

CASE REPORT

Unraveling Uncontrolled Hypertension in a Young Female with Atypical Aortic Coarctation: A Case Review.

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Received 10/02/2024**Accepted** 02/04/2024**First Published** 01/06/2024**Abstract**

Background: Atypical aortic coarctation can lead to significant complications such as uncontrolled hypertension, particularly in young patients. This case highlights the importance of thorough evaluation and timely intervention.

Case Presentation: A 25-year-old female, non-diabetic, presented with uncontrolled hypertension despite a regimen of multiple antihypertensive medications. On examination, she had a blood pressure of 160/90 mmHg in both upper limbs, feeble bilateral femoral pulses, and absent distal pulses. Baseline investigations, including echocardiography, showed an ejection fraction (EF) of 62% with a tri-leaflet aortic valve. Notably, imaging revealed narrowing of the distal descending thoracic aorta with a pressure gradient of 49 mmHg, severe concentric left ventricular hypertrophy (LVH), and grade II diastolic dysfunction.

Results: The patient successfully underwent percutaneous endovascular stenting with a covered stent (Aortic Be Graft, 14x49 mm) for aortic coarctation. Post-procedural hemostasis was achieved after addressing significant oozing. A follow-up aortogram indicated total occlusion of the common and superficial femoral arteries, which was treated with percutaneous transluminal angioplasty (PTA). Discharged on the fourth postoperative day, the patient was prescribed dual antiplatelet therapy (aspirin and clopidogrel, 75 mg each). At one-week follow-up, she reported no complications and maintained stable blood pressure (100/60 mmHg to 120/80 mmHg) without antihypertensive medications. A repeat echocardiogram confirmed a zero-pressure gradient across the descending aorta, demonstrating the intervention's success. The patient expressed satisfaction with her treatment and recognized the importance of medication adherence for long-term health.

Conclusion: This case highlights the importance of early diagnosis and appropriate intervention in young patients with uncontrolled hypertension. Atypical coarctation of the aorta should be considered a potential etiology, and percutaneous endovascular stenting is a safe and effective treatment option.

Keywords

Atypical Coarctation of aorta, percutaneous endovascular stenting, ProGlide, PTA balloon, follow-up.



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Introduction

Atypical aortic coarctation (AAC) is a rare and complex cardiovascular condition characterized by narrowing of the aorta, particularly in the descending thoracic segment. Unlike the more common forms of coarctation that typically occur distal to the left subclavian artery, AAC can manifest in various anatomic configurations, complicating diagnosis and management. This condition can lead to significant cardiovascular complications, including uncontrolled hypertension, which can be particularly problematic in young individuals¹.

In cases of AAC, patients often present with hypertension in the upper extremities and diminished or absent pulses in the lower extremities due to the disruption of normal blood flow. This differential presentation can sometimes lead to misdiagnosis, with conditions such as arteritis being suspected instead². The suspicion of arteritis arises from the common clinical scenario of refractory hypertension in a young patient, especially when accompanied by systemic symptoms such as exertional dyspnea or claudication.

The advent of advanced imaging techniques, such as CT angiography, has greatly enhanced our ability to diagnose vascular anomalies like AAC. This imaging modality allows for detailed visualization of the aorta and its branches, facilitating the identification of coarctations and other structural abnormalities³. In the case presented, CT angiography revealed significant narrowing of the descending thoracic aorta, thereby confirming the diagnosis of coarctation rather than arteritis.

This case highlights the importance of considering atypical aortic coarctation in the differential diagnosis for young patients presenting with refractory hypertension, particularly when initial imaging studies suggest normal findings in other vascular territories. Prompt identification and intervention, such as endovascular stenting, are crucial for preventing long-term complications

associated with uncontrolled hypertension and improving patient outcomes.

Case Presentation

A 25-year-old non-diabetic female presented with uncontrolled hypertension, remaining symptomatic despite a multi-drug regimen that included spironolactone, hydralazine, nifedipine, and nebivolol over the past three months. She reported experiencing exertional dyspnea and intermittent claudication for the last 3-6 months.

On examination, bilateral femoral pulses were feeble, and distal pulses were absent, with blood pressure recorded at 160/90 mmHg in both upper limbs. An electrocardiogram revealed left ventricular hypertrophy. The patient's family history was non-contributory, and there was no history of smoking. Baseline laboratory investigations were unremarkable, and she remained clinically stable during her hospital admission.

Echocardiography showed a tri-leaflet aortic valve and normal dimensions for the ascending aorta and aortic arch (25 mm). However, it revealed coarctation in the distal descending thoracic aorta, with a significant pressure gradient of 49 mmHg. The left atrium was dilated (46 mm) but without evidence of thrombus, and the patient exhibited severe concentric left ventricular hypertrophy (LVH) with preserved systolic function (ejection fraction of 62%) and grade II diastolic dysfunction.

