns, extensive microaneurysms & extensive CNP (capillary non perfusion) zones suggestive of chronic ocular ischemia secondary to vascular disease.
Discussion:
Takayasu’s arteritis is a granulomatous large vessel vasculitis usually affecting young females and more common in Asians. Ophthalmic manifestations are not so uncommon. Occlusive arteritis of aorta and its major branches causes ischemic ocular manifestations and when renal artery is involved it causes uncontrolled systemic hypertension and hence hypertensive retinopathy. Our patient had both but the former was the dominating one and the reason for her sudden bilateral visual loss.

Takayasu’s arteritis is classified as per angiogram into 5 classes. Type I involves the major branches of aortic arch. In Type II there is involvement of ascending aorta, arch and its branches (type IIa) & in addition involvement of thoracic descending aorta (type IIb). In type III it is Type IIb along with abdominal aorta with or without renal arteries. In type IV there is isolated involvement of abdominal aorta and/or renal arteries while type V involves the entire aorta and its branches. Our patient had combination of type (II a + IV). This patient fulfills five of the six criteria described by the American College of Rheumatology. It includes patients age being less than 40 years, limb claudication, bilaterally absent brachial pulse, subclavian bruit & angiographic evidence of obstruction of the large arteries of the upper limbs and the carotids.

The well described ischemic ocular manifestation in Takayasu arteritis is Takayasu retinopathy. It has been classified into four stages by Uyama and Asayma. Stage 1 is characterized by distension of veins, stage 2 by microaneurysm formation, stage 3 by the formation of arterio-venous anastomoses and stage 4 by the presence of ocular complications like cataract, rubeosis iridis, retinal ischemia, neovascularization and vitreous hemorrhage. Anterior ischemic optic neuropathy, central retinal artery occlusions and hypertensive retinopathy are other eye manifestations of Takayasu’s arteritis.

The ischemic retinal changes in Takayasu arteritis depends on which part of the carotid artery becomes involved, the rate and duration of occlusion & the degree of collateral supply. Our patient had severe occlusion of aortic branches leading to ischemic ocular syndrome & anterior ischemic optic neuropathy and hence visual loss. When abdominal aorta with renal artery is involved then patient develops severe systemic hypertension and if not controlled leads to hypertensive retinopathy changes.

Conclusion: Takayasu’s arteritis can cause acute onset visual loss (as in our patient) or as subacute and chronic visual loss. Usually it affects the younger age group and the impact of a vision threatening ocular complication on their lives can be devastating. All patients must undergo baseline ophthalmic examination and regular checkups on followup. Revascularisation procedures can be tried for ischemic ocular syndrome and the disease per se has to be kept under control with steroids and other immunosuppressive drugs like methotrexate. If patient had associated systemic hypertension it has to be adequately controlled with antihypertensives. This patient was given pulse methyl prednisolone followed by oral steroids (1 mg/kg, tapered gradually) plus methotrexate & three antihypertensives. Her vision was status quo with improvement in claudication symptoms. This patient probably had bilateral sequential ischemic ocular syndrome as initial presentation and such cases had been rarely reported in literature.
References:


