

# Takayasu Arteritis Presenting as Neovascular Glaucoma: A Rare Case Report

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## ABSTRACT

Neovascular glaucoma secondary to bilateral ocular ischemic syndrome has never been reported as an initial presenting manifestation of Takayasu arteritis. We report a case of 38-year-old male with no known systemic illness presenting with loss of vision and chronic pain in both eyes since 6 months. On examination, patient had BCVA of FCCF with mid dilated and non-reacting pupil in both eyes. His IOP was 28mmHg bilaterally with GAT. Patient had no NVI on slit lamp examination but had NVA on gonioscopy. On fundus examination, patient had neovascularisation of disc (NVD) with sclerosed vessels with severe tessellation in both eyes. On general physical examination, patient had feeble upper limb pulses and normal pulses in the lower limbs with no audible murmur. Blood pressure could not be recorded in upper limb however BP recorded in lower limb was 120/80 mmHg. Carotid Doppler and computed tomogram (CT) angiography revealed complete occluding thrombus in B/L common carotid artery with minimal central flow in internal carotid artery. Patient was diagnosed as a case of NVG with B/L OIS secondary to Takayasu arteritis. This rare manifestation of Takayasu arteritis highlights the role of ophthalmologists in diagnosing a life-threatening condition where a prompt referral can be life saving for the patient.

**Key Words:** Ocular ischemic syndrome, Neovascular glaucoma, Takayasu arteritis.

## INTRODUCTION

Ocular ischemic syndrome is a rare sight threatening condition, which is caused by ocular hypo perfusion secondary to

severe carotid artery occlusive disease (stenosis or occlusion).<sup>1</sup> Atherosclerosis is the most common cause for changes in carotid arteries. It occurs mainly in elderly at a mean age of 65 years, with no racial predilection. Males are affected more commonly with M:F ratio of 2:1 due to higher incidence of atherosclerotic disease in males.<sup>2</sup> Incidence of ocular ischemic syndrome is reported to be 7.5 cases per million persons every year, however it may be an underestimation because it is usually misdiagnosed as retinal venous occlusions or diabetic retinopathy. Bilateral involvement is uncommon and seen in 22% cases only.<sup>3</sup>

It presents with loss of vision, pain and various signs of anterior and posterior segment ischemia. Signs of anterior segment ischemia are neovascularisation of iris (NVI) and secondary neovascular glaucoma (NVG), iridocyclitis, iris atrophy and sluggish reacting pupil. On posterior segment examination, narrowed retinal arteries, dilated retinal veins, mid peripheral retinal haemorrhages, perifoveal telangiectasia and neovascularisation of disc (NVD) are characteristic findings. Differential diagnosis of ocular ischemic syndrome are diabetic retinopathy and moderate central retinal vein occlusion.<sup>4</sup> Carotid artery imaging and fundus fluorescein angiography (FFA) are important investigations for establishing the diagnosis of ocular ischemic syndrome.<sup>5</sup>

In ocular ischemic syndrome patients, ipsilateral stenosis of  $\geq 90\%$  of

common or internal carotid artery is usually found. Rarely, ophthalmic artery occlusion is responsible for OIS.<sup>6</sup> Most common cause is atherosclerosis in majority of patients. Other less common causes for ocular ischemic syndrome are Takayasu arteritis, giant cell arteritis, dissecting aneurysm of the carotid artery, fibrovascular dysplasia, aortic arch syndrome, Behcet's disease, trauma or inflammation leading to carotid artery stenosis, thyroid orbitopathy, hyperhomocysteinemia and complications after intravitreal anti-VEGF injections and radiotherapy for nasopharyngeal carcinoma.<sup>6-9</sup> As OIS is associated with atherosclerosis, other related co-morbid conditions are also frequently present in these patients. Hypertension is found in 73% patients of OIS and diabetes mellitus is found in 56% cases. The mortality rate in ocular ischemic patients is quite high, which is around 40% within 5 years of onset secondary to cardiovascular disease (66%) and stroke.<sup>10</sup> Therefore patients with OIS should be promptly referred to the cardiologist, neurologist and vascular surgeon.

