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IMAGING

IMAGING VIGNETTE: CLINICAL VIGNETTE

Asymptomatic Takayasu Arteritis With Critical Multivessel Stenosis



Vanshika Tripathi, DO,^a Matthew Montanarella, DO,^b Yixin Zhang, MD,^c Jason R. Young, MD,^d Madeline K. Mahowald, MD^c

ABSTRACT

Takayasu arteritis is characterized by blood vessel inflammation involving the aorta and its branches. We describe a patient with Takayasu arteritis with severe multivessel involvement and classic physical examination findings but virtually no symptoms because of the presence of extensive collateral circulation seen on computed tomography angiography and magnetic resonance angiography imaging. (J Am Coll Cardiol Case Rep 2024;29:102252) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

50-year-old asymptomatic woman was referred to cardiology for a history of Takayasu arteritis diagnosed decades earlier in her home country of Albania. She had a history of hypertension, but antihypertensive agents had been discontinued by her primary physician because of low upper extremity blood pressure (BP) readings. Four-limb BPs taken in her cardiologist's office were noted to be 75/57 mm Hg in the left upper extremity, 75/58 mm Hg in the right upper extremity, 245/104 mm Hg in her left lower extremity, and 235/148 mm Hg in her right lower extremity. Physical examination revealed diminished carotid pulses, a left carotid bruit, a diminished left radial pulse, an absent right radial pulse, and normal lower extremity pulses. Computed tomography angiography (CTA) showed occlusion of the right brachiocephalic and right common carotid arteries with reconstituted flow at the carotid bifurcation, occlusion in the proximal right vertebral artery V1 segment with reconstituted flow in the distal V1 segment, occlusion of the proximal left subclavian artery with reconstituted flow proximal to the origin of the left vertebral artery, and diseased vertebral arteries likely reconstituted by circle of Willis or thyrocervical branches. Prominent collateral vessels, including the lateral thoracic, intercostal, and inferior phrenic arteries supplied the head, neck, and upper extremities (Figures 1A to 1E). Magnetic resonance angiography (MRA) did not show evidence of active vascular inflammation, and serum inflammatory marker values were normal.

Takayasu arteritis is an inflammatory process that most commonly affects the aorta and its branches, with a propensity to affect young women. Its pathogenesis is complex and likely involves immune-mediated granulomatous inflammation of arterial walls in patients with a genetic predisposition. Intimal hyperproliferation and fibrosis can lead to occlusive vasculopathy or aneurysm formation. Because blood flow is compromised, organ damage from ischemia can occur, leading to limb claudication, angina, visual changes, syncope, and stroke. Ischemic injury and symptoms can sometimes be alleviated by collateral circulation. Although

From the ^aDepartment of Internal Medicine, University of Florida College of Medicine-Jacksonville, Jacksonville, Florida, USA; ^bDepartment of Radiology, University of Florida College of Medicine-Jacksonville, Jacksonville, Florida, USA; ^cDivision of Cardiology, Department of Internal Medicine, University of Florida College of Medicine-Jacksonville, Jacksonville, Florida, USA; and the ^dDepartment of Radiology, Mayo Clinic, Jacksonville, Florida, USA.

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ABBREVIATIONS AND ACRONYMS

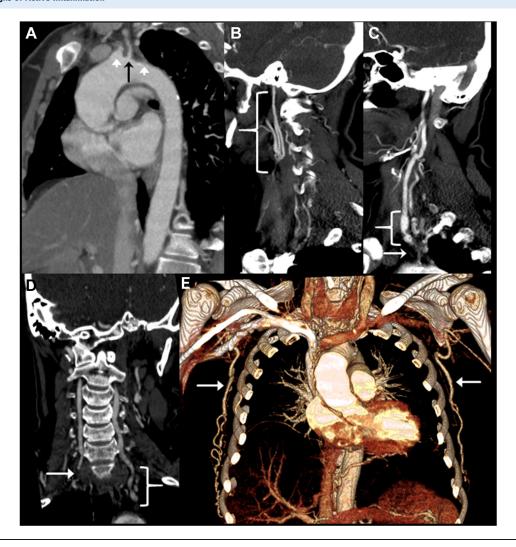
BP = blood pressure

CTA = computed tomography angiography

MRA = magnetic resonance angiography collateral flow serves as a mechanism to counteract ischemic damage, it can also delay diagnosis in patients with severe great vessel involvement who remain asymptomatic.

CTA and MRA are the cornerstone in diagnosing and monitoring Takayasu arteritis.² There is evidence to support the use of fluorine-18 fluorodeoxyglucose positron emission tomography in clarifying disease activity and recurrence, which can be difficult to determine clinically.³

FIGURE 1 Computed Tomography Angiogram With Contrast Reveals Extensive Postinflammatory Changes of the Arch Vessels Without Signs of Active Inflammation



(A) A computed tomography angiogram of the thorax demonstrating chronic occlusions at the origins of the right brachiocephalic and left subclavian arteries (white arrowheads) with severe stenosis of the left carotid artery origin (black arrow) yet sparing of the aorta. (B) A multiplanar computed tomography angiogram of the right carotid artery demonstrating chronic occlusion of the right carotid artery with reconstitution at the bulb with multiple surrounding collateral vessels (white bracket). (C) A multiplanar computed tomography angiogram of the left carotid artery demonstrating tandem severe stenoses at the left carotid artery origin (white arrow) with poststenotic dilatation (white bracket). (D) Coronal computed tomography angiogram of the vertebral arteries showing chronic occlusion of the right vertebral artery at the origin (white arrow) with reconstitution and stenosis of the left vertebral artery at the origin (white bracket). (E) Maximum intensity projection computed tomography angiogram of the thorax highlighting enlarged lateral thoracic arteries (white arrows) supplying the bilateral upper extremities. Perfusion to the head and neck is provided by the left carotid artery and the left vertebral artery, both of which have severe stenoses. Perfusion to the bilateral upper extremities is largely supplied by retrograde flow through the lateral thoracic arteries.

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Our patient's disease pattern was categorized as type 1 Takayasu arteritis, which affects the aortic arch branch vessels but spares the aorta. Regular physical activity in her work as a housekeeper encouraged the development of robust collateral vessels and prevented the development of symptoms. Rheumatology did not recommend immunosuppression in the absence of evidence of active inflammation. She was started on losartan 25 mg daily with a plan for slow up-titration and permissive hypertension to prevent cerebral hypoperfusion. It is imperative to avoid rapid BP correction in patients with chronic hypertension and occluded or stenotic vessels supplying the brain. The optimal BP target is unknown in this patient, and she remains at risk for catastrophic neurologic injury should her left carotid artery become compromised. She is considering total aortic arch replacement.

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ADDRESS FOR CORRESPONDENCE: Dr Madeline K Mahowald, Division of Cardiology, Department of Medicine, University of Florida-Jacksonville, ACC 5th Floor, 655 West 8th Street, Jacksonville, Florida 32209, USA. E-mail: madeline.mahowald@jax.ufl.edu.

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KEY WORDS collateral circulation, CT angiography, pulseless disease, Takayasu arteritis, vasculitis