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ADVANCED

CASE REPORT: CLINICAL CASE

CT—Guided Endovascular Exclusion of Pseudoaneurysmal Subclavian Bypass After Early-Age Surgical Correction of Complex Aortic Coarctation

Lamees I. El Nihum, BS,[a] Zhongyu Li, MS,[b] Ponraj Chinnadurai, MBBS, MMST,[c] Charudatta S. Bavare, MD, MPH,[b] Michael J. Reardon, MD,[b] Thomas E. MacGillivray, MD,[b] C. Huie Lin, MD, PhD[b]

ABSTRACT

We describe a 64-year-old woman with subclavian pseudoaneurysm after aortic coarctation repair, treated using a hybrid approach involving true three-dimensional analysis and image fusion-guided placement of thoracic endovascular aortic repair stents. This case illustrates the potential complications of coarctation repair and need for lifelong surveillance in these patients. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:225–9) © 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 64-year-old woman presented with 3 weeks of hypertension, chest pain, and hemoptysis. Prior work-up revealed a bicuspid aortic valve (BAV) without regurgitation or stenosis and no chest malignancy or infection. Physical examination was remarkable for a continuous murmur at the left side of the chest and a palpable pulsatile fullness above the left clavicle. Troponin levels were within normal range.

LEARNING OBJECTIVES

- To be aware that patients with aortic coarctation are at risk for aortic aneurysm or pseudoaneurysm in adulthood after balloon angioplasty, stent, or surgical repair of aortic coarctation.
- To recognize that persistent hypertension, cerebral aneurysms, early coronary artery disease, and bicuspid aortic valve disease may complicate the course of adults with coarctation even after successful childhood repair.
- To suggest that endovascular or hybrid approach may be the optimal intervention for these complex recurrent lesions when supported by advanced image fusion guidance.

MEDICAL HISTORY

The patient had previously undergone repair of aortic isthmus coarctation with an 18-mm “bucket handle” Hemashield vascular bypass graft (MAQUET, Rastatt, Germany) from the proximal left subclavian artery (LSCA) to the descending thoracic aorta at age 44.

DIFFERENTIAL DIAGNOSIS

We considered: 1) thoracic aortic aneurysm, dissection, or rupture; 2) malignancy; or 3) acute coronary syndrome.

From the [a]Texas A&M College of Medicine, Bryan, Texas, USA; [b]DeBakey Heart & Vascular Center, Houston Methodist Hospital, Houston, Texas, USA; and [c]Siemens Medical Solutions USA, Inc., Malvern, Pennsylvania, USA.

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ABBREVIATIONS AND ACRONYMS

3D = three-dimensional

BAV = bicuspid aortic valve

CTA = computed tomography angiography

LCCA = left common carotid artery

LSCA = left subclavian artery

TEVAR = thoracic endovascular aortic repair

INVESTIGATIONS

Computed tomography angiography (CTA) revealed >80% stenosis of the descending aorta with a minimal coarctation diameter of 2.5 mm and 5-cm aneurysmal dilatation of the subclavian bypass (**Figure 1**), which appeared to be expanding over time.

MANAGEMENT

True three-dimensional (3D) analysis was performed using an EchoPixel workstation (Santa Clara, California) and it aided in multidisciplinary review. Given the extremely high-risk nature of a redo surgical approach requiring deep circulatory arrest and potential for post-surgical collateral bleeding, the decision was made to proceed with a hybrid/endovascular approach.

Intraprocedural image registration of 2-dimensional fluoroscopy with CTA was performed and the origin of the left common carotid artery (LCCA), LSCA, coarctation, and distal anastomosis of the bypass graft were demarcated in 3D as targets for thoracic endovascular aortic repair (TEVAR) coverage. A guidewire was used to cross the aortic coarctation retrograde and snared via right radial artery access to form an arterial-arterial loop for support. The coarctation was dilated with a 6-mm Mustang balloon (Boston Scientific, Marlborough, Massachusetts) to allow passage of a 16-F Mullins sheath after which a 24 mm × 6 cm Covered Cheatham-Platinum Stent (NuMED, Hopkinton, New York) was deployed across the coarctation and the takeoff of the LSCA. Angiography demonstrated successful exclusion of the ostium of the LSCA and successful coarctation dilation.