Management of OIS is a multidisciplinary approach. Ocular treatment is aimed to treat the ocular complications and prevent further damage. Topical steroids are given to suppress anterior segment inflammation and topical cycloplegics are given to stabilize the blood-aqueous barrier and restrict iris movements to decrease chances of spontaneous hyphema. To control IOP, topical beta blockers and alpha agonists are drugs of choice. Prostaglandins and anticholinergic drugs should be avoided as it may increase ocular inflammation. If NVG develops, IOP control becomes refractory to medical treatment. Surgical options in such scenario are trabeculectomy with antimetabolites, aqueous shunt implants and diode laser cyclophotocoagulation. If there is uncontrolled IOP with no visual potential, cycloablation can be performed. If eye is blind and remains painful, retrobulbar injection of alcohol can be given. If still

pain persists in a blind eye, enucleation or evisceration is the last resort.<sup>11</sup>

Carotid Artery Endarterectomy (CEA) is indicated for symptomatic stenosis of 50-99%, if perioperative risk of stroke or death is <6%. In asymptomatic patients, CEA is indicated for 60-99% stenosis, if perioperative risk of stroke or death is <3%.<sup>12</sup>

Carotid Artery Stenting (CAS) is an alternative for CEA in recurrent stenosis after CEA, tracheostomy, carotid stenosis above C2 vertebra, unstable angina, recent MI and congestive heart failure patients.<sup>13</sup>

Extracranial-Intracranial Arterial Bypass Surgery is surgical anastomosis of the superficial temporal artery with a branch of middle cerebral artery. It is indicated in complete occlusion of ICA or CCA or when ICA stenosis is at or above C2 vertebra.<sup>14</sup>

## CASE REPORT

Neovascular glaucoma secondary to bilateral ocular ischemic syndrome has never been reported as an initial presenting manifestation of Takayasu arteritis. We report a case of 38 year old male with no known systemic disease presented to RIO, PGIMS Rohtak with an insidious onset of gradually progressive loss of vision which was associated with chronic pain in both eyes since 6 months. On examination, patient had BCVA of FCCF with mid dilated and non-reacting pupil in both eyes. Anterior segment examination showed mild ciliary congestion with grade 1 flare. Patient had no neovascularisation of iris (NVI) on slit lamp examination but had neovascularisation of angle (NVA) on gonioscopy. His IOP was 28mmHg bilaterally with GAT. On dilated fundus examination, patient had neovascularisation of disc (NVD) with sclerosed vessels with severe tessellation in both eyes.

On general physical examination, patient had feeble upper limb pulses and normal pulses in the lower limbs with no audible murmur. Blood pressure could not be recorded in upper limb however BP

recorded in lower limb was 130/80 mmHg. Other systems were essentially normal.

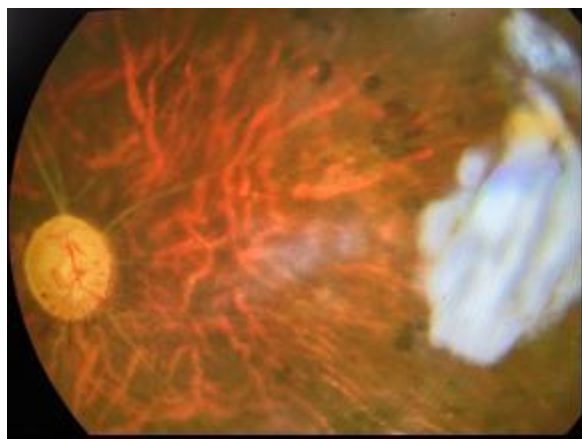


Figure 1: Right eye fundus showing NVD with Sclerosed vessels and bony spicules in midperiphery

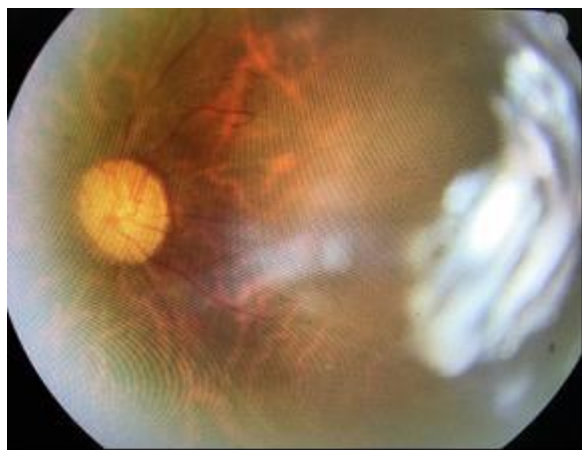


Figure 2: Left eye fundus showing NVD with attenuated arteries with few dot blot haemorrhages

His erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) were elevated, 53 mm/h and 18 mg/l, respectively. On carotid Doppler, B/L common carotid artery showed occlusion with echogenic thrombus till its bifurcation into ICA and ECA. Bilateral ICA showed minimal central flow on color doppler with peripheral thrombus occluding lumen. Patient was advised CT angiography of neck which revealed soft plaque completely occluding bilateral common carotid artery, complete occlusion of 1st part of left subclavian artery, complete occlusion of brachiocephalic trunk and attenuated caliber of right proximal of ICA.

