

Case Report

Takayasu arteritis presenting as sudden onset vision loss simulates multiple sclerosis: a case report

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Abstract

Neurological manifestation may complicate Takayasu arteritis (TA). A 23-year-old girl with sudden onset of vision loss was admitted to hospital. Her brain MRI showed abnormal T2-signal hyperintensity and visual evoked potential revealed prolonged P100 latency. Consequently, optic neuritis was diagnosed. A review on history of dizziness, falling, and weak pulses of upper extremities led to more investigation. Angiography revealed a total occlusion of right and left carotids, left vertebral arteries, aneurismal dilatation of innominate artery and critical stenosis of right vertebral artery. Following diagnosis of TA, stenting of right vertebral artery was done, but she passed away because of subarachnoid hemorrhage.

KEYWORDS: Takayasu Arteritis, Neurologic Manifestations, Blindness.

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Takayasu arteritis (TA) is a chronic inflammatory and stenotic disease of medium and large vessels.¹ It affects predominantly the aorta and its major branches. In addition, TA involves coronary, renal and pulmonary arteries. The clinical manifestations of TA are divided in two phases as prepulseless and pulseless.^{2,3} During the early phase, symptoms such as malaise, fever, fatigue and arthralgia are generally constitutional and nonspecific. In this stage, there is no diagnostic serologic test. In the late phase, the ischemia occurred and the symptoms were secondary to arterial occlusion or aneurismal formation.^{2,4,5} The main clinical presentation of the late phase are pulseless extremities, hypertension, vascular bruits, congestive heart failure and pulmonary arterial involvement.^{2,5}

Neurological manifestations such as headache, stroke, hypertensive encephalopathy, etc. may complicate this disease.^{4,6} Moreover, some unusual neurological presentations of TA in-

clude recurrent epileptic seizures,³ spinal cord compression⁷ and posterior reversible encephalopathy syndrome (PRES).⁸

Here we report a clinical presentation of TA in a 23-year-old girl, at first presenting itself as a loss of vision that had been misdiagnosed as multiple sclerosis.

Case Report

A 23-year-old girl was admitted to hospital with sudden onset of loss of vision in both her eyes. Studying the details of the patient's history revealed episodes of weakness and dizziness since two years ago. She also experienced temporary clouding of sensorium and falling down on several occasions especially in the upright position just after a slight physical activity.

The patient's personal history was significant regarding the opium addiction. On admission, physical examination revealed an oral temperature of 37.2°C, a heart rate of 100 beats

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per minute, a respiratory rate of 18 breaths per minute, blood pressure in right upper arm was 150/100 mmHg and in left upper arm 170/100 mmHg, very weak pulses of the upper extremities and normal pulses of the lower ones. In neurological examination, the abnormal findings were decreased visual acuity and optic atrophy of left fundus. MRI of the brain with contrast agent showed bilateral abnormal T2 signals in white matter. Visual evoked potential (VEP) showed prolonged P₁₀₀ latency in both eyes and electroencephalography (EEG) was reported normal. Findings of performed hematological laboratory tests on patient are listed in table 1. Optic neuritis (ON) caused by multiple sclerosis (MS) was diagnosed and she received IV methylprednisolone 1 gr/day for 3 days. Her vision became better but attention on history of dizzy spell, physical findings of very weak pulses of upper limbs and difference between blood pressure of left and right arms made us investigate more on it. Radiological and sonographic findings of patient are listed in table 2. As noted in table 2, CT and conventional angiographies of chest and cervical arteries were the most diagnostic ones (Figure 1).

So diagnosis of TA was established and due to rheumatologic consultation, she received 1 gr/day IV methylprednisolone for 5 days, 500 mg IV cyclophosphamide in the next day and oral antiplatelet clopidogrel bisulphate (Plavix 75 mg/day).

This patient was a candidate for the stenting of the right vertebral artery which was recommended by an interventional cardiologist. Two

months later, the successful stenting of the right vertebral artery was done (Figures 2 and 3) and the patient was discharged from hospital with good general condition. One week later, she developed a very severe headache, repeated seizures and loss of consciousness. Brain CT scan showed intraventricular and subarachnoid hemorrhage (SAH). Ventriculo-peritoneal shunt was inserted for her, but it was unsuccessful and she passed away after a few days.



