

Takayasu Arteritis Masquerading as Acute Ischaemic Stroke in a Young Patient: A Case Report

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ABSTRACT

Takayasu Arteritis (TA) is a form of vasculitis that involves large vessels. It is quite unusual for patients suffering from TA to present with an acute stroke as the initial symptom. This case involves a 40-year-old woman who presented with weakness of her limbs on the left-side and aphasia since the morning. There was no pulse in the left radial and brachial arteries; however, the left carotid and subclavian arteries had a bruit. Magnetic Resonance Imaging (MRI) and digital subtraction angiography were performed, and based on the results a diagnosis of TA was made. A multidisciplinary treatment approach was planned, and different drugs were administered to the patient, along with appropriate advice for follow-up after a month. The patient successfully underwent stenting for the lesion, and the treatment regimen included Ecospirin, Atorvastatin, methotrexate, and prednisone daily. Computed Tomography (CT) angiograms were conducted to monitor the progression of the vascular disease. The risks and benefits of potential complications in managing long-term care must be cautiously assessed. Early recognition and timely treatment are crucial for the patient's survival. Hence, it is important to consider TA as a differential diagnosis in young patients and to remain alert to the nonspecific symptoms of TA.

Keywords: Large cell arteritis, Neurology, Pulseless disease, Rheumatology

CASE REPORT

A 40-year-old female presented to the hospital with complaints of weakness in her left upper and lower extremities. She also reported experiencing aphasia since morning. Upon arrival at the emergency department, the patient was alert and oriented. She denied any family history of cardiac, cerebrovascular, or rheumatologic diseases. She had no history of hypertension, diabetes mellitus, bronchial asthma, or any thyroid disorder.

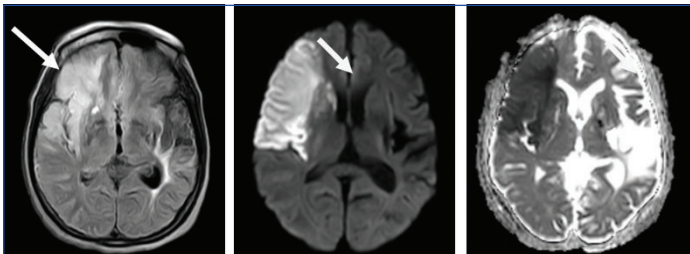
On general examination, her temperature was 97.5°F, and she exhibited a hypovolemic pulse on the right-side, with absent pulse in the left brachial and radial arteries. Carotid and subclavian bruits were present. Blood pressure was non recordable in the left arm, 100/64 mmHg in the right arm, 148/64 mmHg in the right leg, and 126/52 mmHg in the left leg. Her respiratory rate was 16 cycles per minute, and oxygen saturation was 99%. Heart sounds S1 and S2 were present, but there were no murmurs. Bilateral air entry was noted, and the abdomen was soft and non tender.

Higher mental functions were intact, and cranial nerves were normal. Motor strength for the upper and lower limbs on the left-side was graded as 3/5 while on the right-side it was graded as 5/5. Hypotonia was present. Deep tendon reflexes were rated as 3, and the plantar reflex showed a right flexor and left extensor response. The sensory exam yielded normal results, and there were no cerebellar signs. The patient exhibited an apraxic gait. Signs or symptoms of rigidity or bradykinesia, as well as extrapyramidal syndrome and signs of autonomic failure were absent. Her laboratory investigations were recorded as follows [Table/Fig-1].

The MRI of the brain indicated altered signal intensity in the right frontotemporal region, right corona radiata, and right insular cortex. The findings were hyperintense on Fluid-Attenuated Inversion Recovery (FLAIR) [Table/Fig-2a]; A white arrow indicates restriction on Diffusion Weighted Imaging (DWI) [Table/Fig-2b]; and another white arrow shows corresponding low signals on Apparent Diffusion Coefficient (ADC); [Table/Fig-2c]. These findings were suggestive of an acute infarct. [Table/Fig-2a-c] indicates an acute infarct in the right frontotemporal region, corona radiata, and right insular cortex.