A 31 mm × 26 mm × 10 cm Gore C-TAG (W. L. Gore & Associates, Inc., Flagstaff, Arizona) self-expanding stent graft was then deployed and extended by a 28 mm × 28 mm × 10 cm Gore C-TAG stent graft to exclude the distal graft to aorta anastomosis in the descending aorta, thus completing the TEVAR. After post-dilation, we used aortogram to confirm good apposition of the overlapping stents and complete exclusion of the inflow and outflow of the bypass graft and pseudoaneurysm (**Figure 2**).

To prevent retrograde filling of the pseudoaneurysm from the LSCA, the proximal subclavian was occluded with a 16-mm Amplatzer Vascular Plug II (Abbott Vascular, Santa Clara, California) (**Figure 3**). Finally, to ensure adequate perfusion of the left arm, an open surgical LCCA to LSCA bypass was performed (**Figure 3**).

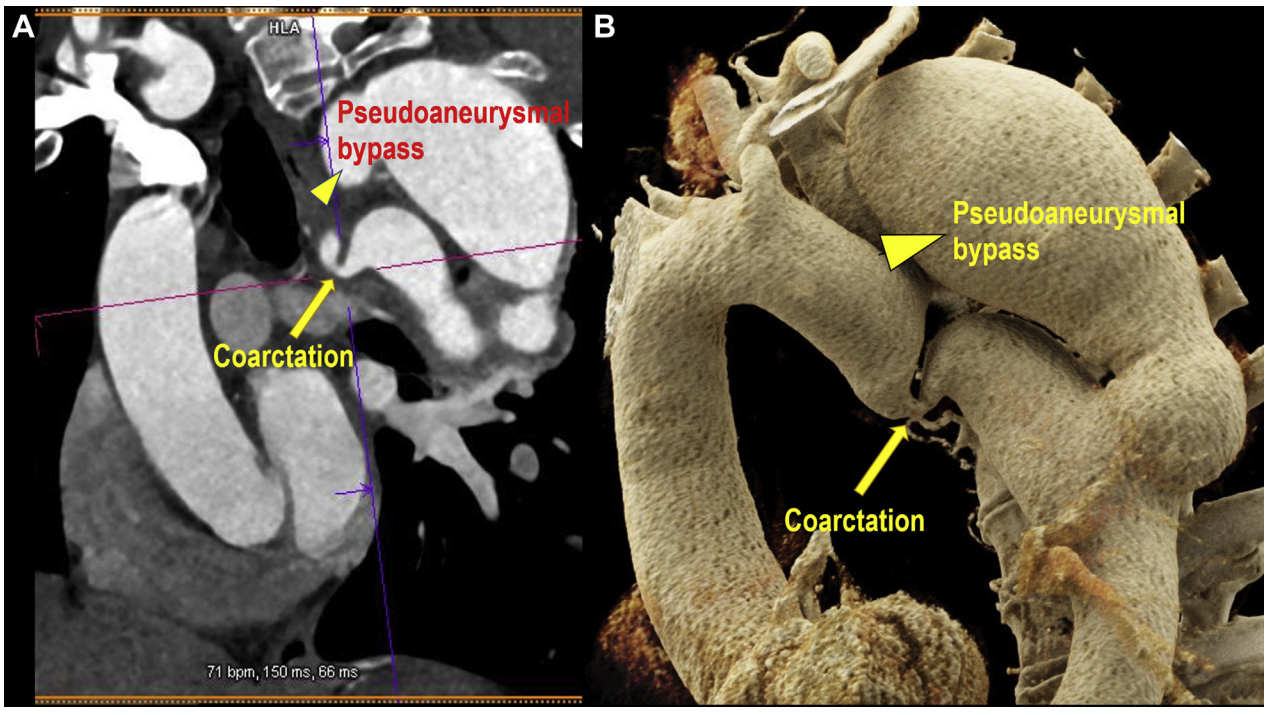
Post-operative computed tomography imaging demonstrated that the overlapping stents had successfully excluded the pseudoaneurysm arising from the LSCA, a patent LCCA to distal LSCA graft, and exclusion of the LSCA pseudoaneurysm connection by the vascular plug. No residual coarctation was visualized.

