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Management of Bilateral Ocular Ischemic Syndrome in Takayasu Arteritis: Our Experience and a Brief Review Stressing Need for Multimodal Approach

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Abstract

Objectives: To elaborate the presentation of three patients of Takayasu Arteritis type V, who presented with acute onset binocular diminution of vision and discuss various aspects of management in Ocular ischemic syndrome (OIS).

Methods: Three patients satisfying the 1990 ACR classification criteria for Takayasu Arteritis, presented with bilateral acute OIS to a rheumatology clinic at a tertiary care center. They were managed by pharmacotherapy and percutaneous transluminal angioplasty. Our experience of managing these patients and a brief review is elaborated. Prior institutional ethics board approval has been obtained.

Results: All three patients with OIS who did not improve with pharmacotherapy including pulse glucocorticoids, had good improvement in best-corrected visual acuity (BCVA) after percutaneous transluminal angioplasty involving bilateral internal/ common carotid arteries.

Conclusion: Acute OIS in TA is a vision threatening emergency. In patients who fail to improve on pharmacotherapy, multidisciplinary approach is warranted. Early revascularization procedures can achieve good visual outcome and prevent irreversible damage.

Keywords: Arteritisa • Takayasu • Pulseless disease • Retinal disease • Hypoperfusion

Introduction

Takayasu Arteritis (TA) is a chronic, granulomatous large vessel vasculitis with a symptom spectrum ranging from asymptomatic to life threatening or organ threatening features like vision loss, transient ischemic attack, acute coronary syndrome and congestive heart failure [1]. Ocular symptoms in TA could be both treatment-related or disease-related and affect up to 8.1 to 68% of the patients [2]. Ocular Ischemic syndrome (OIS) is a rare manifestation in TA which can cause irreversible retinal ischemia and neovascular changes if not treated promptly. OIS denotes hypoperfusion of both anterior and posterior segments of eye and orbital structures supplied by the ophthalmic artery because of common/internal carotid artery occlusion [2,3]. Mild OIS is characterized by generalized vasodilation of retinal vessels with capillary microaneurysms and progression causes iris neovascularization, neovascular glaucoma and neovascularization in the retina. Findings supporting OIS include near total or total stenosis of bilateral common carotid or subclavian arteries, mid-dilated poorly reacting pupil, nonperfusion macular edema, capillary nonperfusion areas, iris/ angle/ retinal neovascularization, microaneurysms, retinal arteriolar narrowing and increased arm-to-retina circulation time. Other disease related causes of blindness in TA include hypertensive retinopathy and mixed retinopathy (presence of both hypoperfusive and hypertensive retinopathy) [4,5]. Although described as a chronic phenomenon, we report three young female patients, diagnosed cases of TA Type V (Angiographic Classification) who presented to us with acute binocular diminution of vision who were managed with individually tailored approaches to restore vision.

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Materials and Methods

Patients who were diagnosed with TA satisfying 1990 ACR classification criteria, presented with acute onset diminution of vision. Detailed history taking was obtained and data regarding patient demographic details, symptoms, clinical signs (peripheral pulses, blood pressure measurement in all four limbs, vascular bruits), neuro-ophthalmological assessment, CT aortography changes and details of intervention were recorded after obtaining patients' written informed consent. Detailed neuro-ophthalmological assessment, including best-corrected visual acuity, perimetry, slit lamp examination, applanation tonometry, fundus photograph and fundus fluorescein angiography (FFA) was performed for each patient. Other causes of acute onset diminution of vision like infections, posterior circulation stroke were ruled out. These patients had a baseline CT angiography of aorta (and major branches) and type of TA based on angiographic findings was noted. Based on findings and discussion with ophthalmologist, a diagnosis of bilateral OIS was made and further management was based on treating team's decision. The patients who did not improve with pharmacotherapy were considered for reperfusion strategies in discussion with interventional radiologist. Disease activity was measured by ESR, CRP levels and ITAS-A (Indian Takayasu Arteritis Score-A) which scores a new onset abnormality in the past 3 months [6]. Patients were considered to have active disease if CRP or ESR was high with new symptoms or signs attributable to vascular ischemia in TA with or without constitutional symptoms or if ITAS-A was more than 4 [7].

