CLINICAL VIGNETTE

A Young Woman with Hypertension

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A 32-year-old woman was referred to cardiology for persistent hypertension. Her blood pressure readings remained elevated despite lifestyle modifications and initiation of antihypertensive medications.

Clinical Presentation and Initial Evaluation

The patient reports hypertension since late adolescence. She was initially told she had "white coat" hypertension. Before she was eventually started on low dose amlodipine with improvement in blood pressure. Evaluation for secondary etiologies of hypertension was unremarkable including screening for renal artery stenosis, hyperaldosteronism, obstructive sleep apnea and pheochromocytoma. On physical exam a systolic cardiac murmur was detected, prompting echocardiogram, which revealed mild concentric left ventricular hypertrophy with normal systolic function and ejection fraction of 60-65%. The aortic valve was bicuspid with mild regurgitation and no stenosis. Flow velocities in the decending aorta were in the normal range. The finding of a bicuspid aortic valve (BAV) led to additional advanced imaging to further delineate the anatomy and assess for an associated aortopathy. Magnetic resonance angiography (MRA) showed coarctation of the aorta, with minimal diameter of 8 mm, and extensive neck, chest, transpleural collateral vasculature (Figure 1). Review of MRA raised suspicion for possible separated aorta and the need for precise anatomical delineation. Computed tomography (CT) scan confirmed the diagnosis of aortic coarctation (Figure 2).

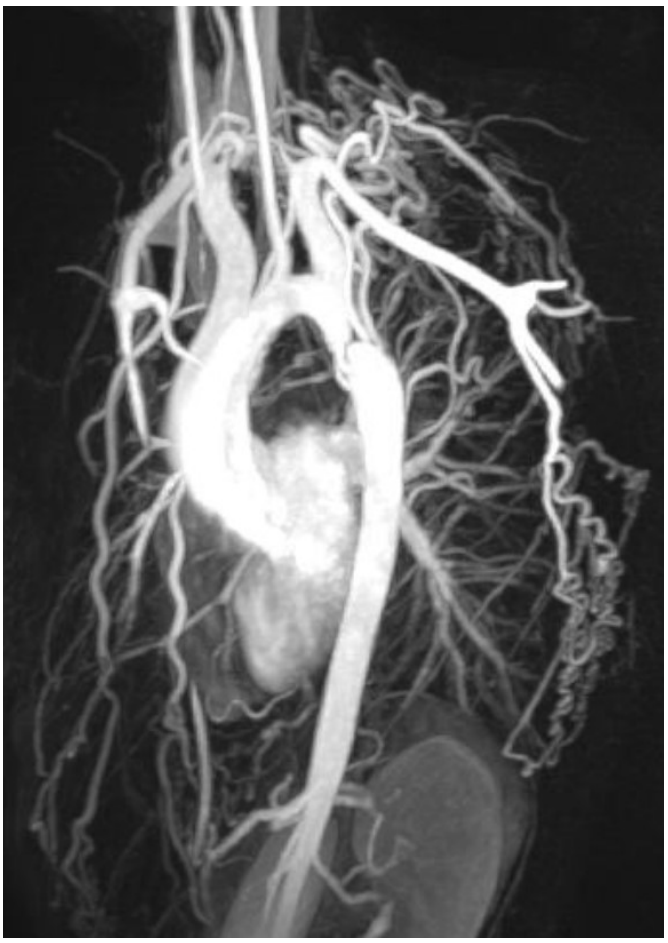

Figure 1.