Laboratory investigations	Value	Biological reference range
Haemoglobin	10.1	13-15 g/dL
Mean corpuscular volume	84	79-100 fL
Total leukocyte count	7,700	4000-11,000/cumm
Platelet count	460,000	1,50,000-4,50,000/cumm
Prothrombin time/international normalised ratio	0.7	<=1.1
Serum urea	14	5-20 mg/dL
Serum creatinine	0.5	0.3-1.2 mg/dL
Serum sodium	140	135-145 mmol/L
Erythrocyte Sedimentation Rate (ESR)	25	<20 mm/hr
C reactive protein	2.48	<1.0 mg/dL
Antinuclear antibody	1:1000	<1:40
Antithrombin III	Normal	
Protein C	Normal	
Protein S	Normal	

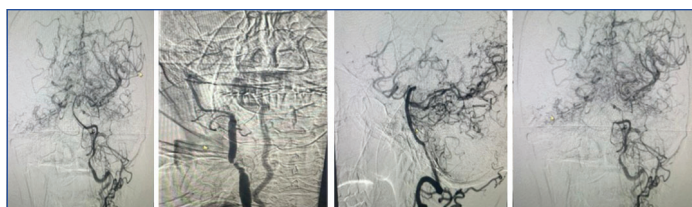
[Table/Fig-1]: Laboratory investigations.



[Table/Fig-2]: Magnetic Resonance Imaging (MRI) of brain suggestive of acute infarct in right frontotemporal, corona radiata and right insular cortex. a) Fluid attenuated inversion recovery; b) Diffusion weighted image; c) Apparent diffusion coefficient. (Images from left to right)

[Table/Fig-3] shows the a Digital Subtraction Angiogram (DSA) indicating diffuse vasculitis. The patient was taken immediately for the DSA procedure, which was uneventful.

With the patient under local anaesthesia, following regular prepping and draping of the right groin, a 5F Picard catheter was advanced



[Table/Fig-3]: Digital Subtraction Angiogram (DSA) suggestive of diffuse vasculitis.

into the aortic arch. Selective catheterisation of the following vessels was performed. There was evidence of luminal irregularity with narrowing involving the mid-right Common Carotid Artery (CCA), causing approximately 70-80% stenosis with poor anterograde flow. The right Internal Carotid Artery (ICA) was occluded from its origin. The right Anterior Cerebral Artery (ACA) and Middle Cerebral Artery (MCA) were being partially perfused by the Right Posterior Communicating Artery (PCOM) and leptomeningeal collaterals from the right Posterior Cerebral Artery (PCA). The venous phase appeared normal. There was also evidence of luminal irregularity with narrowing involving the left CCA origin, leading to poor anterograde flow. Additionally, there was luminal irregularity with narrowing involving the left proximal ICA, causing approximately 80-90% stenosis with sluggish anterograde flow. The left ACA and MCA were being partially perfused by the PCOM and leptomeningeal collaterals from the left PCA. The venous phase also appeared normal. The findings in the vessels suggested large vessel vasculitis. Based on clinical findings, laboratory investigations, radiological investigations, and digital subtraction angiography, a presumptive diagnosis of Takayasu arteritis was made.

Upon hospital discharge, the patient had notably regained motor function, showing 4 out of 5 strength on the left-side though she still exhibited mild residual expressive aphasia. She was transferred to an inpatient rehabilitation facility for intensive physical, speech, and occupational therapy. Six months later, during a routine follow-up, the patient still displayed significant differences in blood pressure between her upper extremities and bruits over her subclavian and carotid arteries. The patient successfully underwent stenting for the lesion. Her treatment regimen included Ecosprin 150 mg once a day, Atorvastatin 40 mg at bedtime, Methotrexate at 25 mg weekly, and Prednisone at 7.5 mg daily, with scheduled interval CT angiograms to monitor the progression of the vascular disease. Limb physiotherapy was also provided for the patient.

