

An unusual case of Takayasu's arteritis: Evaluation by CT angiography

Mukund Vidhate, Ravindra Kumar Garg, Rajesh Yadav¹, Neera Kohli¹, Praveen Naphade, H. K. Anuradha

Departments of Neurology and ¹Radiodiagnosis, Chhatrapati Shahuji Maharaj Medical University, Lucknow, Uttar Pradesh, India

Abstract

Takayasu's arteritis is a chronic, idiopathic, medium and large vessel vasculitis involving aorta and its main branches. Frequent neurological manifestations include postural syncope, seizures, and blindness. Stroke, as presenting feature of Takayasu's arteritis, is unusual. CT angiography reveals characteristic involvement of aortic arch and its branches. Involvement of intracranial vasculature is rather unusual. We are describing an unusual patient of Takayasu's arteritis who presented with recurrent disabling syncopal attacks and had extensive involvement of intracranial vasculature. CT angiography revealed severe involvement of aortic arch. There was near complete occlusion at origins of both subclavian arteries, distal flow was maintained by collateral vessels along the chest wall. There was near total occlusion (at origin) of right common carotid with normal flow in distal part. The left common carotid was more severely involved showing greater than 80% narrowing in proximal half of the vessel. CT angiography also revealed involvement of left internal carotid artery, narrowing of left middle cerebral artery and involvement of cortical vessels. Patient was treated with oral corticosteroids. She improved remarkably after two and half months of follow up.

Key Words

Takayasu's arteritis, vasculitis, stroke

For correspondence:

Dr. Ravindra Kumar Garg, Department of Neurology, Chhatrapati Shahuji Maharaj Medical University, Lucknow - 226 003, Uttar Pradesh, India. E-mail: garg50@yahoo.com

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Introduction

Takayasu's arteritis is an idiopathic chronic inflammatory disease of unknown etiology, pathologically characterized by granulomatous changes of the aorta and its major branches.^[1] Takayasu's arteritis has been reported worldwide; it is more common in young Asian women. Females in the second or third decade of life are at greatest risk for this disease.^[2]

In Takayasu's arteritis, in fact, is a panarteritis, beginning with inflammation of the adventitia with subsequent involvement of the media and intima. The characteristic pathological features include arteritis wall vessel inflammation leads to thickening, fibrosis, segmental stenosis, and aneurysm and thrombus formation. The exact etiopathogenesis is unknown.^[1,2]

Conventional angiography demonstrates luminal irregularity, vessel stenosis, occlusion, dilatation or aneurysms in the aorta or its primary branches. Computed tomography angiography is a non-invasive technique capable of demonstrating mural and luminal changes in both the aorta and aortic branches. Computed tomography angiography has a greater diagnostic accuracy than conventional angiography.^[3]

Neurologic involvement frequently occurs and sometimes, this may be the initial presentation of the disease. The disease may have varied neurological manifestations. Patients may present with headache, syncope, and blindness. Involvement of intracranial vasculature is rather unusual.^[4] Cerebral ischemia can give rise to devastating neurological symptoms. Approximately 10–20% of patients with Takayasu's arteritis are likely to have cerebrovascular accidents.^[5] Occlusion of the vertebral or carotid arteries may cause ischemic stroke. Patients with Takayasu disease may also develop intracranial aneurysms.^[4]

Orthostatic syncope may, at times, be disabling.^[6] We are describing an unusual patient of Takayasu's arteritis who presented with recurrent disabling syncopal attacks and had extensive involvement of intracranial vasculature.

Case Report

A 24-year-old housewife, presented with recurrent episodes

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of loss of consciousness of 1 month duration. These episodes used to occur with change of posture and were common in sitting upright and on standing. Patient was not able to sit for more than few seconds. There was no history suggestive of seizures. Patient had history of stroke approximately a year back. The patient did have fever, joint pain, rash or other systemic symptoms.

On examination, all the peripheral pulses of upper limbs and neck (bilateral carotid, superficial temporal, brachial and radial) were absent. Pulses in lower limbs were palpable normally. Blood pressure, measured in lower limbs, was 130/80 mm Hg. Neurologic examination revealed Broca's aphasia, right hemiparesis (power of Medical Research Council grade-1 in upper limb and 4 in lower limb), generalized hyperreflexia and extensor plantar response on right side. Optic fundi were normal. Cardiac examination was normal.

Hematological evaluation revealed hemoglobin of 10 g/dl, total leukocyte count 6800/cmm with 65% polymorphs and 25% lymphocytes. Erythrocyte sedimentation rate was 22 mm at 1 hour. C-reactive protein level was 11 mg/L (Normal upper limit 5 mg/L). All other serum biochemical parameters were normal. Prothrombin time and activated partial thromboplastin time were within normal limits. tests for human immunodeficiency virus antibodies, hepatitis C virus antibodies and Australia antigen were also negative. Chest X-ray, electrocardiogram, echocardiography, electroencephalography were normal. CT brain revealed an old infarct in left middle cerebral artery territory. CT angiography revealed severe involvement of aortic arch. There was gross thickening of wall of arch of aorta and proximal ascending aorta. There was no aneurysmal dilatation of aorta. There was near complete occlusion (at origin) of both subclavian arteries, distal flow was maintained by collaterals along chest wall. There was significant (near total) occlusion at origin of right common carotid with normal flow in distal part [Figures 1 and 2]. The left common carotid was more severely involved showing >80% narrowing in proximal half with involvement of left internal carotid artery [Figure 2]. Angiography of intracranial vessels revealed significant narrowing of left middle cerebral artery mainstem as well as cortical vessels [Figure 3].

