


CASE REPORT

Ischemic stroke as the first presentation of takayasu's arteritis in young male

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Abstract

Takayasu's arteritis should be kept under the differential diagnosis of stroke in all young patients. Early, proper diagnosis and treatment are necessary to reduce any further progression, morbidity, and mortality rates of the disease.

KEY WORDS

cerebrovascular accident, ischemic stroke, takayasu's arteritis, vasculitis

1 | INTRODUCTION

Takayasu's arteritis (TA) is a chronic inflammatory disease of unknown cause characterized by granulomatous vasculitis of medium and large arteries, principally the aorta and its branches. TA also tends to affect women more often, with a ratio of 1.2-29:1. Neurological involvement may occur in about 50% of patients, and the occurrence of acute stroke as the initial presentation in patients with Takayasu's arteritis is rarely reported. Herein, we report a case of a 26-year-old Nepali male who was presented with acute weakness in the left limbs and aphasia after 3 days history of right elbow fracture open reduction, internal fixation surgery, and insertion of Kirschner wires (K-wires). Asymmetric arterial blood pressure of the upper and lower extremities and bruits over subclavian arteries were observed. Imaging studies revealed lenticulostriate infarction, narrowing, and inflammatory changes of the carotid arteries and aorta. Takayasu's arteritis presented with acute ischemic stroke was diagnosed, and the patient was started on steroids and oral methotrexate with good clinical response.

Takayasu's arteritis, also known as pulseless disease or occlusive thromboaropathy, is a granulomatous vasculitis of unknown etiology that primarily affecting the aorta and its major branches.¹

It is an estimated annual incidence rate of 1 case per 1 million persons annually. It is mainly affecting young women (9:1 ratio) with a typical age at onset between 15 and 25 years. It has a worldwide distribution, being rare in North America but more prevalent in the Far East.²

Inflammatory injury to the vessel wall leads to patchy disappearance of the elastica and smooth muscle layer and subsequent intimal hyperplasia, resulting in vascular stenosis in virtually all patients and dilatation and aneurysm in about 25%.² Neurological involvement in Takayasu's arteritis occurs in about half of all cases but stroke occurs in only 10% of patients with Takayasu's arteritis.³

Here, we are reporting a case of acute ischemic stroke as a first presentation of Takayasu's arteritis in a young male who presented with weakness of the left half of the body with motor aphasia. This study aimed to highlight the need to complete the evaluation of the underlying etiology of young

patients with stroke to initiate the therapeutic plan to reduce the chance of second events and to promote appropriate follow-up to improve outcomes.

2 | CASE REPORT

A 26-year-old Asian male from Nepal developed an acute onset of left-sided weakness and aphasia, after 3 days history of a right elbow fracture open reduction and internal fixation surgery.

On examination, he was conscious, afebrile. Blood pressure in the right upper limb was 125/89 mm Hg and left upper limb 102/75 mm Hg. Blood pressure recorded in the lower limbs—176/91 on both sides. There was a bruit over both subclavian arteries. His left arm was cold with weak pulses. No abnormality was detected on the chest and abdominal examination.

Neurological examinations revealed left dense hemiplegia (power grade 1/5), left hyperreflexia, and extensor plantar response on the left side. Cranial nerve examination was unremarkable.

A brain CT scan showed extensive hypodensity within the right temporoparietal region, representing the right lenticulostriate infarction (Figure 1).

Echocardiography revealed a normal heart structure with normal LV functions. Duplex Doppler study of carotid arteries shows a long segment of uniform circumferential wall thickening of both common carotid arteries causing 96% stenosis on the right and 92% stenosis on the left. The wall thickening is extending to the brachiocephalic trunk, the origin of both subclavian arteries. The carotid bulbs and internal and external carotid arteries have normal wall thickness with abnormal damped flow. The waveform of the left CCA was biphasic and of moderate peak systolic velocity (50 cm/s) compared to a more damped monophasic waveform of the right CCA which has a peak velocity of 20 cm/s (Figure 2).

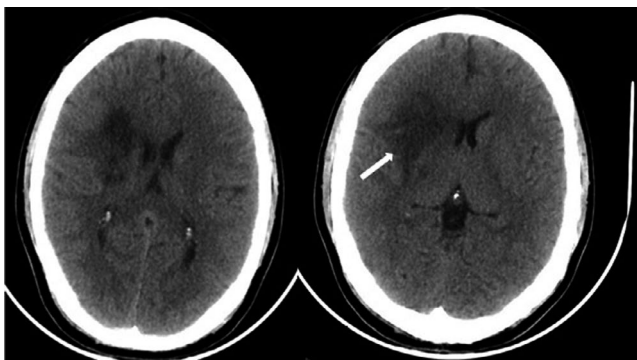


FIGURE 1 Non enhanced axial CT scan brain showing right frontoparietal hypodense area of infarction at the territory of the middle cerebral artery (arrowed)

CT angiography revealed concentric mural thickening of aortic arch branches around their origins extending to involve both CCAs causing thin linear interrupted filling of their lumen. The carotid divisions up to the circle of Willis are well opacified. Both subclavian arteries show stenosis of their proximal aspect and distal filling. There are no mural calcifications, no perivascular inflammatory changes, no aneurysm, or collateral vessels. The descending thoracic, abdominal aorta and its branches are of normal caliber and filling (Figure 3).