DISCUSSION

Coarctation of the aorta accounts for 5% to 8% of congenital heart disease, and is often identified and treated during childhood (1). Primary surgical repair of coarctation can be associated with long-term issues such as recurrent coarctation, occurring in 5% to 24% of patients, and late hypertension despite repair (1). Aortic aneurysm and pseudoaneurysm occur in 5% to 10% of patients undergoing coarctation repair and have been reported after every type of coarctation repair (end-to-end, subclavian flap, prosthesis interposition, and patch aortoplasty) (2,3). Late aneurysm formation is particularly prevalent when Dacron patches are used, wherein the rate may increase to 38%; resection of the coarctation ridge may predispose to aneurysm formation (2,3). In our patient, the pseudoaneurysm arose from repair using a "bucket handle" Hemashield vascular bypass graft from the LSCA to the descending thoracic aorta. Pseudoaneurysm of the subclavian artery is uncommon, typically occurring after major trauma or iatrogenic injury.

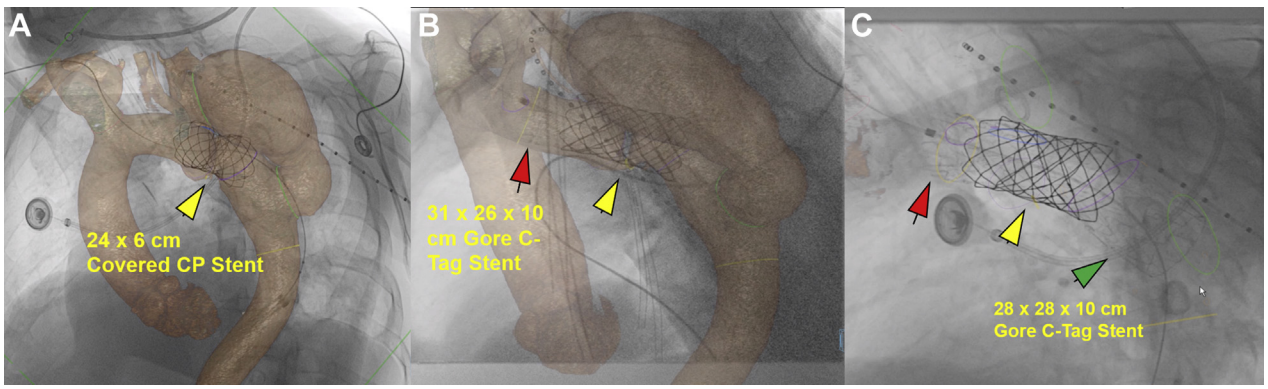
The etiology of aortic dilation, dissection, and pseudo/aneurysm may be related to a generalized arteriopathy because intracerebral aneurysms are seen in up to 10% of patients and early development of coronary artery disease has been described (2,4,5). Hypotheses point to deficiency of microfibrillar proteins, hyperactivity of matrix metalloproteinases and tunica medial degeneration, mutations in genes such as NOTCH1, and developmental abnormalities of neural crest tissue that give rise to muscular arteries of the heart, aortic arch, and cervicocephalic arteries as potential contributors (4,5). Additionally, 70% to 75% of patients with coarctation have concomitant BAV (4), and incidence of aortic valve disease in cases of associated coarctation is higher than that of isolated BAV (2). Furthermore, concomitant BAV and coarctation is more often associated with aortic dilation as compared with isolated BAV or coarctation cases (4). Finally, persistent hypertension and

