

Severe carotid obstruction in a patient with takayasu's arteritis: case report and literature review

Abstract

Takayasu's arteritis (TA) is a rare large-vessel vasculitis that predominantly affects women, usually in their second or third decade of life. Accurate diagnosis relies on clinical evaluation and imaging studies such as angiography, computed tomography (CT), Doppler ultrasound, and magnetic resonance imaging (MRI). Diagnostic criteria have evolved over the past quarter-century, with the American College of Rheumatology (ACR) establishing definitions and clinical classifications that facilitate a stepwise approach to diagnosis, leading to timely identification and prevention of severe complications. This case report describes a 35-year-old female patient whose imaging findings prompted a diagnostic approach based on the literature, ultimately leading to a diagnosis of Takayasu's arteritis.

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Introduction

Takayasu's arteritis (TA) is a chronic, idiopathic vasculitis primarily affecting large-caliber arteries, including the aorta and its major branches. The prevalence of TA is relatively low, estimated at around 4 to 300 per million people, and it is more common in young women in their second and third decades of life, although it can occur at any age.¹ The clinical manifestations of TA are variable, which can delay diagnosis. Initial nonspecific symptoms such as fatigue and fever often precede more specific vascular manifestations, which vary depending on the arterial involvement. Diagnostic suspicion is established through clinical findings and imaging studies, such as angiography, CT, or MRI, which reveal significant vessel stenoses, occlusions, or dilatations. Diagnosis is confirmed based on clinical and classification criteria defined by the American College of Rheumatology.²

Here, we present a clinical case of a 35-year-old female patient with bilateral carotid obstruction as an imaging diagnostic finding of TA, followed by a discussion of the case in the current literature.

Case report

A 35-year-old female housewife with no significant medical history presented with symptoms lasting 7 months. These symptoms included repeated transient ischemic attacks before the onset of her current symptoms. Her current symptoms included headaches that were felt all over the head, had a pulsating sensation, and occurred intermittently. The intensity of these headaches was 7 out of 10 on the visual analog pain scale. The headaches did not follow a specific pattern, got worse with position changes, improved with oral analgesics, happened daily, lasted approximately 24 hours. The patient experienced symptoms such as nausea, dizziness, and sensitivity to sound. During the physical examination, the patient appeared stable with normal vital signs, but a bruit was detected in the right carotid artery. Initial laboratory tests, including complete blood count, renal function, electrolytes, glucose, and liver function tests, revealed no abnormalities.

A Doppler ultrasound of the neck vessels in B-mode revealed regular circumferential wall thickening of the right common carotid artery with a mean intima-media thickness of 1.8 mm. Color Doppler showed long-segment intima-media thickening with a regular wall,

resulting in 70% stenosis of the vessel lumen. The left common carotid artery was occluded entirely from its origin, with collateral flow through the internal carotid artery. A non-contrast CT of the brain showed no evidence of lesions.

Subsequent cerebral angiography revealed irregularities in the right common carotid artery with 70% stenosis at the origin of the internal carotid artery and complete occlusion of the left common carotid artery at its origin (Figure 1). These imaging and angiographic findings suggest a primary diagnosis of TA, prompting consultation and multidisciplinary management by neurology and rheumatology.



Image 1

Image A: Cross-sectional carotid ultrasound showing circumferential thickening of the right common carotid artery, referred to as the "macaroni sign."

Image B: Longitudinal section of the same right common carotid artery showing increased arterial wall thickening with extended, smooth distribution. Blood flow is visible, circulating endoluminally.

Image C: Selective angiography of the right common carotid artery showing a longitudinal filling defect along its ordinary course and focal repletion defect in the proximal segment of the internal carotid artery.

Discussion

TA is a chronic inflammatory disease predominantly affecting large arteries, especially the aorta and its major branches, with a higher incidence in young women, having a female-to-male ratio of approximately 8:1. The typical age of onset is between 10 and 40 years.² Our patient, a 35-year-old woman, aligns with the demographic described in the literature for this disease.