Baseline investigations revealed raised erythrocyte sedimentation rate of 115 mm 1st hour, serum C-reactive protein level of 24 mg/dL and mild normocytic normochromic anemia. Homocysteine, protein C, protein S, antithrombin III, and antiphospholipid antibodies were within normal limits.

Based on the clinical findings and the aortogram abnormalities, the patient was diagnosed as Takayasu's arteritis. The patient was treated high-dose prednisolone as well as oral methotrexate 20 mg/wk, acetylsalicylic acid 200 mg/d, and simvastatin 20 mg/d. The patient afterward shows significant clinical recovery with normalization of ESR and CRP at the end of hospitalization.

3 | DISCUSSION

Takayasu's arteritis is an idiopathic, inflammatory disorder affecting the aorta and its major branches. The name comes after a Japanese ophthalmologist, Dr Mikito Takayasu, who in 1908 first reported characteristic symptoms of this disease.⁴

Takayasu's arteritis is commonly affecting women, with a female to male ratio of approximately 8:1. The incidence varies worldwide, with Japan reporting an incidence of approximately 150 cases per million per year. In the United States, the incidence falls to approximately two cases per million per year.⁵

Although numerous genetic and environmental causes have been investigated, etiologic risk factors for Takayasu's arteritis are not well understood.⁶ The disease involves aorta and its branches, rarely including pulmonary or smaller arteries (eg, coronary). The time from first clinical symptoms to diagnosis varies and is usually 2-36 months (mean 24 months).⁴

Most patients develop nonspecific generalized symptoms such as malaise, fever, night sweats, arthralgias, anorexia, and weight loss, which may occur months before vessel involvement is apparent. Later in the course of the disease, patients often present with hypertension and contribute to renal, cardiac, and cerebral injury.⁷

About half of the patients of Takayasu's arteritis can have neurological symptoms, the most common being visual symptoms. Strokes occur in 10% of the patients.³ There are many possible mechanisms of stroke in Takayasu's arteritis

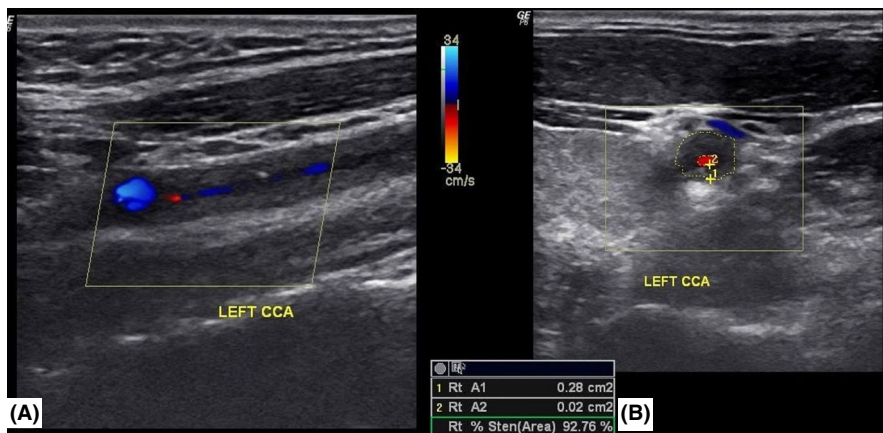


FIGURE 2 Color flow mapping of the left CCA shows same changes as the right (A) with 92% stenosis (B)



FIGURE 3 AP and oblique 3D computed tomography angiogram showing stenosis and filling defects of both CCAs (arrowed), noted the venous contamination where filling of the dural sinuses, jugular veins and superior vena cava due to a slight delay in shooting

which include embolism of stenotic or occlusive lesions of the aorta and its branches, hypertension, cardioembolism, and cerebral hypoflow.⁶

In 1990, the American College of Rheumatology has developed a set of criteria for the diagnosis of Takayasu's arteritis:

1. Age of onset < 40 years
2. Claudication of extremities.
3. Decreased brachial artery pressure.
4. Blood pressure difference > 10 mm Hg
5. Bruit over subclavian arteries and aorta.
6. Aortogram abnormalities.

At least 3 of the above 6 criteria are to be met for the diagnosis and demonstrate a sensitivity of 90.5% and a specificity of 97.8%.⁸

Standard treatment for Takayasu's arteritis is high-dose steroids and other immunomodulators, such as methotrexate, cyclophosphamide, and azathioprine. Patients with structural damage to affected vessels may require angioplasty to improve blood flow or arterial bypass or reconstruction in severe cases.^{8,9}

The course of the disease is variable; most patients have a chronic relapsing and remitting course. Survival rates have increased greatly recently so that 10-year survival rates of 80%-90% have become common. Disease-related mortality most often occurs from heart failure, stroke, myocardial infarction, aneurysm rupture, renal failure, or infectious complications of immunosuppressive treatment.¹⁰

Our patient had no history of generalized manifestations like malaise, fever, night sweats, arthralgias, anorexia, and weight loss. Acute ischemic stroke was the first clinical presentation. Our patient met 5 of the 6 of the criteria and was therefore diagnosed as Takayasu's arteritis. The patient showed a good response to high-dose prednisolone and oral methotrexate, with a return of motor strength on the left side to grade 4/5 and a mild Broca's aphasia. He was discharged with the plan for interval CT angiograms, ESR, and CRP for monitoring of the progression of the disease; also, he was referred to neuro-rehabilitation center for further rehabilitation.

CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest.

AUTHOR CONTRIBUTIONS

All authors were involved in drafting the article or revising it, and all authors approved the final version to be published.

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