DISCUSSION

The TA is a chronically progressive inflammatory disease of unknown aetiology, primarily involving the aorta and its main branches as well as the pulmonary arteries. Given that this condition predominantly affects large arteries, it has sometimes been referred to as the "pulseless" disease [1]. Common causes of ischaemic stroke in both younger and older populations include atherosclerosis and embolism [2]. However, stroke as the presenting manifestation of TA is extremely uncommon, with only a few cases reported in the literature [3,4]. During the acute phase, many patients experience associated systemic symptoms such as fever, malaise, and weight loss. The Erythrocyte Sedimentation Rate (ESR) and C-Reactive Protein (CRP) are commonly elevated in this context. Glucocorticoids are usually effective during the acute phase for the majority of patients. To reduce the use of glucocorticoids and manage relapses, immunosuppressive drugs such as methotrexate, azathioprine, leflunomide, or mycophenolate are used. Although biologically targeted agents have sometimes been prescribed, their effectiveness has not been proven in randomised clinical trials. In severe cases with critically narrowed arteries, surgical bypass or endovascular procedures may be required. TA is a rare disease that predominantly affects young female patients and is characterised by an inflammatory process involving large vessels,

particularly the aorta and its major branches. The exact aetiology of TA remains unknown, and it may also occur in the paediatric age group. The presentation of TA is highly variable and may include either impalpable pulses or bruits, most commonly detected at the carotid or radial arteries, or hypertension secondary to critical renal stenosis, myocardial ischaemia resulting from coronary involvement, or cerebrovascular diseases [5].

Stroke is an unusual presenting feature of TA [6]. With an estimated frequency of 10-20%, strokes are a common complication of TA. However, stroke as the initial manifestation is uncommon, and there are not many case reports in the literature [7]. TA should be considered in the differential diagnosis of stroke, particularly in the absence of usual risk factors but in the presence of dyslipidemia and/or hypertension [8].

Cerebrovascular problems can significantly impact morbidity and mortality in patients with TA, and physicians need to pay close attention to this complication. The management of stroke in patients with TA involves implementing a well-planned strategy to avoid morbidity and prevent unnecessary deaths. Early recognition of nonspecific symptoms, especially in younger stroke patients, is crucial. Such nonspecific symptoms may include fever, fatigue, dizziness, an irregular pulse, and fluctuations in blood pressure [9]. A South Korean study revealed that cerebral infarction associated with TA was less common in females than in males. Moreover, most stroke events in males occurred within six months of diagnosis, while in females, such events were more frequently observed after three years [10]. According to Russo RAG and Katsicas MM, immunosuppressant therapy, along with glucocorticoids, proved to be very effective in patients diagnosed with TA [11]. However, there is currently no summary of well-designed clinical studies available on the treatment strategy for patients with TA.

The rarity of TA makes it challenging to diagnose and treat, especially when the initial manifestation is neurological symptoms such as acute stroke. In cases of stroke in young patients with asymmetric pulses, blood pressure differences, systemic symptoms, limb claudication, and high ESR, clinicians should maintain a high index of suspicion.

In 1990, the American College of Rheumatology developed a set of criteria for the diagnosis of TA:

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

At least three of the above six criteria must be met for diagnosis, demonstrating a sensitivity of 90.5% and a specificity of 97.8% [12].

The treatment of TA mainly involves corticosteroid therapy. In some patients who do not respond to steroids, cytotoxic agents such as cyclophosphamide, azathioprine, or methotrexate can also be used. In an acute vascular condition, such as a patient suffering from a stroke, thrombolysis or even surgical interventions like revascularisation may be necessary to avoid increased vascular damage. The difference in blood pressure between the limbs, along with the loss of a pulse in the right upper limb, indicated the urgent need for revascularisation to prevent the aggravation of existing arterial stenosis. In this patient, the observation of blood pressure differences among all four limbs and the absence of a pulse in the right upper limb at the distal arteries suggested an urgent need for revascularisation to prevent worsening stenosis.

CONCLUSION(S)

In conclusion, stroke is a very rare clinical presentation of TA, and a proper workup of a patient presenting with stroke should be

conducted. Vasculitis and infarcts were detected through laboratory investigations and imaging. Considering the low success rates of endovascular approaches, it is prudent to explore the feasibility of combined treatment options, taking into account the potential risks and benefits. In this case, a multidisciplinary approach was utilised, and the patient was discharged from the hospital on outpatient treatment, which includes rehabilitative therapy and methylprednisolone pulse therapy, followed by oral steroids and antiplatelet drugs.

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