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CASE REPORT

BEGINNER

CLINICAL CASE

Management of Severe Coarctation of the Aorta and Bilateral Carotid Artery Stenosis

An Interventional Catch-22

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ABSTRACT

We present a case of an adult with concurrent severe aortic coarctation, bilateral carotid artery stenosis, and anomalous right subclavian artery, posing the interventional dilemma of accepting potential cerebral hyperperfusion syndrome vs hypoperfusion ischemic injury. Transcatheter stenting of the aortic coarctation was successfully performed without any neurological deficits. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2023;6:101693) © 2023 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/).

HISTORY OF PRESENTATION

We present the case of a 34-year-old woman from Afghanistan who was advised to present to the hospital with hypertensive crisis. On presentation, her blood pressure was 220/95 mm Hg on the background of known severe resistant hypertension. Examination revealed a significant difference in her left (220 mm Hg) and right arm (90 mm Hg) systolic blood pressure (SBP), with a weak distal right radial pulse. On auscultation, there was an early systolic murmur loudest at the sternal angle with radiation to her back. Altogether, these clinical findings raised the suspicion of aortic coarctation.

LEARNING OBJECTIVES

- To examine the fine physiological balance in aortic coarctation with concurrent carotid artery stenosis.
- To understand the pathophysiology of subclavian steal syndrome in a patient with anomalous vasculature.
- To appreciate the unique interventional dilemma in such patients, balancing the risk of postoperative cerebral hyperemia vs postrepair hypoxia.

PAST MEDICAL HISTORY

Our patient was diagnosed with hypertension at age 12 years, which was subsequently complicated by a stroke with upper and lower limb motor deficits that have since completely resolved. Following her arrival to Australia as a refugee 3 months before, her hypertension was more optimally managed by her new

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ABBREVIATIONS AND ACRONYMS

LV = left ventricular

SBP = systolic blood pressure

general practitioner, who aimed to achieve tighter blood pressure control with multiple antihypertensive agents at maximal doses: telmisartan, hydrochlorothiazide, amlodipine, and bisoprolol. Despite the addition of hydralazine and isosorbide mononitrate, her hypertension remained poorly controlled, and was thus referred for further inpatient investigation.

DIFFERENTIAL DIAGNOSIS

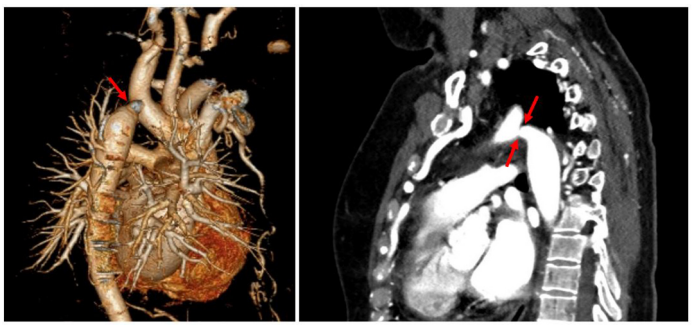
Our patient's presentation was consistent with a new diagnosis of coarctation of the aorta. However, the upper limb pressure gradient, which was higher in the left arm, is reverse of the classical phenotype, suggesting an underlying vascular anomaly. The presence of cerebral aneurysms was also considered in the context of suspected severe aortic coarctation and her history of recurrent headaches and intermittent visual deficits.

INVESTIGATIONS

Computed tomography angiogram of the aorta revealed severe preductal aortic coarctation with critical narrowing and an anomalous right subclavian artery originating as the fourth branch of the arch, distal to the coarctation with a retroesophageal course with no resultant tracheal or esophageal compromise (Figure 1). The majority of flow into the thoracic aorta and right arm appeared to be via a left to right subclavian steal with retrograde flow through the aberrant right subclavian artery (Figure 2), which is supplied by retrograde flow through the right vertebral artery. On transthoracic echocardiography, peak velocity across the stenosis was 5.3 m/s, with a pressure gradient of 112 mm Hg, confirming a severe degree of narrowing. There was no significant left ventricular (LV) hypertrophy (LV mass 75 g/m^2, posterior wall end-diastolic thickness 8 mm, interventricular septal thickness 9 mm) or LV systolic dysfunction (LV ejection fraction 52%).