All patients were subjected to percutaneous transluminal angioplasty after informed consent and explaining the procedure related benefits and risks. Transfemoral approach by accessing right common femoral artery was accessed under ultrasound guidance in all patients and 5F 11cm vascular sheath was secured. Intraarterial heparin was given at 70-100U/kg body weight. With the help of angiographic catheter (Headhunter (H1), Cook

Beacon tip 5F 100cm) and hydrophilic guide wire (Terumo 150 cm, 0.035"), all arch vessels (right brachiocephalic, left common carotid and left subclavian) were cannulated. After cannulating the arch vessels, an angiogram was done which revealed variable degree of stenosis in internal carotids, subclavian and vertebral arteries with multiple arterial collaterals. In patients with moderate narrowing, lesion was crossed with Terumo J-angled tip and straight-tip guide wire (0.035", 150 cm, 0.018"180 cm), followed by over-the-wire balloon angioplasty using Mustang balloon (Boston Scientific). In patients with severe/near total occlusion of arch vessels CTO guide wire (0.014",300 cm length) was used to cross the narrowing followed by plain balloon angioplasty. Serial balloon dilatation of affected artery was done with sterling monorail balloon (3 mm x 100 mm, 4 mm x 100 mm) (Figure 2).

Prophylactic atropine (0.5mg single dose) was given at the time of internal carotid artery angioplasty due to risk of bradycardia because of possible carotid sinus activation during angioplasty. Post-angioplasty angiogram shows optimal results with subsidence of all visible collaterals. Post procedure, patients were prescribed a single antiplatelet (Aspirin 75mg once daily for 15days) with overlap of injection Clexane 60mg subcutaneously for 5 days.

Results

Case 1

18-year-old female, who had left upper limb claudication for past 1 year duration presented with acute onset painless diminution of vision in both eyes and severe vertigo for 1 day duration. She appreciated h and motion on visual acuity testing at 2 meters distance. At presentation, her both brachial and radial pulses were not palpable. CT angiography of aorta and its branches showed stenosis of both proximal subclavian arteries, common carotid, intrarenal aorta, coeliac and superior mesenteric arteries. A diagnosis of TA was made and was started on IV methylprednisolone pulse therapy for 3 days. Her fundus examination, tonometry and slit-lamp examination were normal. FFA was reported to have delayed arm-to-retina circulation time of 19 sec (normal range 11-15 sec) and capillary non-perfusion areas. In view of acute vertebrobasilar insufficiency and visual symptoms not improving on pharmacological therapy, she underwent left subclavian, left internal carotid and right internal carotid artery percutaneous transluminal angioplasty (PTA) on the sixth day after onset of symptoms. The post-procedure period was uneventful and she started appreciating colour vision by the third day of procedure. There was no further worsening and she tested 20/80 BCVA in both eyes and ambulated without support by two weeks after procedure without any further vertigo episodes. Currently after 1 year of follow-up, she continues to maintain vision of 20/80 and without new/worsening symptoms.

Case 2

15-year-old female presented with episodic vertigo, transient vision loss and claudication of both upper and lower limbs of 2 years duration. Clinical examination revealed absence of bilateral radial, brachial, right popliteal and dorsalis pedis pulses. Her CT aortogram revealed type V TA with severe stenosis of both subclavian arteries, common carotid, renal and iliac arteries with >50% narrowing of abdominal aorta. She was on methotrexate at a dose of 25mg/week and tapering doses of prednisolone and was in clinical remission (ITAS-A less than 4) for few months. When dose of prednisolone reached less than 0.25mg/kg, she developed acute binocular vision loss with absent pupillary reflexes. Visual acuity testing revealed perception of h and motion in both eyes at 1 meter distance. CT Brain was normal, fundus examination revealed hypertensive retinopathy stage 2. FFA revealed delayed arm-to-retina circulation time (18.5 sec) and capillary non-perfusion areas. Her vision partially improved to 20/ 40 within 2 weeks in right eye after increasing steroid dose to 0.5mg/kg and addition of tocilizumab. Visual field charting using Humphrey field analyzer showed peripheral defects in both eyes. Due to persistent left eye symptoms (BCVA 20/ 200) and recurrent vertigo episodes, she was taken up for left common carotid, subclavian and right internal carotid PTA, following which her vision improved to a BCVA of 20/40 in both eyes and vertigo episodes subsided. Patient is on regular follow-up with us for the past 1 year and has no worsening or new complaints.

