

CASE REPORTS

Stroke as an initial presentation of Takayasu's arteritis

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Abstract

Takayasu's arteritis is an idiopathic inflammatory disease involving the main vessels. Acute stroke as the initial presentation in patients with Takayasu's arteritis is rarely reported. Herein, we report the case of a 36-year-old woman who complained of acute weakness in the left limbs. The lack of a pulse in the distal artery and the difference in blood pressure between limbs were observed in the emergency department. Takayasu's arteritis complicated with acute ischemic stroke was diagnosed based on the angiographs. The relevant literature is also reviewed.

INTRODUCTION

Takayasu's arteritis is a chronic inflammatory vasculopathy of unknown etiology, and mainly affects the aorta, its large branches and the pulmonary arteries.¹ It is most common seen in Japan, Southeast Asia, India and Mexico. It predominantly affects women in their second to third decade of life.^{1,2} During the progressive phase with fibrosis and thickening of the arterial wall, cerebrovascular accidents including transient ischemic attack and stroke occur in 10 to 20% of these patients.³ However, stroke as an initial presentation of Takayasu's arteritis has rarely been reported in the literature.^{4,5} Herein, we report a case of stroke as an initial presentation of Takayasu's arteritis to heighten the awareness as a cause of young stroke.

CASE REPORT

A 36-year-old female developed an acute onset of dizziness and weakness in the left extremities. At our Emergency Department, her consciousness was clear. Her body temperature was 37.2 degrees Celsius; pulse rate 76 per minute; and respiratory rate 18 per minute. Blood pressure in the right arm was 74/59 mmHg; in the left arm, 74/61 mmHg; in the right leg 149/70 mmHg; and in the left leg, 120/66 mmHg. Arterial pulse was not palpable in either arm. There was no bruit at the carotid artery. Neurological examinations revealed the

muscle strength of the left extremities at grade 4+ over 5.

Laboratory examinations showed the white blood cell count was 7560 per cubic millimeter with normal differential count. Platelet count was also normal. Hemoglobin was 8.9 g per deciliter and hematocrit was 28.2%. Platelet count was 454,000 per cubic millimeter. Erythrocyte sedimentation rate (ESR) was 27 mm per hour. C-reactive protein was 1.040 mg per deciliter. Her anti-nuclear antibody was positive. C3 and C4 levels were 112 mg/dl and 19.6 mg/dl, respectively. Antithrombin III, protein C, protein S and rheumatic antigen were within normal limits.

The computed tomography (CT) scan of the brain revealed focal mild hypodensity with effacement of sulci in the right hemisphere and persistent hyperdensity at the right middle cerebral artery (Figure 1A). No obvious midline shift was noted. The subsequent magnetic resonance image (MRI) of the brain confirmed the diagnosis of acute cerebral infarction in the right middle cerebral artery territory (Figure 2A). The MR angiography revealed narrowing of the lumens of the right brachiocephalic and proximal left common carotid artery (Figure 2B). There was occlusion of the left subclavian artery, right common carotid artery and middle cerebral artery. The transcranial Doppler study revealed near total occlusion of the right common carotid

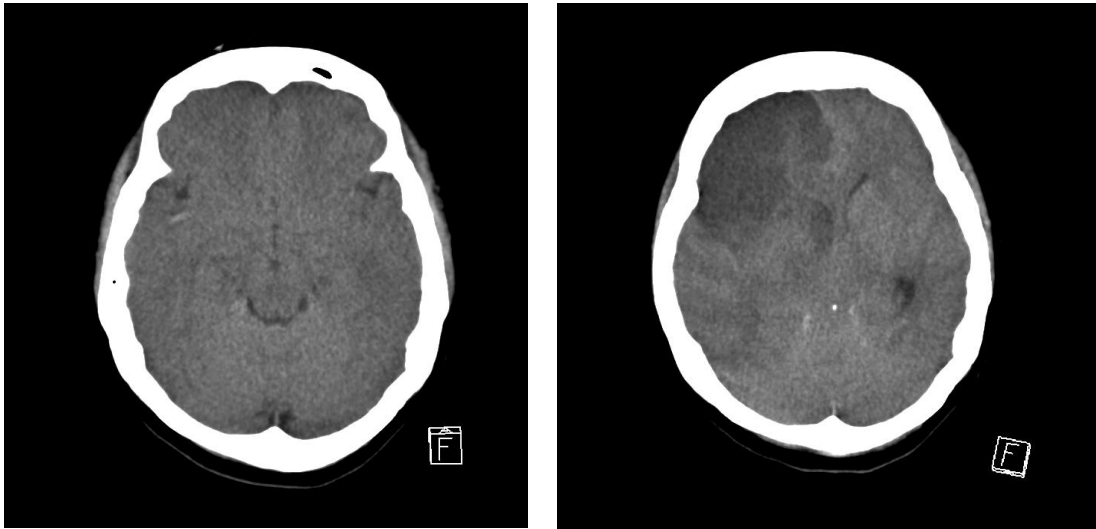


Figure 1. Computed tomography scan of the brain revealed hyperdensity at the right middle cerebral artery at admission (A) and areas with marked hypodensity in the right hemisphere with an obvious midline shift five days after admission (B).