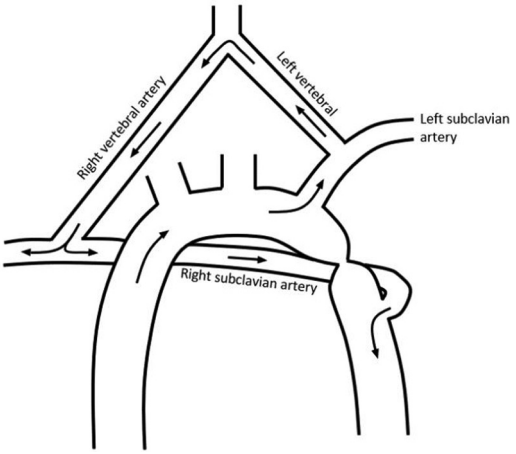
Computed tomography and cardiac magnetic resonance imaging revealed 50% stenosis of the high cervical left internal carotid artery and very severe stenosis of the supraclinoid segment of the intracranial right internal carotid artery. There is a tight stenosis of the origin of the A1 segment of the right anterior cerebral artery. There is no appreciable right posterior communicating artery, in effect making this an isolated intracranial vascular territory, supplied by a very stenotic artery (Figure 3). This was confirmed by magnetic resonance imaging and nuclear medicine perfusion imaging demonstrating a previous right external capsule infarct, with further cerebral perfusion imaging with acetazolamide confirming an area of abnormal cerebral perfusion reserve in the right internal carotid artery territory watershed regions. This raised concerns regarding possible right middle cerebral artery territory ischemia with any precipitous drops in blood pressure.

FIGURE 1 Computed Tomography Angiogram of the Aorta



The **arrows** point to severe preductal aortic coarctation with critical narrowing.

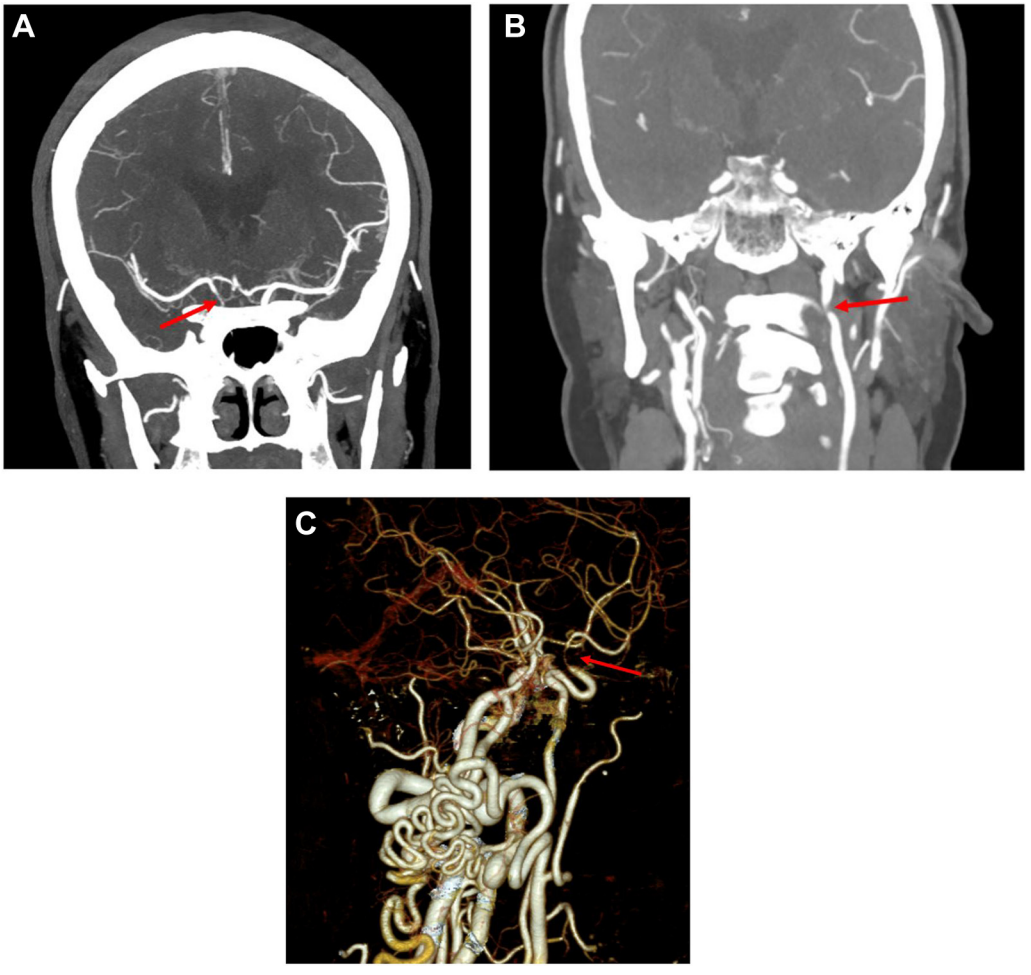
FIGURE 2 Left to Right Subclavian Steal Phenomenon With Retrograde Flow Through the Aberrant Right Subclavian Artery