On the basis of the clinical picture and carotid doppler, diagnosis of NVG secondary to bilateral ocular ischemia was

made. The bilateral ocular ischemic syndrome in young patients can occur secondary to giant cell arteritis, Takayasu arteritis, hyperhomocysteinemia, polyarteritis nodosa and Wegener's granulomatosis. The young age of onset with clinical findings of pulselessness, elevated ESR and CRP along with the radiological features of vascular narrowing of the proximal aortic branches suggested the diagnosis of Takayasu disease.

The patient was started on systemic steroids with a dose of 1 mg/kg body weight with topical beta blockers, alpha agonist and carbonic anhydrase inhibitors to control IOP. He was urgently referred to vascular surgeon for Extracranial-intracranial arterial bypass surgery as there was complete occlusion of CCA and ICA.

The patient came for follow up after 1 month after his bypass surgery. His symptoms got improved slightly with a decrease in ESR and CRP. His IOP was 18mmHg in both eyes. However, unfortunately his vision did not get improved. But diagnosing a life-threatening condition like Takayasu arteritis and a prompt referral can be life saving for the patient.

## DISCUSSION

Ocular ischemic syndrome (OIS) is a disorder which occurs secondary to chronic ocular hypoperfusion. Atherosclerosis of carotid artery is the main etiology for majority of OIS cases. 90% or more stenosis of ipsilateral carotid artery reduces the central retinal artery perfusion pressure by 50% leading to ocular hypoperfusion.<sup>15</sup> Other less common causes for ocular ischemic syndrome are Takayasu arteritis, giant cell arteritis, dissecting aneurysm of the carotid artery, fibrovascular dysplasia, aortic arch syndrome, Behcet's disease, trauma or inflammation leading to carotid artery stenosis, thyroid orbitopathy, hyperhomocysteinemia and complications after intravitreal anti-VEGF injections and radiotherapy for nasopharyngeal carcinoma.<sup>6-9</sup> Takayasu arteritis, also known

as aortic arch syndrome/ pulseless disease, is a large vessel granulomatous vasculitis with massive intimal fibrosis and vascular narrowing. It mostly affects young or middle age women of Asian descent, though it can affect anyone. Females are more commonly affected with a M:F ratio of 9:1.<sup>16</sup> It mainly affects aorta and its branches, as well as pulmonary arteries.<sup>17</sup>

If there is conjunctival chemosis and congestion with mild inflammatory reaction in AC, fixed semi-dilated pupil with retinal opacification with no cherry red spot and mid peripheral retinal haemorrhages, it immediately clinch to the diagnosis of ocular ischemic syndrome. In OIS despite of anterior segment neovascularization and uveal ectropion, there is normal intraocular pressure which is thought to be due to ischemia of ciliary body and reduced aqueous humor production leading to normal IOP.<sup>18</sup> On FFA, delayed choroidal perfusion and non-filling of retinal vessels confirms the diagnosis.

Whenever a patient of ocular ischemic syndrome presents to an ophthalmologist, a detailed workup is must in all patients to look for systemic associations such as carotid or coronary occlusive disease, and atherosclerosis. In the absence of these, we should look for rare associations such as Takayasu arteritis, giant cell arteritis, dissecting aneurysm of the carotid artery, fibrovascular dysplasia, Behcet's disease, trauma or inflammation leading to carotid artery stenosis, thyroid orbitopathy, hyperhomocysteinemia and complications after intravitreal anti-VEGF injections and radiotherapy for nasopharyngeal carcinoma.<sup>6-9</sup>

The differential diagnosis of OIS is diabetic retinopathy and central retinal vein occlusion (CRVO). In CRVO, there are dilated and tortuous veins while in OIS, veins are only dilated with no tortuosity. In OIS, intraretinal hemorrhages are less numerous than in diabetic retinopathy and presence of hard exudates suggests diabetic retinopathy. Diabetic retinopathy may coexist with ocular ischemic syndrome,

therefore in diabetic patients, if there is marked asymmetry of retinopathy, patient should be examined for possible carotid artery stenosis as almost 20% of such patients may have a coexisting haemodynamically significant carotid artery stenosis.<sup>19</sup> On FFA, absence of retinal arterial stasis and choroidal filling defects is an important feature to distinguish diabetic retinopathy and CRVO from OIS.<sup>20</sup>

In literature, NVG has never been reported as a presenting manifestation of Takayasu arteritis secondary to bilateral OIS. The mean age of presentation of ocular ischemic syndrome is 65 years and it is rare before the age of 50, however our patient was only 38 year male with bilateral OIS, which is seen in only 22% cases. There was no known systemic disease in our patient. Moreover, he had no history of episodes of transient vision loss. On general physical examination, radial pulse was non-palpable which arouse a suspicion of Takayasu arteritis. His erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) were elevated, 53 mm/h and 18 mg/l, respectively. On carotid Doppler, B/L common carotid artery showed occlusion with echogenic thrombus till its bifurcation into ICA and ECA. Bilateral ICA showed minimal central flow on color doppler with peripheral thrombus occluding lumen. Patient was advised CT angiography of neck which revealed soft plaque completely occluding bilateral common carotid artery, complete occlusion of 1st part of left subclavian artery, complete occlusion of brachiocephalic trunk and attenuated caliber of right proximal of ICA. After managing his NVG, patient was urgently referred to vascular surgeon for Extracranial-intracranial arterial bypass surgery as there was complete occlusion of CCA and ICA.

