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IMAGING VIGNETTE

INTERMEDIATE

CLINICAL VIGNETTE

Short-Segment Type B Interrupted Aortic Arch Presenting With Subarachnoid Hemorrhage With Subsequent Primary Percutaneous Repair

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ABSTRACT

This report describes a young adult man presenting with subarachnoid hemorrhage secondary to an intracranial aneurysm who was found to have a short-segment type B interrupted aortic arch. We describe the clinical presentation, evaluation, and management of this patient and highlight imaging findings and percutaneous repair of the aneurysm and interrupted aortic arch. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:1607–1609) © 2021 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/).

A 34-year-old male patient presented with a severe posterior headache, dizziness, nausea, photophobia, and bilateral foot numbness. Physical examination revealed anisocoria, as well as right arm blood pressure 174/74 mm Hg, with a 50 mm Hg systolic blood pressure difference between arms.

Computed tomography angiography (CTA) revealed a subarachnoid hemorrhage with a 1.1-cm ruptured left vertebrobasilar aneurysm. Angiography of the right vertebral artery through right radial access identified the vertebrobasilar aneurysm, which was successfully coiled without complications (**Figure 1A**). During percutaneous repair, angiography demonstrated retrograde flow in the left vertebral artery toward the left subclavian artery and into the descending aorta, consistent with subclavian steal syndrome. Angiography of the aortic arch through right radial access demonstrated an interrupted aortic arch (IAA) proximal to the left subclavian artery (**Figure 1B**).

The patient's headache improved. He received labetalol for hypertension and nimodipine to prevent cerebral vasospasm. CTA of the chest and abdomen again demonstrated a short-segment type B IAA with a dense network of collateral vessels in the lower neck, mediastinum, and thoracic wall reconstituting the descending aorta, mainly through the left subclavian artery and supreme intercostal artery (**Figure 1C**, Video 1). The electrocardiogram showed left ventricular (LV) hypertrophy (LVH) with a repolarization abnormality, and transthoracic echocardiography (TTE) demonstrated moderate concentric LVH with normal LV systolic function.