FIGURE 1 Pseudoaneurysmal Bypass



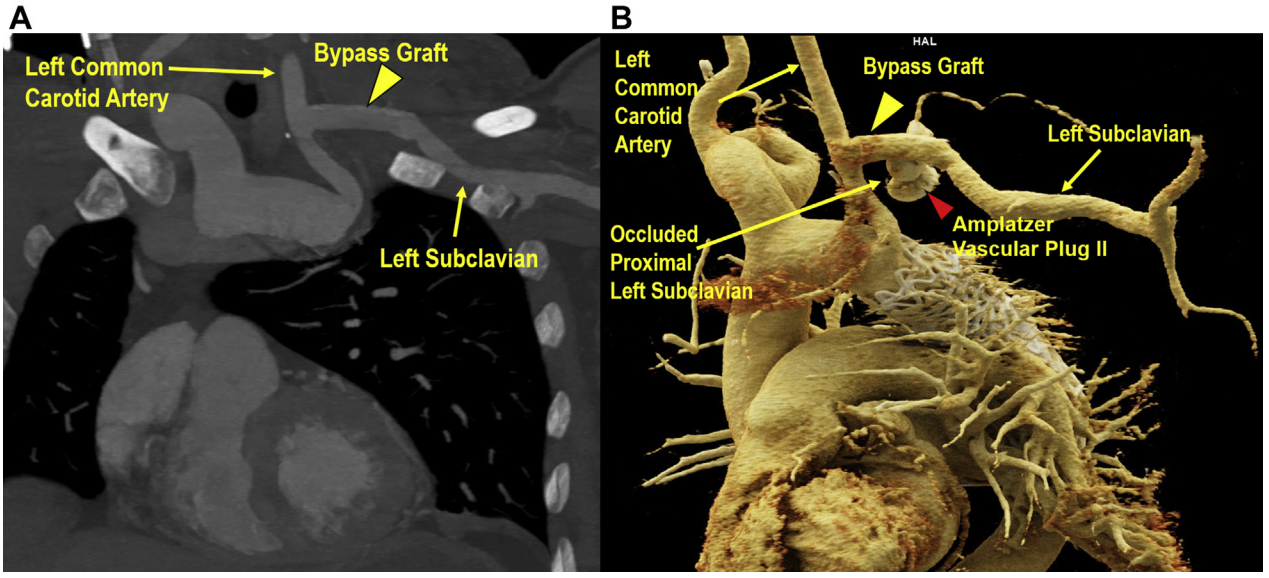
Computed tomography **(A)** and 3-dimensional cinematic computed tomography angiography rendering **(B)** revealed a focally kinked distal aortic arch with >80% stenosis and a minimal coarctation neck diameter of 2.5 mm **(arrow)**. A 5-cm pseudoaneurysmal dilatation of the subclavian bypass was also present **(arrowhead)**.

FIGURE 2 Bypass Exclusion



Under computed tomography-overlay guidance, 24 mm x 6 cm covered CP stent **(yellow arrowhead)** was deployed across the aortic coarctation and left subclavian artery takeoff **(A)**. A 31 mm × 26 mm × 10 cm Gore C-TAG thoracic endovascular aortic repair (TEVAR) stent **(red arrowhead) (B)** was placed and extended by a 28 mm × 28 mm × 10 cm Gore C-TAG TEVAR stent **(green arrowhead) (C)** to exclude the inflow and outflow of the bucket-handle bypass.

FIGURE 3 Post-Procedural Imaging



Computed tomography **(A)** and 3-dimensional computed tomography angiography rendering **(B)** depicting a 16-mm Amplatzer Vascular Plug II **(red arrowhead)** sealing the subclavian attachment of the pseudoaneurysmal graft. A carotid-subclavian bypass **(yellow arrowhead)** was performed after the procedure.

development of late hypertension despite successful coarctation repair may be related to maladaptive changes in the vasculature, although this is not fully understood (3).

Endovascular treatment of recurrent coarctation may be the treatment of choice if anatomy is favorable (4) but the context of underlying arterial pathology must be considered given the risk of aortic wall injury including dissection, rupture, or aneurysm both within and at the margins of the stent (6). The true incidence of aortic aneurysm following intravascular stent therapy remains unascertained with studies reporting rates of 5% to 9% (7). Covered CP stents may reduce but not eliminate risk of aortic wall injury in coarctation therapy (6) and 2 cases of pseudoaneurysm have been reported after covered CP stenting (8). Advances in cardiovascular imaging now provide excellent guidance for complex anatomy such as in our case (9), however, whether or not this improves long-term outcomes has not been demonstrated.

FOLLOW-UP

At 6 months, the patient had resolution of chest pain, hemoptysis, and secondary hypertension with discontinuation of multiple antihypertensive medications.

CONCLUSIONS

Despite successful childhood intervention for coarctation of the aorta, late complications such as hypertension, recurrent coarctation, cerebral aneurysms, aortic valve disease, aortic pseudo/aneurysm, and aortic dissection may present in adulthood, some of which may require high-risk surgery or complex transcatheter intervention. Routine lifelong surveillance and treatment is required.

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ADDRESS FOR CORRESPONDENCE: Dr. C. Huie Lin, Houston Methodist DeBakey Heart & Vascular Center, 6550 Fannin Street, Suite 1901 Smith Tower, Houston, Texas 77030, USA. E-mail: clin@houstonmethodist.org. Twitter: @HuieLin.

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