## CONCLUSION

Ophthalmologists need to be especially alert while dealing with ocular ischemic syndrome, as it is not only a sight threatening condition but also associated with underlying life threatening conditions.

We should always look beyond the eye when we see a patient of NVG as it can be secondary to chronic ocular hypoperfusion. OIS may present for the first time as NVG without any preceding ischemic events elsewhere in the body. It has poor visual prognosis but main concern is that five-year mortality rate is around 40% in these cases, which is significantly high. A careful clinical examination combined with targeted ophthalmic investigations, systemic examination and required investigations may clinch the diagnosis. Along with ocular treatment, a prompt referral to the concerned specialists for appropriate minimally invasive or surgical intervention may be life saving for the patient.

### REFERENCES

1. Bioussé V. Carotid disease and the eye. *Curr Opin Ophthalmol* 1997;8:16-26.
2. Mendrinos E, Machinis TG, Pournaras CJ. Ocular ischaemic syndrome. *Surv Ophthalmol*. 2010;55:2-34.
3. Sturrock GD, Mueller HR. Chronic ocular ischemia. *Br J Ophthalmol* 1984;68:716-23.
4. Sharma S, Brown GC: Ocular Ischaemic Syndrome. Ryan SJ, Hinton DR, Schachat AP et al (ed.). Elsevier, 2006; 1491–502.
5. Brown GC, Magargal LE: The ocular ischaemic syndrome. Clinical, fluorescein angiographic and carotid angiographic features. *Int Ophthalmol*, 1988; 11(4): 239–51
6. Brown GC. Macular edema in association with severe carotid artery obstruction. *Am J Ophthalmol* 1986;10(2):442-8.
7. Imrie FR, Hammer HM, Jay JL. Bilateral ocular ischaemic syndrome in association with hyperhomocysteinemia. *Eye*. Jul 2002; 16(4):497-500.
8. Mizener JB, Podhajsky P, Hayreh SS. Ocular ischaemic syndrome. *Ophthalmology*. 1997;104(5):859-64.
9. Sturrock GD, Muller HR. Chronic ocular ischaemia. *Br J Ophthalmol* 1984;68(10): 716-23.
10. Sivalingam A, Brown GC, Menduke H. The ocular ischemic syndrome: Mortality and systemic morbidity. *Int Ophthalmol* 1989; 13(3):187-91.
11. Bloom PA, Sharma K et al. Trans-scleral diode laser cyclophotocoagulation in advanced refractory glaucoma. *Ophthalmology*. 104;1508-19.
12. Goldstein LB, Adams R, Alberts MJ et al. Primary prevention of ischemic stroke: A guideline from American Heart Association. *Circulation* 2006;113(24):873-923.
13. Narins CR, Illig KA. Patient selection for carotid stenting vs endarterectomy: a systemic review. *Journal of vascular surgery* 2006;44(3):661-72.
14. Sundt TM, Piepgras DG, Houser OW. Bypass surgery for vascular disease of carotid system. *Mayo Clinic* 1976;51(11): 677-92.
15. Kearns TP. Ophthalmology and the carotid artery. *Am J Ophthalmol* 1979;88:714-22.
16. John B, Kiyoshi M. FDG-PET of Takayasu arteritis. *Journal of General Internal Medicine* 2014;29(7):1072-3.
17. Milan B, Josip K. Ocular manifestation of aortic arch syndrome. *Annales d'Oculistique* 1967;200(11):1168-79.
18. Furino C, Guerriero S, Boscia F, Ferrari TM, Cardascia N, Sborgia L, et al. *In vivo* evidence of hypotrophic ciliary body in ocular ischemic syndrome by ultrasound biomicroscopy. *Ophthalmic Surg Lasers Imaging* 2007;38:505-7.
19. Mendrinos E, Machinie TG, Pournaras CJ: Ocular Ischemic Syndrome. *Surv Ophthalmol*. 2010; 55(1): 2–34.
20. Sharma S, Brown GC: Ocular Ischemic Syndrome. Ryan SJ, Hinton DR, Schachat AP et al (ed.). Elsevier, 2006; 1491–502

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