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ABBREVIATIONS AND ACRONYMS

CTA = computed tomography angiography

IAA = interrupted aortic arch

LV = left ventricular

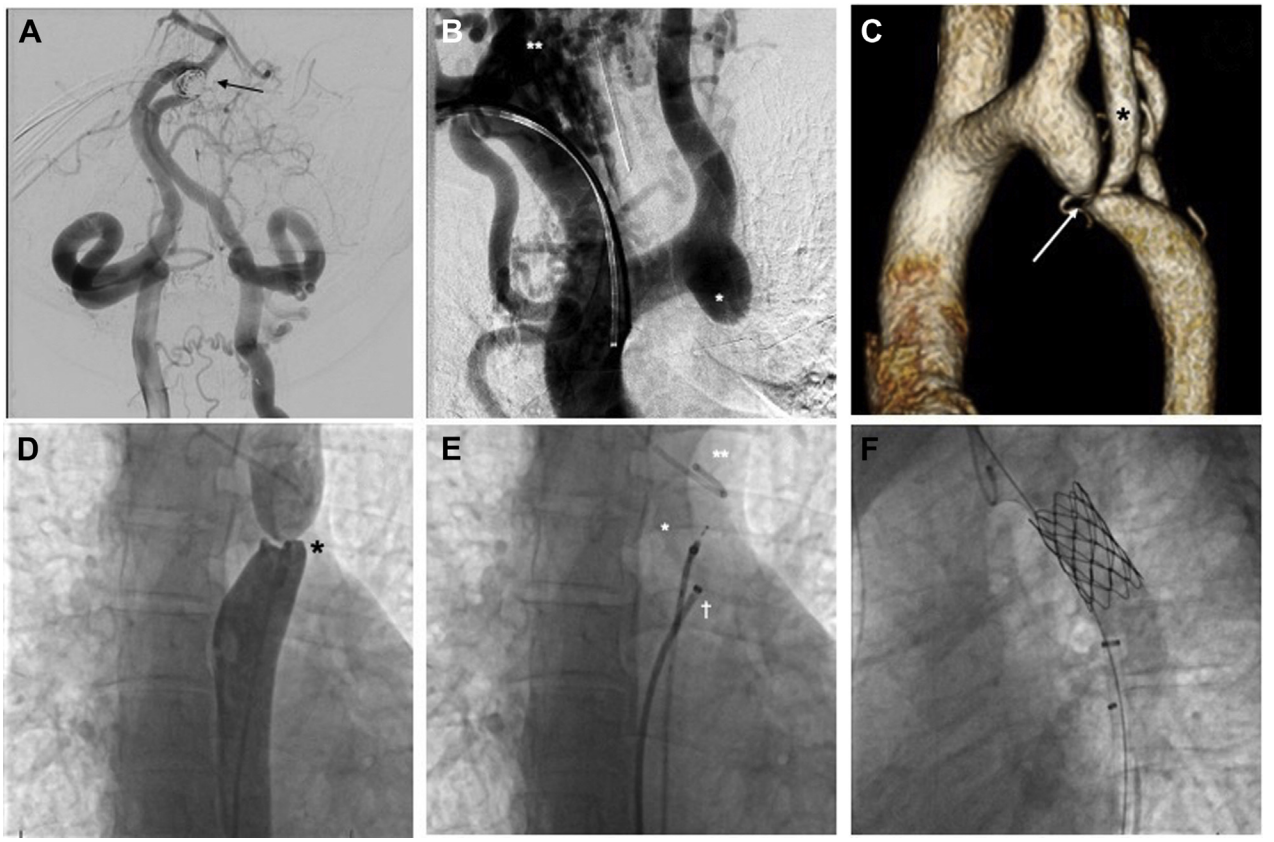
LVH = left ventricular hypertrophy

TTE = transthoracic echocardiography

Two weeks later, the patient was considered to be at acceptable risk to undergo transcatheter recanalization of his IAA. At cardiac catheterization, a short-segment type B IAA was confirmed, with peak-to-peak gradient of 80 mm Hg across the obstruction (Figure 1D). Radiofrequency energy allowed a 0.035-inch Powerwire (Baylis) to be advanced across the 1-mm-thick atretic plane (Figure 1E). Afterward, a 3.9-cm Covered CP Stent was delivered over a 16 mm × 4 cm BIB balloon catheter (B. Braun Interventional Systems Inc) and post-dilated with an 18 mm × 4 cm VIDA balloon (Becton, Dickinson and Company) (Figure 1F). Following stent placement, antegrade flow was confirmed without evidence of aortic dissection. Aortic arch to descending aorta peak-to-peak pressure difference was 8 mm Hg at the conclusion of the case.

The decision was made during the procedure to allow the covered stent to occlude the left subclavian artery takeoff. The left subclavian artery would still be perfused through the circle of Willis, retrograde through the left vertebral artery. Because flow was established through the aortic arch, the degree of residual subclavian steal to the left from the right was thought to be acceptable. Puncture of the covered stent

FIGURE 1 Diagnosis and Percutaneous Repair of Vertebrobasilar Aneurysm and Short-Segment Type B Interrupted Aortic Arch



(A) Successful coiling of vertebrobasilar aneurysm **(arrow):** There is evidence of subclavian steal as contrast material fills the left vertebral artery. The catheter is in the right vertebral artery. **(B)** Digital subtraction angiography demonstrating an interrupted aortic arch **(asterisk):** There is a dense network of collaterals in the neck **(double asterisk).** The catheter is in the ascending aorta. **(C)** 3-dimensional computed tomography angiographic reconstruction demonstrates the interrupted aortic arch **(arrow):** The descending aorta is reconstituted through collateral vessels, namely, the left subclavian artery **(asterisk). (D)** Simultaneous proximal and distal aortography of the interrupted aortic arch: A 1-mm atretic plane **(asterisk)** is seen with a peak-to-peak gradient of 80 mm Hg. **(E)** Radiofrequency perforation of the atretic plane: Radiofrequency energy allowed a 0.035-inch Powerwire (Baylis) to cross the atretic plane **(asterisk).** A pigtail catheter **(double asterisk)** and a long sheath **(dagger)** are in the proximal and distal aortic arch, respectively. **(F)** Covered stent deployed within the interrupted aortic arch: A 3.9-cm Covered CP Stent was delivered over a 16 mm × 4 cm BIB balloon (B. Braun Interventional Systems Inc) and post-dilated with an 18 mm × 4 cm VIDA balloon (Becton, Dickinson and Company).

and deployment of an additional stent to maintain ostial left subclavian artery patency were considered but not thought to be needed. It was acknowledged that surgical repair would have included reimplantation of the left subclavian artery to maintain ostial patency.

The postprocedure course was complicated by right radial artery occlusion and right upper extremity superficial venous thrombosis, both asymptomatic. Aspirin was initiated, and the patient was discharged on postprocedure day 3.

Follow-up 2 weeks after the procedure revealed a 6 mm Hg mean gradient across the stent by echocardiographic Doppler interrogation. The patient felt well. Lifelong surveillance will include regular evaluation for hypertension, stroke, coronary artery disease, and heart failure, along with annual TTE and CTA every 3 to 5 years to assess for stent fracture, aortic aneurysms, and restenosis.

IAA is a rare congenital heart malformation seen in approximately 0.012% of births worldwide and is often fatal once the ductus arteriosus closes if it is not immediately surgically repaired. Thus, new presentations of IAA in adulthood are extremely rare. Here we present a patient with subarachnoid hemorrhage from a ruptured vertebrobasilar aneurysm secondary to a high-flow state from subclavian steal syndrome from the right to the left, with the descending thoracic aorta reconstituted by collateral vessels. Management of IAA historically has been surgical but carries a risk of spinal cord ischemia as a result of cross-clamping (1). Several case series have demonstrated safety and efficacy of endovascular repair of IAA, including radiofrequency perforation, transseptal needle, and stiff guide wire techniques (2,3). Although no study has compared surgical with transcatheter techniques, suitable anatomy for endovascular repair is required, including a short atretic segment and lack of malalignment of the proximal and distal segments of the aorta, as was the case here. Further advances in percutaneous techniques can be expected to improve endovascular repair outcomes in the future.

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KEY WORDS 3-dimensional imaging, aortic coarctation, computed tomography, congenital heart defect, imaging

APPENDIX For a supplemental video, please see the online version of this paper.