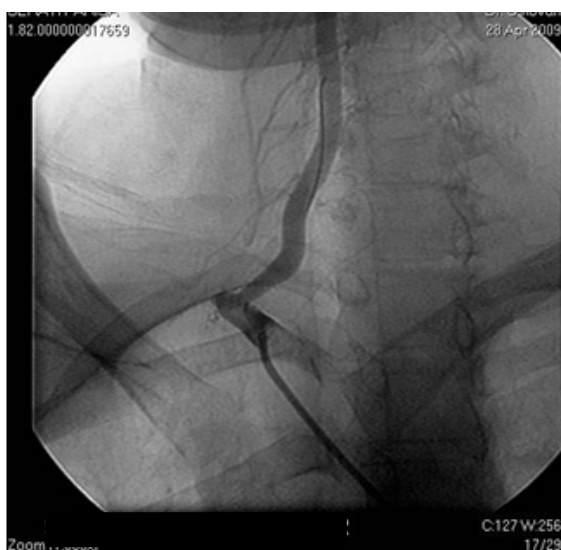
Figure 1. CT angiography of aortic arch and cervical vessels of the patient showed extensive stenosis of more cervical vessels, aneurysmal dilation of innominate artery

Table 1. Hematological findings of patient

Test	Result
Hemoglobin (Hb)	11.5 mg/dl
White blood cell (WBC)	9750 cells/mm
Erythrocyte sedimentation rate (ESR)	19 mm at the first hour
CRP	12 mg/dl
Platelet count	422000/mm
Viral markers (HBS-Ag, HCV-Ab, HIV-Ab)	Negative
Antinuclear antibody (ANA) titer	Slightly increased
Antidouble strength antibody (Anti Ds DNA)	Negative
Anticardiolipin antibody (ACLA)	Negative
VDRL	Negative
Antibrucella antibody	Negative

Table 2. Radiological and sonographic findings of patient

Test	Result
Transesophageal echocardiography (TEE)	Diffuse intimal thickening in aorta, dilated ascending aorta suggesting aortitis
CT and conventional angiographies of chest and cervical arteries	Total occlusion of the right and left carotids, total occlusion of left vertebral artery, aneurismal dilation of innominate artery, severe stenosis of right vertebral artery
Color doppler sonography of four limbs	No flow in axillary, brachial, ulnar and radial arteries bilaterally Normal flow in lower limbs arteries
Color doppler sonography of renal arteries	Normal flow in bilateral renal arteries

**Figure 2.** Stenting of right vertebral artery in the patient**Figure 3.** Stenting of right vertebral artery in the patient**Table 3.** The American College of Rheumatology criteria for the diagnosis of Takayasu arteritis⁹

Criterion	Definition
Age at disease onset must be less than 40 years	Development of symptoms or findings related to Takayasu's arteritis at age < 40 years
Claudication of limbs	Development and worsening of fatigue and discomfort in muscles of 1 or more limb while in use, especially the arms
Decreased brachial arterial pulse	Decreased pulsation of one or both brachial arteries
Blood pressure difference greater than 10 mmHg	Difference of more than 10 mmHg in systolic blood pressure between arms
Bruit over subclavian arteries or aorta	Bruit audible on auscultation over 1 or both subclavian arteries or abdominal aorta
Arteriographic abnormality	Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal limbs, not caused by arteriosclerosis, fibromuscular dysplasia, or similar causes; changes usually focal or segmental

Discussion

Takayasu arteritis is an inflammatory aortitis which mainly affects young females in the age group of 10-30 years^{5,10}; its male to female ratio is 1 to 8.5.¹¹ It is more prevalent in Asians, but it has a widespread distribution.^{5,12,13}

