CASE REPORT

Ocular Ischemic Syndrome and Ischemic Optic Neuropathy in Takayasu Arteritis

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Abstract

Ocular Ischemic Syndrome (OIS) is a disorder, which consists of constellation of signs and symptoms secondary to chronic ocular hypoperfusion. We report a case of 35 years old Pakistani female who presented to us with gradual fall of vision in both eyes. On examination, she had bilateral OIS and ischemic optic neuropathy. The patient had past history of transient ischemic attack and facial palsy almost 20 years back. We referred her to physician for the diagnosis of Takayasu arteritis associated with relapsing polychondritis. She was given 60 mg of dexamethasone per day as initial treatment. At her last follow-up after approximately 3 months, she was quite comfortable systemically and her general condition was improved. Unfortunately, her vision did not improve. These two autoimmune diseases are rarely reported together in the same patient in Pakistan. Whether this association is rare or is underdiagnosed still remains to be answered.

Key Words: *Ocular ischemic syndrome. Takayasu arteritis. Relapsing polychondritis. Ischemic optic neuropathy. Carotid artery stenosis.*

INTRODUCTION

Takayasu arteritis is an inflammatory and stenotic disease of medium and large sized arteries with a strong predilection for aortic arch and its branches. The Chapel Hill Consensus Conference on the nomenclature of systemic vasculitis defined Takayasu arteritis as "granulomatous inflammation of aorta and its major branches."[1] The aortic branches are more commonly involved at the origin than distally. A small percentage of Takayasu patients have associated Relapsing Polychondritis (RPC).[2]

Ocular ischemic syndrome is one of the ocular manifestations of Takayasu arteritis. The authors hereby describe a patient with decreased intra ocular pressure, arteriolar narrowing, venous dilation and ischemic optic neuropathy in both eyes.

CASE REPORT

A 35 years old Pakistani female presented in outpatient department in March 2012, with gradual loss of vision in both eyes (more marked in the right eye) for 5 years. There was no history of ocular redness, pain, transient loss of vision, ocular trauma, haloes around light and no history of any discharge from the eyes. Family history regarding vision was unremarkable. Probing the systemic history revealed fever, malaise and weight loss. She had depressed nasal bridge for the last one year, joint pains for 2 years, breathlessness for 4 years, pain in neck, shoulders and back for 5 years. There was morning stiffness and poor healing of wounds.

On enquiring the past history, it was found that she had weakness of the right half of the body at the age of 20 years, which came out to be left ischemic cerebro-vascular accident on CT scan. One year later, she developed facial nerve palsy of the left side.

On ocular examination, her visual acuity was hand movements in right eye and 6/36 in left eye with no improvement with pinhole. Colour vision was defective. On applanation tonometry, intra ocular pressures were 5 mm and 4 mm of Hg in the right and left eye respectively. Pupillary reactions to light were absent in right and sluggish in the left eye. Extra ocular movements were of full range. On slit lamp examination, anterior segment was normal. Fundoscopy revealed bilateral disc pallor. There was retinal venous dilatation and arteriolar attenuation. Blood flow in the retinal arterioles was sluggish revealing cattle trucking phenomenon (Figure 1). A slight pressure on the globe revealed central retinal artery pulsations. Macular reflex was dull on both sides.

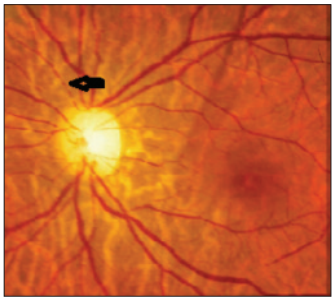

Figure 1: Left fundus photograph showing arteriolar attenuation, disc pallor and interrupted flow in the arterioles (Black arrow).

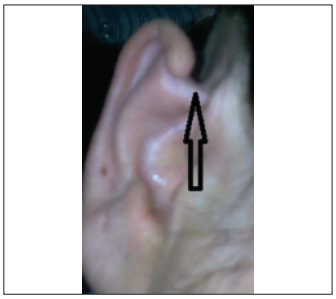

Figure 2: Deformed right ear with loss of auricle at root of helix.

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On general physical examination, the patient had a pale look but was well oriented in time and space. Radial pulses were not palpable and blood pressure was not recordable in the upper limbs and 80/50 mmHg in the lower limbs. Respiratory rate was 24/minute. Cardiovascular examination revealed absent pulses in upper limb and carotid arteries. Pulsations in lower limbs of both sides were weak. There was no bruit over the carotids. Respiratory and gastrointestinal systems were normal. On CNS examination, optic nerve dysfunction was found on both sides and vestibulo-cochlear dysfunction on the right side. ENT examination showed marked saddle shaped nose deformity and a large septal perforation. Right ear was deformed with loss of auricle at root of helix (Figure 2). Ear canals and ear drums were normal. Rinne's test was positive bilaterally and Weber's test was lateralized to the left indicating sensorineural loss in the right ear.

Patient was advised blood, urine, serological and radiological tests. Haemoglobin was 11.9 g/dl, RPR (rapid plasma regain) for syphilis and RA factors were negative, cANCA and pANCA were < 0.5 U/ml (normal).

On Carotid Doppler, right and left carotids were occluded by intimo-medial thickening. Only streak of blood could be seen flowing through them. Vertebral arteries had the similar morphology. Doppler ultrasound of upper limb showed no appreciable flow in subclavian arteries and its subsequent branches on either side. The arterial tree of lower limbs was patent bilaterally. However, progressively dampened waveform was discernible below the popliteal arteries and very weak flow in the tibial circulation. Renal perfusion was good. CT scan of the chest and echocardiography were normal. Temporal artery biopsy was performed to rule out giant cell arteritis and it was found normal.