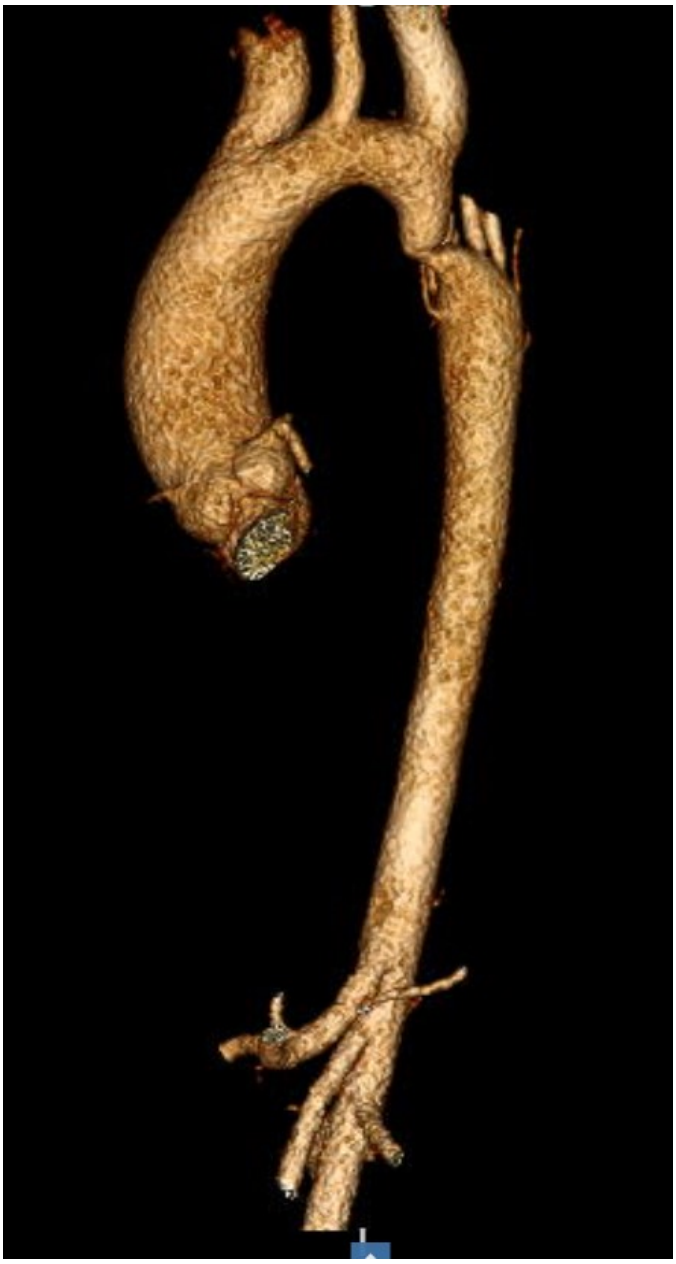

Figure 2.

Pathophysiology

The coexistence of BAV and aortic coarctation represents a complex pathophysiological scenario. BAV, which occurs in 1-2% of the population, is a congenital malformation that can lead to turbulent blood flow, resulting in shear stress on the vessel walls. This abnormal flow pattern, combined with the coarctation of the aorta, contributes to the development and perpetuation of hypertension.[1]

Aortic coarctation is a narrowing of the aorta, typically occurring near the site where the ductus arteriosus inserts during fetal development. This narrowing obstructs blood flow, leading to increased pressure proximal to the coarctation and decreased pressure distal to it. Collateral vessels may develop to maintain blood supply to the lower part of the body, as a compensatory mechanism. Aortic coarctation is found in approximately 6% of patients with BAV.[1]

The increased workload on the heart, due to obstruction and hypertension, can lead to left ventricular hypertrophy. Over time, hypertrophy may progress to heart failure if left untreated. Additionally, the turbulent flow and shear stress on the vessel walls can predispose patients to formation of aneurysms and dissections.

Management

Indications for intervention in patients with coarctation include one of any of the following: a resting gradient across the coarctation greater than 20 mmHg, radiologic evidence of significant collateral circulation, and systemic hypertension or heart failure attributable to coarctation. Percutaneous interventions are increasingly favored due to their less invasive nature, shorter recovery times, and comparable effectiveness to surgical repair. The choice between surgical and percutaneous interventions depends on various factors, including the anatomy of the coarctation, the patient's overall health, and the expertise of the medical team. In certain cases, stenting may be less successful such as in the presence of severe aortic tortuosity or transverse arch hypoplasia.[2] Given the severity of this patient's coarctation, the presence of significant collaterals and suitable anatomy, the decision was made to proceed with percutaneous repair following a collaborative decision-making process involving cardiovascular surgery and interventional cardiology.

The patient was brought to the cardiac catheterization lab. Via femoral arterial access, the coarctation was crossed with a wire and a catheter was subsequently passed. Aortography was performed for stent sizing. Peak to peak gradient across the coarctation was 39 mmHg. A 3.4 cm covered CP stent followed by a Palmaz 3110 stent used as a buttress were deployed to a maximum diameter of 16 mm within the coarctation resulting in no residual gradient.

Post-procedure, the patient did well. She was started on a low dose aspirin and noted significant improvement in blood pressure. The need for antihypertensive medications diminished, allowing for a gradual reduction under close medical supervision. Regular follow-up appointments were scheduled to monitor her cardiovascular health, including valve function, blood pressure, and any potential complications.

Conclusion

This young woman with a bicuspid aortic valve underscores the importance of a comprehensive approach to the diagnosis and management of complex cardiovascular disease. Importantly, whenever a bicuspid aortic valve is diagnosed, evaluation for associated aortopathy is needed.[3] The integration of advanced imaging modalities, collaborative decision-making, and minimally invasive interventions has the potential to optimize outcomes and improve the quality of life for individuals with

these congenital anomalies. Staying abreast of evolving diagnostic and therapeutic strategies is needed to provide optimal care for these complex patients.

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