The occurrence of neurological manifestations was reported in 52.7% of patients with TA.^{2,6} Headache was the most common symptom (55%).⁶ Major neurological events occurred in about one-half of the patients were TIA, cerebral infarction, hypertensive encephalopathy, seizure, paraplegia and even Moya-Moya phenomenon.^{6,14} These events are related to a combination of carotid and vertebral artery disease and the complication of hypertension and thromboembolism would lead to stroke.^{2,7}

Differential diagnosis of TA are other causes of large vessels vasculitis including infectious aortitis (syphilis, mycobacterial, fungal), autoimmune causes (systemic lupus, Behcet's disease, giant-cell arteritis and Kawasaki disease) and some developmental abnormalities (coarctation of aorta and marfan's syndrome),^{2,5,15} most of which have clinical presentation or specific laboratory findings, however, they were not found in our patient.

Although sarcoidosis may have clinical and radiological presentation similar to TA,¹⁵ lack of some specific features such as skin lesion and hilar lymphadenopathy ruled it out.

Diagnosis of TA needs at least 3 of 6 criteria of the American College of Rheumatology (Table 3).⁹ Our patient's criteria were being 23 years old, having pulseless arms, having different blood pressure between two arms (20 mmHg) and arteriographic findings of stenosis in aortic arc with its main branches (Tables 1 and 2). The presence of 3 or more of these 6 criteria demonstrates 90.5% and 97.8% sensitivity and specificity, respectively.⁹

Optic neuritis caused by MS was a misdiagnosis in our patient and combinations of visual loss, abnormal VEPs, and white matter signals of MRI were misleading. Our case had no MS clinical and MRI criteria. MRI criteria of McDonald supports clinically definite diagnosis of MS¹⁶ which needs evidences of at least two episodes of neurological deficits and phys-

ical findings of two sites involving in central nervous system. However, pulsetherapy by methylprednisolone is the best treatment of optic neuritis.

In our patient, two main arterial systems of central nervous system (vertebrals and carotids) were involved. Vision loss was attributed to hypoperfusion of optic nerves and retina. Abnormal white matter T2 signals of MRI were ischemia in watershed zones secondary to severe hypoperfusion in carotids and vertebral arteries circulation. Dizziness and falling episodes were secondary to hypoperfusion of brain stem structures. Classical ophthalmic features of TA are because of the reduced ocular perfusion.² The occlusion of retinal artery branches has also been reported.¹⁷ Anterior ischemic optic neuropathy (AION) is rare in TA.¹²

Treatment of TA is divided into medical and surgical or endovascular therapy. Glucocorticoid therapy is the first line in medical treatment. Second-line drugs, including cyclophosphamide, azathioprine and methotrexate, should be taken in refractory cases or in patients with recurrence of disease activity.^{2,5} Antiplatelet or anticoagulant agents prevent thromboembolic events. Surgical or endovascular therapy of TA has been performed in critical stenosis of renal, coronary or cerebral vessels. Clinical features of cerebrovascular ischaemia or critical stenosis of at least three cerebral vessels need interventional procedures.² Although angioplasty is not successful in total occlusions, using it opens short-segment occlusions.¹⁸ In spite of high initial success rate of angioplasty in TA, there are some different reports on long-term restenosis rate.^{19,20} Our patient was treated by IV methylprednisolone, cyclophosphamide and oral clopidogrel.

It seemed that performing angioplasty with stenting of right vertebral artery as the only perfusing brain vessel was lifesaving. We did it but she passed away after stenting and it might be because of SAH resulting from hyperperfusion phenomenon.

Conclusions

TA is an uncommon stenotic arterial disease. Some studies reported that it may have un-

usual neurologic presentations such as seizure, myelitis, and PRES. Our patient with hyperintense white matter lesions in brain MRI and vision loss was found as an example of TA which simulates MS.

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Conflict of Interests

Authors have no conflict of interests.

Authors' Contributions

NA carried out the design, coordinated the study, participated in most of the experiments and prepared the manuscript. SS gathered patient data and history. MAO participated in manuscript preparation. All authors have read and approved the content of the manuscript.