On the basis of history, examination and investigations, a diagnosis of bilateral ocular ischemic syndrome with ischemic optic neuropathy secondary to Takayasu arteritis and relapsing polychondritis was made. She fulfilled the McAdam criteria for relapsing polychondritis.

She was given 60 mg of dexamethasone per day as initial treatment. At her last follow-up after approximately 3 months, she was quite comfortable systemically and her general condition was improved with no joint pains and breathlessness. Unfortunately, her vision did not improve.

DISCUSSION

Ocular Ischemic Syndrome (OIS) is a condition, which results from chronic hypoperfusion secondary to severe ipsilateral carotid stenosis. The most common cause of OIS is atherosclerosis. Hence, OIS is uncommon in patients younger than 50 years but Takayasu arteritis (which was the cause of OIS in this case) is common in young females less than 50 years of age. Takayasu was an ophthalmologist who first described this disease in a 21-year woman with arteriovenous anastomosis around papilla in 1908.[3]

Takayasu arteritis commonly affects women of child bearing age and is more common in Asians. Constitutional symptoms include weight loss, headache, malaise, arthralgia and fever. There is carotid artery bruit in 80% cases, difference in the blood pressure of extremities, claudication, carotodynia, hypertension, congestive heart failure and aortic regurgitation. TIA, stroke, OIS and seizures are also associated with Takayasu arteritis.

Ocular signs of Takayasu disease include episcleritis, corneal oedema, ischemic pseudoiritis, iris atrophy, low intra-ocular pressure, neovascular glaucoma, cataract, retinal vascular changes, cotton wool spots, disc oedema, exudative retinal detachment and macular oedema. Spontaneous arterial pulsations are also seen in OIS.[4] In this patient, there was hypotony, bilateral arteriolar narrowing, venous dilation, box carrying phenomenon in arterioles due to slow blood flow and disc pallor. All these signs were indicative of ocular ischemia.

Hypertensive as well as hypoperfusive retinopathy has been described in Takayasu arteritis. When there is renal involvement, hypertensive retinopathy occurs. However, stenosis of common carotid artery results in chronic ophthalmic artery insufficiency, which is the main source of blood supply to the eyeball. This results in hypoperfusive retinopathy. Peter described 2 patients with hypertensive and 2 patients with hypoperfusive ocular changes.[5] Hypoperfusive changes were ocular ischemic syndrome and anterior ischemic optic neuropathy. Hypertensive changes were exudative retinal detachment and papilloedema. In a retrospective study of 11 patients, Loukil *et al*. gave an account of hypoperfusive retinopathy with Takayasu disease.[6] In another retrospective study of 15 cases (30 eyes), Wang and colleagues discussed 16 eyes with ischemic retinopathy and 14 eyes with hypertensive retinopathy.[7]

This patient had sensorineural hearing loss. Takayasu arteritis as well as polychondritis are associated with sensorineural hearing disturbance. Whether this particular patient suffered hearing problem from Takayasu arteritis or polychondritis was not further evaluated as treatment in both cases was oral corticosteroids, which were started as soon as the diagnosis was made.

From Pakistan, only few case reports of ocular Takayasu arteritis are available.[8] Pelegrin reported bilateral OIS and unilateral ION as initial manifestation of TA in a 42-year Pakistani male.[9] Mc Adam *et al*. reported 25 - 35% patients of RPC with other autoimmune diseases.[2] Among them, only 2% of RPC had Takayasu arteritis. Patients with RPC can have retinal occlusive vasculitis

but in this case, the retinal findings were due to carotid stenosis and retinal vasculitis was not present.

Ophthalmologists have an important role in early and accurate diagnosis of many systemic diseases where a prompt referral to the concerned specialist can save many lives. This patient had cerebrovascular accident and facial palsy at the age of 15 years but she was not investigated for the cause until she developed bilateral vision loss at 35 years of age.

OIS and ischemic optic neuropathy in Takayasu arteritis with relapsing polychondritis is rarely reported in Pakistan. Whether this association is rare or is under-diagnosed still remains to be answered.

REFERENCES

1. Jennette JC, Falk RJ, Andrassy K, Bacon PA, Churg J, Gross WL. Nomenclature of systemic vasculitides. Proposal of an international consensus conference. *Arthritis Rheum* 1994; **37**: 187-92.
2. McAdam LP, O'Hanlan MA, Bluestone R, Pearson CM. Relapsing polychondritis: prospective study of 23 patients and a review of the literature. *Medicine (Baltimore)* 1976; **55**:193-215.
3. Takayasu M. A case with peculiar changes of the central retinal vessels. *Acta Soc Ophthalmol Jpn* 1908; **12**:554.
4. Brown GC, Magargal LE. The ocular ischemic syndrome. Clinical, fluorescein angiographic and carotid angiographic features. *Int Ophthalmol* 1988; **11**:239-51.
5. Peter J, David S, Joseph G, Horo S, Danda D. Hypoperfusive and hypertensive ocular manifestations in Takayasu arteritis. *Clin Ophthalmol* 2010; **21**:1173-6.
6. Loukil I, Mallouch N, Hachicha F, Bhiri R, Hijazi A, Blouza J. Ischemic retinopathy in Takayasu disease. *Presse Med* 2012; **42**:e594-8.
7. Wang WW, Ye JJ, Chen YX, Dai RP. Fundus manifestations and treatment of Takayasu arteritis. *Zhonghua Yan Ke Za Zhi* 2012; **48**:124-30.
8. Khealani BA, Baig SM. Takayasu's arteritis presenting as ischemic stroke: case report. *J Pak Med Assoc* 2002; **52**: 263-5.
9. Pelegrin L, Mesquida M, Rey A, Sanchez DB, Frohman L. Blind runner. *Surv Ophthalmol* 2012; **57**:486-94.