To further investigate, a CT angiogram was performed, confirming normal findings in the ascending aorta, aortic arch, and great vessels. However, it revealed significant narrowing at the T9-T10 level of the descending thoracic aorta, measuring 40 mm in length, with a proximal diameter of 16 mm and a distal diameter of 14 mm. These findings led to the diagnosis of atypical coarctation of the aorta. Consequently, percutaneous endovascular stenting was planned as the definitive management approach.

Diagnostic Assessment

The diagnostic approach for this case involved a thorough clinical evaluation and advanced imaging. A 25-year-old female with refractory hypertension presented with blood pressure of 160/90 mmHg in both upper limbs, weak femoral pulses, and absent distal pulses. An ECG indicated left ventricular hypertrophy, while echocardiography confirmed severe concentric LVH, grade II diastolic dysfunction, and a pressure gradient of 49 mmHg across the distal descending aorta, suggesting coarctation.

Therapeutic Intervention

The therapeutic intervention commenced with dual access via the right radial and right femoral arteries. An 11F sheath was introduced through the right femoral artery, allowing for an aortogram to be performed via the right radial sheath. This imaging was essential to assess the narrowest diameter, length of the coarctation, and the diameters above and below the coarctation, guiding the selection of the appropriate stent.

Invasive measurements indicated a pressure gradient of 73 mmHg across the coarctation. A covered stent (Aortic Be Graft) sized 14x49 mm was then carefully positioned and deployed at a nominal pressure of 6 atm. Initially, transient underexpansion was observed, but after 10-15 seconds, there was gradual improvement. A repeat aortogram confirmed the underexpansion, prompting the need for post-dilation using a 14x40 mm non-compliant balloon at 12 atm. This successfully achieved full stent expansion and a zero-pressure gradient across the coarctation.

Following stent placement, ProGlide was utilized for hemostasis; however, significant oozing occurred during deployment, leading to the decision to place an additional ProGlide to achieve hemostatic control. As per standard protocols, an aortogram was performed at the aortic bifurcation to evaluate the access site. This revealed total occlusion of both the common femoral artery and the superficial femoral artery, which was being supplied through collateral circulation.

To address this complication, a crossover sheath was introduced via contralateral femoral access, allowing for the deployment of percutaneous transluminal angioplasty (PTA) balloons sized 4x100 mm and 5x40 mm. A final check injection confirmed the restoration of normal flow in the femoral artery. The patient was subsequently transferred to the ICU, initiated on anticoagulation therapy, and all antihypertensive medications were discontinued.

Follow-Up and Outcomes

The patient was discharged on the fourth postoperative day with a prescription for dual antiplatelet therapy, consisting of aspirin and clopidogrel at a daily dosage of 75 mg each. At the one-week follow-up, she demonstrated a stable clinical condition, reporting no episodes of chest pain, intermittent claudication, or other complications.

During the two-week follow-up, the patient continued to exhibit excellent functional capacity, with no chest pain or dyspnea upon exertion. Her blood pressure readings consistently ranged between 100/60 mmHg and 120/80 mmHg, all achieved without the need for antihypertensive medications. A repeat echocardiogram showed a zero-pressure gradient across the descending aorta, confirming the success of the intervention.

The patient expressed high satisfaction with her treatment outcomes and demonstrated a comprehensive understanding of the importance of adherence to her prescribed medication regimen for optimal long-term health.

Discussion

In young patients presenting with uncontrolled hypertension and significant discrepancies in blood pressure between the upper and lower limbs, the suspicion of arteritis often arises. In this case, CT angiography was pivotal in identifying the underlying cause. The patient was diagnosed with atypical coarctation of the aorta, a rare condition that affects a small percentage of individuals with aortic coarctation. This variant typically extends beyond the usual site at the aortic isthmus and can

be associated with conditions such as arteritis or fibromuscular dysplasia; however, these were ruled out based on normal findings in the ascending aorta, aortic arch, great vessels, and the rest of the vascular system.

Clinically, atypical coarctation is characterized by hypertension in the upper limbs and diminished or absent pulses in the lower limbs,^{4,5} aligning with the symptoms observed in this patient. Initial symptomatic management primarily involves antihypertensive medications, but definitive treatment options include percutaneous transluminal angioplasty with or without endovascular stent placement, or surgical intervention.^{4,5} Due to its favorable safety profile, percutaneous endovascular stenting is often preferred over surgical options^{6,7}.

In this case, a covered stent was chosen for treatment. The selection of stent type is crucial; available options include the Cheatham Platinum (CP) stent and covered stents. The covered stent deployed successfully addressed the patient's condition, underscoring the effectiveness of this intervention for atypical aortic coarctation.

Conclusion

In young patients presenting with uncontrolled hypertension, atypical aortic coarctation should be considered as a potential diagnosis. Percutaneous endovascular stenting is generally safer than surgical options for treating coarctation of the aorta, making it a preferred intervention. Compliance with guideline-directed medical therapy (GDMT) is crucial for achieving optimal patient outcomes. Additionally, a follow-up CT angiogram should be conducted at six months to evaluate for complications such as pseudoaneurysms, concealed perforation, or stent under-expansion.