Case 3

14-year-old female presented with acute binocular painless vision loss with history of limb claudication. Examination revealed no light perception on visual acuity testing in both eyes with absent pupillary reflexes. There was absent left brachial and left radial pulses. CT angiography of the aorta and its branches revealed involvement of both subclavian, renal, iliac arteries and stenosis of more than 90% in bilateral common carotid arteries. Fundus examination showed cherry red spot, hyperemia of disc with blurred margins. FFA revealed increased foveal avascular zone, capillary non perfusion areas. She was started on IV methylprednisolone pulse for 3 days. There was no improvement in visual acuity, so we proceeded to manage with a bilateral carotid PTA. She developed seizures post-procedure and was managed conservatively. MRI Brain revealed diffuse T2 STIR hyperintensities in bilateral occipital and parietal lobes suggestive of ischemic reperfusion injury. She

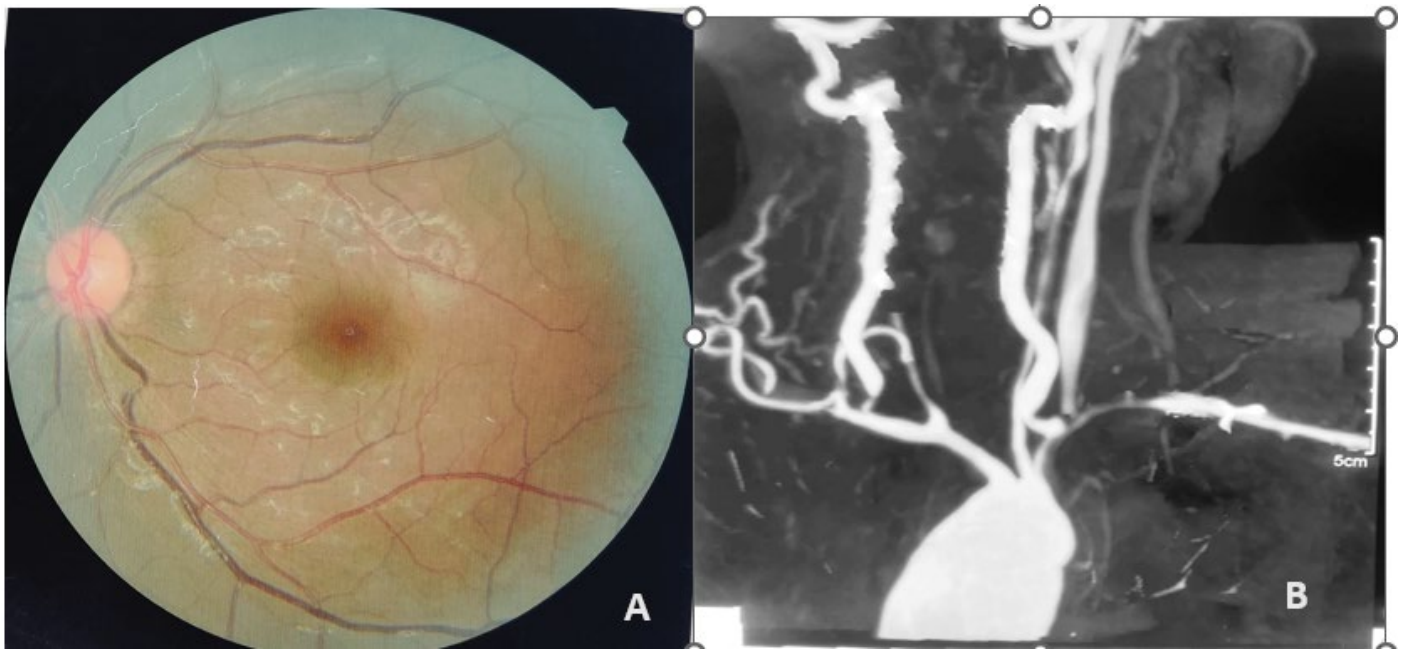

Figure 1 . (A) Fundus image of patient 3 with cherry red spot, hyperemia of disc with blurred margins and (**B)** CT Aortogram of patient 2 showing bilateral carotid and subclavian stenosis.

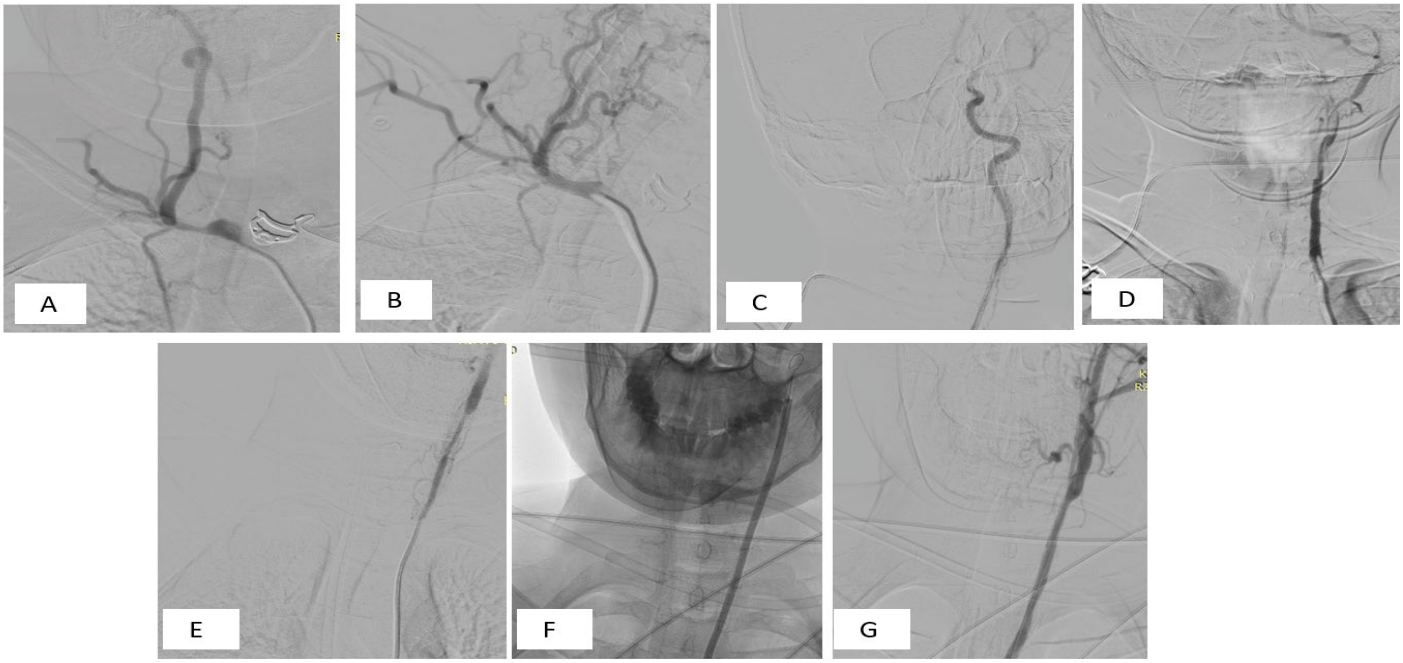

Figure 2. Shows (**A**) and (**B**) showing DSA images of filling of left subclavian artery of patient 1 pre- and post-PTA, (**C**) and (**D**) showing DSA images of filling of left internal carotid artery of patient 2 pre- and post-PTA and (**E**), (**F**) and (**G**) showing DSA images of filling of left internal carotid artery of patient 3 pre-, intra-procedure balloon in ICA and post-PTA.