The exact cause of TA is unknown, but it is believed to have an autoimmune basis, with recent studies implicating genetic factors, infections, and autoimmunity in its development.³ In our patient, we

have not yet identified specific risk factors associated with the onset of the disease. Clinically, TA presents a broad spectrum of symptoms that vary depending on the affected arteries and is generally divided into two phases: an early or inflammatory phase characterized by symptoms such as fever, fatigue, weight loss, arthralgia, and general malaise and a late or occlusive phase, where specific symptoms associated with arterial obstruction or damage appear, including headache, neurological symptoms, claudication of limbs (due to ischemia), absent or weakened peripheral pulses, arterial bruits, renovascular hypertension, strokes, heart failure, and chest pain (due to aortic involvement).^{4,5} Our patient exhibited a headache associated with neurological symptoms and a carotid bruit secondary to carotid artery involvement, as evidenced by angiography.

Table 1 Classification Criteria for TA according to the American College of Rheumatology 2022. Adapted from (2).

Absolute criteria	Additional clinical criteria	Additional imaging criteria
	Female sex +1	
	Angina or cardiac pain with ischemic characteristics +2	Number of arterial territories affected
Age less than or equal to 60 years at the time of diagnosis	Intermittent claudication of arms or legs +2	- An arterial territory +1 - Two arterial territories +2
Evidence of vasculitis in images	vascular murmur +2	- Three or more arterial territories +3
	Reduced pulses in upper limbs +2	Symmetrical involvement of paired arteries +1
	Carotid artery abnormality +2	Commitment of the abdominal aorta with involvement of the renal or mesenteric arteries +3
	Systolic pressure difference in upper limbs >20 mmHg +2	

A score greater than 5 on additional criteria is necessary for the diagnosis of AT.

The gold standard for diagnosis includes imaging studies, with conventional angiography being the primary method as it allows for evaluating arterial stenosis and occlusions. Alternatively, CT and MRI with contrast are useful for visualizing arterial wall thickening and luminal changes. Ultrasound Doppler can be a non-invasive method to assess blood flow and arterial lesions.³ In our patient, the initial Doppler ultrasound was performed due to clinical concerns at this level, followed by a non-contrast CT of the brain that showed no abnormalities, and finally, carotid angiography, which confirmed the diagnosis.

Treatment for TA aims to reduce inflammation and prevent vascular complications. First-line therapy includes immunosuppressive medications such as corticosteroids, the initial treatment to control inflammation. Other immunosuppressants such as methotrexate, azathioprine, mycophenolate mofetil, or biological agents (e.g., tocilizumab) may be necessary to reduce corticosteroid dosage and maintain remission.⁷ Other therapeutic options include surgical or endovascular interventions such as angioplasty with or without stent placement, arterial bypass, or revascularization surgery to address severe stenosis or occlusions. Additionally, a comprehensive approach is required for managing complications, including strict control of hypertension, prevention of ischemic events with anticoagulants or antiplatelet agents, treatment of heart failure, and other complications.⁸

Conclusion

Takayasu's arteritis is a rare and potentially severe vasculitis that primarily affects young women. Early diagnosis and aggressive treatment are essential to improving the prognosis and quality of life.

The diagnosis of TA is based on a combination of clinical criteria, laboratory findings, and imaging studies. The clinical criteria were outlined by the American College of Rheumatology (ACR) in 1990 and include age at onset <40 years, limb claudication, decreased brachial pulses, a difference in blood pressure >10 mmHg between arms, bruits over the aorta or large arteries, and abnormal arteriography without evidence of atherosclerosis.⁶ In 2022, the criteria were updated and categorized into absolute, additional clinical, and additional imaging criteria (Table 1).² Our patient meets all the absolute criteria and five additional clinical criteria, including female sex, presence of a vascular bruit, and abnormal carotid artery findings. Regarding additional imaging findings, we observed alterations in two arterial territories (+2) and symmetrical involvement of paired arteries (+1).

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Multidisciplinary collaboration among rheumatology, cardiology, neurology, and vascular surgery is crucial for comprehensive disease management.

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None.

Conflicts of interest

The authors declare that there are no conflicts of interest.

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