artery, severe stenosis of the right brachiocephalic artery and left subclavian artery with compromised hemodynamics in their branches. Conventional digital subtraction angiography revealed marked stenosis of the brachiocephalic trunk and left subclavian artery, which affected the patency of both common carotid arteries.

The patient had sudden deterioration of consciousness, increased weakness in the left limbs and a dilated right pupil 5 days after admission. Marked cerebral infarction in the

right hemisphere with an obvious midline shift was seen in the follow-up CT scan (Figure 1B). An emergency right decompressive craniectomy was performed. Her clinical symptoms improved gradually during the next 10 days. Carotid stenting was performed to prevent progressive occlusion of the main arteries. Steroid was prescribed to manage the inflammation status. The patient had clear consciousness and left hemiparesis during the one-year follow-up.

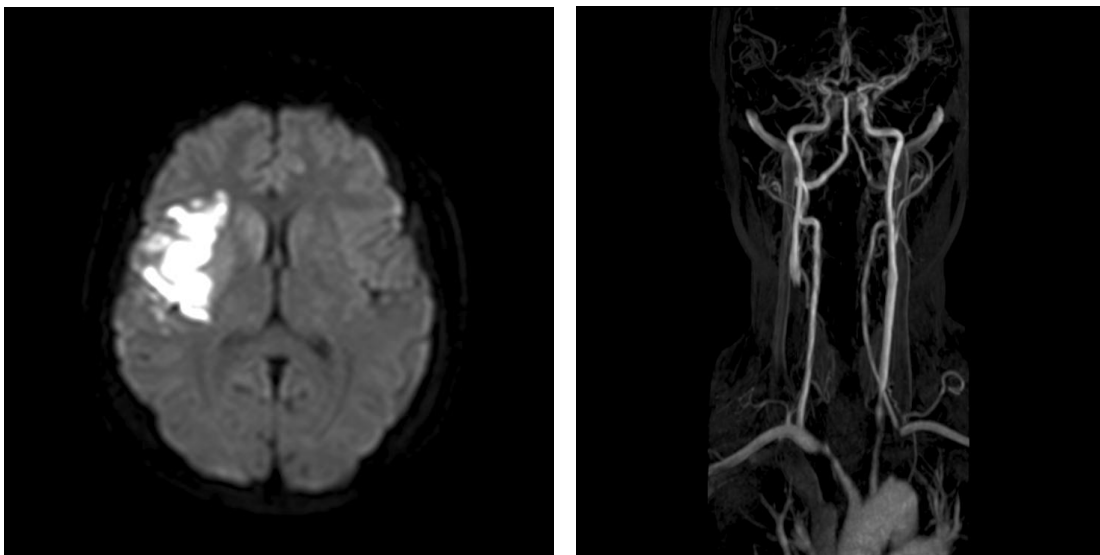


Figure 2. (A) The diffuse weighted magnetic resonance images revealed hyperdensity in the right middle cerebral artery territory, indicating the acute ischemic stroke. (B) Magnetic resonance angiography revealed narrowing lumens of the right brachiocephalic and proximal left common carotid artery and occlusion of the left subclavian artery, right common carotid artery and middle cerebral artery.

DISCUSSION

Takayasu's arteritis is an idiopathic, chronic inflammatory large-vessel arteriopathy that primarily affects the aorta and its main branches.¹ It is a rare disease and was first reported in 1905 by Mikito Takayasu, an ophthalmologist, in a case with peculiar changes in the central retinal vessels.⁶ This disorder is most common in Japan and to date more than 5000 patients have been registered by the Japanese government.¹ Women are most affected and it most commonly occurs in the second and third decades of life.⁵