Table 1. illustrating features of patients with OIS in TA.

S.No	Age	Duration(years)	Absent peripheral pulse	Hypertension	Acute phase reactants	CT aortogram involvement	TA type	Fundus angiogram findings	Fundus examination
1	18	1	Both brachial and radial	No	CRP and ESR High	Both proximal subclavian arteries, common carotid, Infrarenal aorta, coeliac and superior mesenteric arteries	V	capillary non perfusion areas, delayed arm-to-retina circulation time (19 sec)	Normal
2	15	1.5	Both brachial, radial, right popliteal, and dorsalis pedis.	Yes	CRP High	Both subclavian arteries, common carotid, renal and iliac arteries with >50% narrowing of abdominal aorta	V	capillary non perfusion areas, delayed arm-to-retina circulation time (18.5 sec)	Grade II Hypertensive retinopathy
3	14	1.5	Left Brachial and radial		CRP and ESR Normal	Both subclavian, renal, iliac arteries and >90% diffuse stenosis of bilateral common carotid arteries	V	increased foveal avascular zone, capillary non perfusion areas	Cherry red spot, hyperemia of disc with blurred margins

improved with conservative management and was discharged as there were no further seizure episodes. Her vision improved to identifying colours and appreciating h and motion at 1 meter distance at 10 days after procedure. It further improved to 20/ 80 BCVA in both eyes and is on follow-up for the past 3 months without any seizure episodes.

Discussion

All patients in our series had acute binocular sudden diminution of vision not completely responding to pharmacotherapy. A review by Malhotra, et al showed sudden onset vision loss in 41% of patients with OIS. OIS can rarely present with postpr andial or photic (on bright light) amaurosis also [8]. Fundus examination was normal in one patient, another showed Grade 2 hypertensive retinopathy with arterial narrowing and focal irregularities (ischemic changes) whereas the third patient had cherry rod spot at the macula with hyperemia of disc and mild blurring of disc margins suggesting possible anterior ischemic optic neuropathy (AION), although hyperemia was not characteristic of AION (Figure 1).

None of the patients showed neovascularization probably because the duration of symptoms of TA was less than 2 years. All patients had capillary non-perfusion areas in the fundus fluorescein angiography and 2 patients had increased arm-to-retina circulation time suggesting chronic hypoperfusive retinopathy. In a study on ocular manifestations in 78 patients with TA, delayed arm-to-retina circulation time was the most common finding in fundus fluorescein angiography while both arm-to-retina circulation time and presence of capillary non perfusion areas were seen commonly in our patients. Type V TA according to angiography findings was seen in all 3 patients. It affirmed previous knowledge that type V TA was the most common angiographic type in Indian cohort of TA patients with ocular manifestations [9,10] enlists the characteristics of our patients (Table 1).

Interventions in TA patients are indicated in setting of hypertension with renal artery stenosis, cerebral ischemia and/or symptomatic cerebral vessel occlusion, symptomatic aortic regurgitation, critical limb ischemia or any critical ischemia causing end-organ damage. Revascularization procedures in TA include endarterectomy, patch angioplasty, percutaneous transluminal angioplasty and surgical bypass. There have been conflicting suggestions in literature addressing optimal modality of revascularization in stenotic lesions of TA [11-13]. Malhotra, et al also noticed that patients with OIS had improvement in chronic ischemic changes in fundus angiography post carotid surgery. Saadoun, et al in his study, analyzed complications in 104 surgical bypass procedures and 62 endovascular repair procedures and they reported that outcomes were not statistically different between both the groups. They drew the conclusion that active vascular inflammation at the time of procedure increased the likelihood of complications by 7 times. Another study concluded that vascular surgery during active disease was the main risk factor contributing to mortality in TA patients [12,13]. A 10-year retrospective study about endovascular and surgical revascularization showed similar patency

rates and similar rate of complications in both revascularization procedures [14]. All our patients had active vascular inflammation as evidenced by ITAS-A (CRP) more than 4. Since the manifestations in these patients were acute and vision threatening, the revascularization procedure was performed on an urgent basis. In this series, we noticed good response in terms of visual outcome with percutaneous transluminal angioplasty in all three patients except for a single complication.