MANAGEMENT

Following multidisciplinary team discussion, involving surgical, anesthetic, radiology, neurosurgery, and cardiology teams, a decision was made for transcatheter repair of the aortic coarctation, with close operative and postoperative monitoring. In the

FIGURE 3 **Computed Tomography Angiogram Carotid With Circle of Willis**



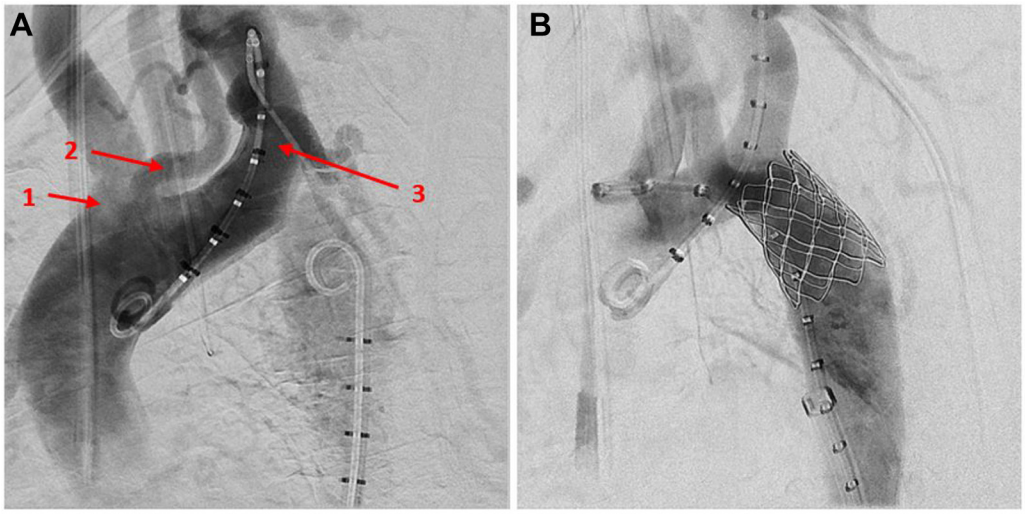
(A and C) Severe stenosis of supraclinoid right internal carotid artery. **(B)** Stenosis (50%) of the left internal carotid artery at the level of C1.

event of any neurological deficit, immediate carotid artery stenting was ready to be initiated.