References

1. Johnston SL, Lock RJ, Gompels MM. Takayasu arteritis: a review. *J Clin Pathol* 2002; 55(7): 481-6.
2. Tann OR, Tulloh RM, Hamilton MC. Takayasu's disease: a review. *Cardio Young* 2008; 18(3): 250-9.
3. Ioannides MA, Eftychiou C, Georgiou GM, Nicolaides E. Takayasu arteritis presenting as epileptic seizures: a case report and brief review of the literature. *Rheumatol Int* 2009; 29(6): 703-5.
4. Vanoli M, Bacchiani G, Origg L, Scorza R. Takayasu's arteritis: a changing disease. *J Nephrol* 2001; 14(6): 497-505.
5. Maffei S, Di Renzo M, Bova G, Auteri A, Pasqui AL. Takayasu's arterities: a review of the literature. *Intern Emerg Med* 2006; 1(2): 105-12.
6. Wang JZ. Neurological manifestation of Takayasu's arteritis. *Zhonghua Shen Jing Jing Shen Ke Za Zhi* 1992; 25(6): 369-71, 385-6. (Chinese).
7. Kim HA, Kim JH, Won JH, Suh CH. An unusual clinical manifestation of Takayasu's arteritis: spinal cord compression. *Joint Bone Spine* 2009; 76(2): 209-12.
8. Karande S, Jagtap S, Joshi A. Posterior reversible encephalopathy syndrome revealing Takayasu's arteritis. *Indian J Pediatr* 2009; 76(2): 218-20.
9. Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. *Arthritis Rheum* 1990; 33(8): 1129-34.
10. Singh S, Bali HK, Salaria M, Lal R, Pandav SS, Kumar L. Takayasu's arteritis in young children: a potentially treatable condition. *Indian Pediatr* 1999; 36(3): 291-6.
11. Lupi-Herrera E, Sánchez-Torres G, Marcushamer J, Mispireta J, Horowitz S, Vela JE. Takayasu's arteritis. Clinical study of 107 cases. *Am Heart J* 1977; 93(1): 94-103.
12. Liang P, Hoffman GS. Advances in the medical and surgical treatment of Takayasu's arteritis. *Curr Opin Rheumatol* 2005; 17(1): 16-24.
13. Malik KP, Kapoor K, Mehta A, Dadeya S, Guliani BP, Aggarwal S, et al. Bilateral anterior ischemic optic neuropathy in Takayasu arteritis. *Indian J Ophthalmol* 2002; 50(1): 52-4.
14. Kerr GS, Hallahan CW, Giordans J, Leavitt RY, Fauci AS, Rottem M, et al. Takayasu arteritis. *Ann Intern Med* 1994; 120(11): 919-29.
15. Saab F, Giugliano RP, Giugliano GR. Takayasu arteritis in a young woman: a 4-year case history. *Tex Heart Inst J* 2009; 36(5): 470-4.
16. Polman CH, Reingold SC, Edan G, Filippi M, Hartung HP, Kappos L, et al. Diagnostic criteria for multiple sclerosis: 2005 revisions to the "McDonald Criteria". *Ann Neurol* 2005; 58(6): 840-6.
17. Kaushik S, Gupta A, Gupta V, Jain S, Lal V. Retinal artery occlusion in Takayasu's arteritis. *Indian J Ophthalmol* 2005; 53(3): 194-6.
18. Tyagi S, Verma PK, Gambhir DS, Kaul UA, Saha R, Arora R. Early and long-term results of subclavian angioplasty in aortoarteritis (Takayasu disease): comparison with atherosclerosis. *Cardiovasc Intervent Radiol* 1998; 21(3): 219-24.
19. Liang P, Tan-Ong M, Hoffman GS. Takayasu's arteritis: vascular interventions and outcomes. *J Rheumatol* 2004; 31(1): 102-6.
20. Sharma S, Saxena A, Talwar KK, Kaul U, Mehta SN, Rajani M. Renal artery stenosis caused by nonspecific arteritis (Takayasu disease): results of treatment with percutaneous transluminal angioplasty. *AJR Am J Roengenol* 1992; 158(2): 417-22.