The symptoms of Takayasu's arteritis vary depending on the site and degree of arterial lesions. The pattern of Takayasu's arteritis is typically triphasic, consisting of a systemic non-vascular phase, a vascular inflammatory phase, and a quiescent "burnt out" phase.⁷ Most patients initially present with non-specific symptoms, such as fever, night sweats, malaise and arthralgia. As the disease progresses, symptoms of end-organ disease due to the ischemia, including renovascular hypertension or coronary artery disease, may develop. Neurological complication occurring in the chronic phase of the disease, range from asymptomatic disease to catastrophic neurological impairment. In a study of 63 consecutive patients with Takayasu's arteritis, neurological manifestations were common symptoms in the chronic phase. It included dizziness, visual disturbance, headache, ischemic stroke, seizures, and some unusual manifestations such as reversible posterior encephalopathy syndrome.⁸ Although neurological involvement, including transient ischemic stroke, acute stroke

or cranial nerve palsies, has been reported in 10 to 24% of patients with Takayasu's arteritis^{2,9-11}, stroke as the initial presentation is rarely described in the literature and these cases are summarized in Table 1.^{4,5,12-14}

The exact etiology of ischemic stroke in Takayasu's arteritis remains unclear. The decreasing of cerebral blood flow from the stenotic or occlusive lesion in the aortic arch and its main branches, and cardiac diseases such as aortic regurgitations have been described to account for the stroke in most patients.¹⁵ Intracranial stenosis as a consequence of vasculitic involvement, or prior embolization into the vessel are hypothesized.¹⁶ In a review of 190 patients with Takayasu's arteritis, the lesion patterns and possible mechanisms in 21 ischemic stroke events with relevant ischemic lesions on MRI were investigated.¹¹ These ischemic lesions were categorized into five subgroups (cortical border-zone, internal border-zone, large lobar, large deep subcortical, and small subcortical infarctions) depending on the lesion size and topographic distribution. The authors reported that most of the ischemic lesions were located at middle cerebral artery branches or in the internal/cortical border-zone area. The authors proposed that hemodynamic compromise in large-artery stenosis and thromboembolic mechanisms played significant roles in ischemic stroke associated with Takayasu's arteritis.

Several criteria for the diagnosis of Takayasu's arteritis were proposed by the American College of Rheumatology in 1990. They included (1) Age at onset of less than 40 years, (2) Claudication of the extremities, (3) Decreased brachial artery

Table 1: The summary of published cases presenting stroke as the first manifestation of Takayasu's arteritis

Author	Year	Age	Gender	Presentation	Ischemic region	Recovery
Khealani&Baig ⁴	2002	19	Female	Right hemiparesis, aphasia	Left basal ganglia and parietal lobe	Partial
		36	Female	Left hemiparesis	Unknown	Unknown
Sikaroodiet al. ⁵	2007	50	Female	Repeated and aggravated hemiparesis and mental decline	Left frontal, left internal capsule, right basal ganglia and bilateral parietal lobe	Stable
Gao &Wang ¹²	2013	35	Female	Left hemiparesis, pain and numbness of arms	Right middle cerebral artery territory	Stable
Humayunet al. ¹³	2014	28	Female	Left hemiparesis	Right frontal area	Stable
Pereira et al. ¹⁴	2014	19	Female	Right hemiplegia, dysarthria, aphasia	Left cerebral hemisphere	Stable
Our case	2015	36	Female	Left hemiparesis, dizziness	Right middle cerebral artery territory	Stable

pressure, (4) Blood pressure difference of more than 10mmHg, (5) Bruit at the subclavian arteries and aorta, and (6) Aortograph abnormalities.⁶ The presence of three of the six criteria was required, which was associated with a sensitivity of 90.5% and a specificity of 97.8%. However, because of the rarity of Takayasu's arteritis, the diagnosis and treatment may be delayed, especially when neurological symptom such as acute stroke occurs as an initial presentation. A high index of suspicion should arise when the stroke develops in young patients with asymmetrical pulses and blood pressure, presence of systemic symptoms, claudication of limbs, and elevated ESR.⁵

Steroid is the mainstay of treatment for Takayasu's arteritis. Cytotoxic drugs, including cyclophosphamide, azathioprine, and methotrexate have been used to manage steroid-unresponsive patients. However, when acute vascular compromise such as acute stroke occurs, thrombolytic, surgical or interventional revascularization should be considered. The survival rates at 5, 10 and 15 years are 91%, 84% and 83%, respectively.⁵

In our patient, only asymmetry of blood pressure in the four limbs and the lack of a pulse at the distal arteries were seen. Ischemic stroke caused by embolic materials resulting from an active inflammatory process was hypothesized.¹⁷ Decompressive craniectomy was performed to decrease the mass effect resulting from the large cerebral infarction. Considering our patient's condition, revascularization of the main vessels was urgently performed to prevent the progression of stenosis. Steroid was also administered to control the inflammatory process.

In conclusion, although neurological manifestations are common in patients with Takayasu's arteritis in the chronic phase, acute stroke as an initial presentation has rarely been reported. Our patient shows that Takayasu's arteritis should be considered in the differential diagnosis of young stroke.

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