While awaiting transcatheter intervention, the patient was continued on 6 antihypertensive agents with an SBP target of 140 to 160 mm Hg in the left arm. The stenting procedure was performed with general anesthetic and vascular surgery support. Cerebral oximetry values were maintained within 5% of baseline preinduction values throughout the case. Using a 6-F multipurpose catheter, the aortic coarctation was crossed with a slippery wire, which was then placed in the brachiocephalic artery. The catheter was then exchanged for an Amplatz Extra Stiff Wire, and a long 14-F Mullins sheath was placed across the coarctation. A 39-mm-long CP stent (PFM Medical) was mounted on a 20-mm (D) × 40-mm (L) balloon and was advanced across the sheath and over the coarctation point. Using fluoroscopy, positioning was confirmed and the stent was deployed. There was good contrast flow over the coarctation point and in the left and right subclavian arteries, with no evidence of aortic dissection or perforation (Figure 4).

The patient was transferred postprocedure to the intensive care unit for close monitoring of cerebral perfusion in the setting of bilateral carotid stenosis. Initial higher postoperative SBP targets of 160 to 180 mm Hg were achieved with a noradrenaline infusion for 2 days. On day 3, the patient was successfully weaned off noradrenaline with SBP targets relaxed to 140 to 160 mm Hg. She was discharged home on the fourth postoperative day with all

FIGURE 4 Angiographic Images Before and After Intervention



Aortic coarctation before **(A)** and after **(B)** stent placement. Note the significant change in aortic caliber. (1) Right common carotid, (2) left common carotid, (3) left subclavian artery.

antihypertensive agents ceased and without any neurological deficits.

DISCUSSION

Coarctation of the aorta occurs in approximately 4 of 10,000 births and is an isolated finding in 82% of cases.[1] Its coexistence with other vascular anomalies such as carotid stenoses is even rarer and raises the question of whether repairing coexisting lesions within the same procedure is beneficial.

The coexisting vascular anomalies pose a delicate risk in managing our patient's aortic coarctation. Her anomalous right subclavian artery (1% of all aortic coarctation cases) causes a subclavian steal phenomenon. Although the reversed vertebral arterial flow poses a risk of inadequate flow to her posterior circulation, the left to right subclavian collateralization provides the majority of flow to the thoracic aorta and right upper limb through retrograde flow in the aberrant right subclavian artery (**Figure 2**).

Despite the presence of bilateral carotid artery stenoses, which act as a protective mechanism against the cerebral sequalae of hypertension, her severe resistant hypertension posed significant long-term cardiovascular risk and thus required urgent treatment. Her natural compensations, however, pose an interventional dilemma, because the correction of the aortic coarctation may cause a precipitous drop in cerebral perfusion, magnified by her bilateral stenoses. The choice to rectify the carotid artery stenosis first while the patient remains grossly hypertensive poses the risk of reperfusion intracerebral hemorrhage. The coexistence of all 3 vascular conditions is exceedingly rare, and to our knowledge this is the first reported case in an adult of her age.

Surgical repair or catheter-based stenting is a Class I therapeutic recommendation for adults with hypertension and significant native coarctation of the aorta, defined as upper and lower extremity resting peak-to-peak gradient >20 mm Hg, which is present in this case.[2] The 2018 American College of Cardiology/American Heart Association guidelines state that the presence of systemic hypertension or significant upper and lower extremity pressure gradients support the decision to proceed with intervention.[2] Transcatheter intervention with stenting remains the preferred intervention for native aortic coarctation, with surgical intervention traditionally reserved for cases with features such as aortic aneurysm or hypoplasia of the aortic arch, or following failed catheter intervention.[3]

FOLLOW-UP

Three months following transcatheter intervention of her aortic coarctation, the patient's SBP remains within the target range of 140 to 160 mm Hg without

any antihypertensive agents and she remains free of any neurological deficits.

CONCLUSIONS

We report the successful transcatheter stenting of the aorta in a 34-year-old woman with the coexistence of aortic coarctation, an aberrant right subclavian artery, and severe bilateral carotid artery stenosis, which posed a unique physiological management dilemma. Careful preoperative evaluation by multidisciplinary cardiology, vascular, and neurosurgical teams enabled the successful management of this patient.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS bilateral carotid stenosis, coarctation of the aorta, secondary hypertension, transcatheter interventions