Conclusion

We suggest that combining vascular surgical and pharmacological intervention in acute vision loss in TA may restore vision, even in active disease. A longer follow-up will tell us about long term effects of angioplasty on visual outcome in TA. Visual outcome in OIS is poor unless there is timely intervention and there is scope of immediate improvement in patients with internal carotid disease who develop OIS without fundus evidence of neovascularization.

Conflict of Interest

Abstract published in EULAR abstract archive July 2022.

Statement of Ethics and Consent

Informed consent obtained from all patients included in the study.

References

1. Panja, Manotosh and P.C. Mondal. "Current status of aortoarteritis in India." (2004).
2. Peter, Jayanthi, Sarada David, George Joseph and Saban Horo, et al. "Hypoperfusive and hypertensive ocular manifestations in t akayasu arteritis." *Clin Ophthalm* 4 (2010): 1173.
3. Peter, Jayanthi, Sarada David, Debashish D anda and John Victor Peter, et al. "Ocular manifestations of Takayasu arteritis: A cross-sectional study." *Retina* 31 (2011): 1170-1178.
4. Lin, Xuemei, Jinxin Song, Wei Gao and Songdi Wu. "Acute ocular ischemic syndrome." *QJM: An Internat J Med* 110 (2017): 831-832.
5. Mathew, Ashish J, Ruchika Goel, Sathish Kumar and Debashish D anda. "Childhood-onset Takayasu arteritis: An update." *Internat J Rheum Dis* 19 (2016): 116-126.
6. Misra, Ramnath, Debashish D anda, Sivakumar M Rajappa and Alakendu Ghosh, et al. "Development and initial validation of the Indian Takayasu clinical activity score (ITAS2010)." *Rheumat* 52 (2013): 1795-1801.
7. Misra, Durga Prasanna, Neeraj Jain, Manish Ora and Kritika Singh, et al. "Outcome measures and biomarkers for disease assessment in Takayasu arteritis." *Diagnost* 12 (2022): 2565.
8. Malhotra, Raman and Kevin Gregory-Evans. "Management of ocular ischaemic syndrome." *Br J Ophthalmol* 84 (2000): 1428-1431.
9. Chun, Yeoun Sook, Seok Joon Park, In Kipark and Hum Chung. "The clinical and ocular manifestations of Takayasu arteritis." *Retina* 21 (2001): 132-140.
10. Hata, Akihiro, Makoto Noda, Ryutaro Moriwaki and Fujio Numano. "Angiographic findings of Takayasu arteritis: New classification." *Int J Cardiol* 54 (1996): S155-S163.
11. Tyagi, Sanjay, Prattay Guha Sarkar, Mohit D. Gupta and M. P. Girish. "Restoration of vision by endovascular revascularization in Takayasu arteritis: A case series." *J Cardiol Cases* 18 (2018): 123-127.
12. Rosa Neto, Nilton Salles, Samuel Katsuyuki Shinjo, Maurício Levy Neto and Rosa Maria Rodrigues Pereira. "Vascular surgery: The main risk factor for mortality in 146 Takayasu arteritis patients." *Rheumatol Int* 37 (2017): 1065-1073.
13. Saadoun, David, Marc Lambert, Tristan Mirault and Mathieu Resche Rigon, et al. "Retrospective analysis of surgery *vs*. endovascular intervention in Takayasu arteritis: A multicenter experience." *Circulation* 125 (2012): 813-819.
14. Diao, Yongpeng, Sheng Yan, Shyamal Premaratne and Yuexin Chen, et al. "Surgery and endovascular management in patients with Takayasu's arteritis: A Ten-Year Retrospective Study." *J Vasc Surg Cases* 63 (2020): 34-44.

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