Learning points

- Timely identification of underlying conditions, such as atypical aortic coarctation, is critical in young patients presenting with uncontrolled hypertension.
- This technique serves as a safer alternative to traditional surgical interventions for treating aortic coarctation, minimizing patient risk and recovery time.
- Continuous monitoring is vital to evaluate patient outcomes and address any complications that may arise post-procedure.
- Following angioplasty, strict adherence to prescribed medications is essential for maintaining stable health and preventing the recurrence of hypertension.

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Figure/Video

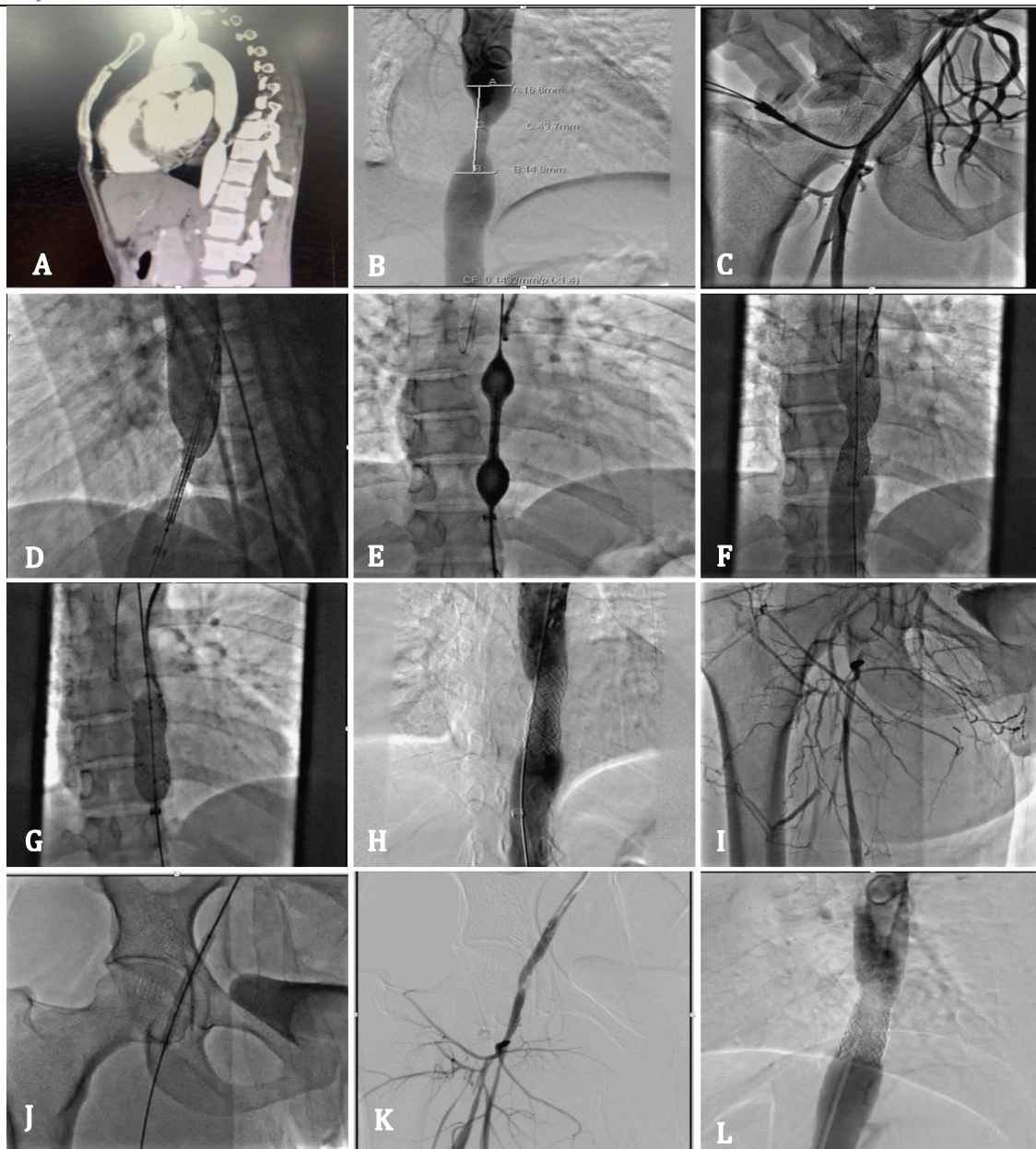


Figure 1:

A) CT Angiography

B) Aortogram done with dimensions measured under fluoroscopy via right radial access.

C) Ultrasound Guided Femoral Access.

D) Stent Placement

E) Stent Deployment and

F) Under-expansion of stent.

G&H) Post Dilatation using NC Balloon with fully expanded stent.

I) Aortogram at aortic bifurcation showing total occlusion of CFA and SFA filling from collaterals.

J) PTA balloon.

K) Some residual occlusion post dilatation (most likely thrombotic).

L) Final check injection of stent