Patient was treated with corticosteroids (prednisone 60 mg/day) and aspirin. Patient improved considerably after two and half months of follow up. At present patient is independent of all her daily activities. Now patient is receiving prednisone 60 mg every alternate day.

Discussion

Our patient fulfilled American College of Rheumatology criteria for Takayasu's arteritis [Table 1].^[7] She had type-I (as per angiographic classification proposed by the International Cooperative Study on Takayasu's arteritis) disease showing severe involvement of aortic arch and its branches.^[8]

Our patient, at the time of presentation, had disabling orthostatic syncope making it impossible for her to sit. Patient had critical stenosis at origin of both carotids possibly, causing significant fall in cerebral perfusion pressure while

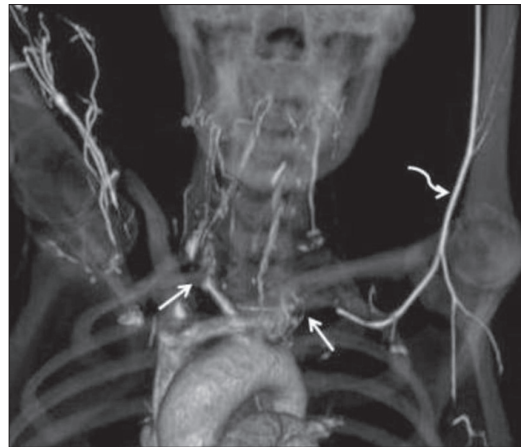


Figure 1: Three dimensional CT aortic angiography showing total occlusion of origin of bilateral subclavian arteries (arrows), distal flow established by collaterals



Figure 2: Three dimensional CT angiography showing occlusion at origin of right common carotid artery with normal flow in distal extracranial and intracranial portion of right internal carotid artery (arrows); left common carotid artery showing involvement of both proximal and distal extracranial portion (arrowheads)

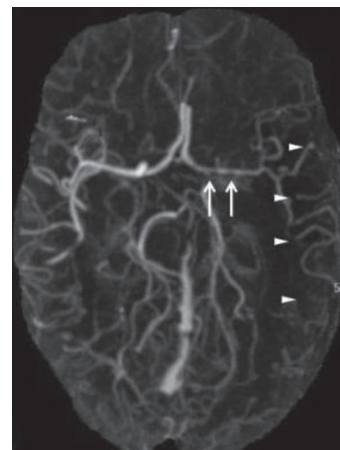


Figure 3: CT angiography at circle of Willis showing irregular narrowing of M₁ portion of left middle cerebral artery (arrows) with decreased filling of left sided cortical vessels (arrowheads). Left posterior cerebral artery was normal

Table 1: Diagnostic criteria for Takayasu arteritis^[6] (The presence of 3 or more of these 6 criteria)

Onset at age less than or equal to 40 years
Claudication of an extremity
Decreased brachial artery pulse
Greater than 10mm Hg difference in systolic blood pressure between arms
A bruit over the subclavian arteries or the aorta
Arteriographic evidence of narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities

she attempted to sit upright. Intracranial hemodynamics in these patients is characterized by dampened or blunted spectra with low flow pulsatility observed by transcranial Doppler study. This phenomenon occurred mainly when both carotid arteries were affected, was also more prominent when the vertebral arteries were involved. There is altered cerebral hemodynamics and metabolism in patients of Takayasu's arteritis with neurologic symptoms.^[9]

Our patient also suffered a major stroke before we made the diagnosis of Takayasu's arteritis. Approximately 10–20% of patients with Takayasu's arteritis have ischaemic stroke or transient ischaemic attacks. Intracranial stenoses in Takayasu's arteritis could be due to either vasculitic involvement or a prior embolization into the vessel. Other postulated mechanisms for stroke in Takayasu's arteritis are stenocclusive extracranial vessels, hypertension and premature atherosclerosis.^[10,11] Intracranial involvement in Takayasu's arteritis may be under-recognized as evaluation of intracranial vasculature had infrequently been performed.^[5,12] In our patient there was multiple small segmental narrowing of left middle cerebral artery suggestive of probable vasculitic involvement. Vertebral arteries were visualized normally.

The patients of Takayasu's arteritis require long term immunosuppressive therapy to control vessel wall inflammation. The immunosuppressive therapy includes glucocorticoids, which are often used in combination with other immunosuppressive agents, such as methotrexate, azathioprine, or mycophenolate mofetil. The remission of disease is defined as the absence of new arterial lesions, lack of systemic symptoms, and ability to taper prednisone to less than 10 mg/d.^[13] Our patient responded very well to prednisolone therapy. Certain patients, who do not respond to immunosuppressive therapy, may require surgical intervention. Revascularization procedures for the surgical treatment of Takayasu's arteritis include bypass grafting, angioplasty, endarterectomy, transluminal angioplasty and stenting.

Diagnosis of Takayasu's arteritis was established nearly one year after stroke as simple clinical examination of peripheral pulses was missed till patient presented to us with progressive cerebral ischemia and disabling orthostatic hypotension. Hence we again emphasize the importance of clinical examination of peripheral pulses in evaluating stroke in young.

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