CATHALOGUE

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CASE REPORT

A Case of Aortic Stenosis & Coarctation of Aorta in an Adult

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Abstract

Background: Aortic coarctation, a congenital heart defect characterized by narrowing of the aorta, typically manifests and is addressed during childhood. Often, aortic coarctation coexists with other cardiac anomalies such as aortic stenosis and bicuspid aortic valve, complicating its clinical presentation and management.

Case Presentation: Herein, we present the case of a 54-year-old man who was admitted to our cardiology unit due to the sudden onset of chest heaviness, dyspnea, and palpitations. Upon thorough examination and diagnostic workup, the patient was diagnosed with aortic coarctation accompanied by aortic valve stenosis. Given the severity of the condition and its potential complications, a multidisciplinary approach was undertaken to address both issues.

Management: In the first stage of management, the patient underwent successful aortic valve replacement to alleviate the symptoms associated with aortic valve stenosis. Subsequently, attention turned to treating the aortic coarctation. Due to the complexity of the case and the need for precise intervention, a staged approach was adopted. The coarctation was addressed in a separate procedure following the initial valve replacement. This sequential approach ensured optimal management of both conditions while minimizing the risk of complications.

Conclusion: This case highlights the importance of recognizing and managing concurrent cardiac pathologies in patients with aortic coarctation. A comprehensive evaluation and a multidisciplinary approach are essential for successful treatment outcomes. The staged management strategy employed in this case effectively addressed both aortic valve stenosis and aortic coarctation, underscoring its utility in managing complex congenital heart diseases in adult patients.

Keywords

Aortic valve pathology, aortic coarctation, aortic valve stenosis, case report

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Introduction

Coronary bifurcation lesions present a unique Coarctation of the aorta, a congenital vascular anomaly characterized by a narrowing of the aortic lumen, is typically identified and addressed during childhood or early adulthood. However, there is a growing body of literature documenting cases where individuals are first diagnosed with coarctation of the aorta in later stages of life, particularly beyond the age of 50. This phenomenon underscores the importance of considering congenital heart diseases, even in older adults, and highlights the potential challenges in diagnosing and managing such conditions in this demographic[1].

While coarctation of the aorta is relatively uncommon in older adults, its diagnosis in this age group poses unique clinical and therapeutic implications. In contrast to cases identified in childhood, where the focus is often on early detection and intervention to mitigate long-term complications, the presentation of coarctation of the aorta in older adults may be more insidious, leading to delayed diagnosis and treatment initiation. This delay can result in the progression of associated cardiac pathologies and an increased risk of cardiovascular events[2].

Furthermore, coarctation of the aorta is frequently associated with other congenital heart diseases, such as bicuspid aortic valve and aortic valve stenosis, complicating its clinical presentation and management. The presence of multiple cardiac anomalies necessitates a comprehensive diagnostic approach and a tailored treatment plan to address all underlying pathologies effectively.

In the context of aortic valve stenosis, a common coexisting condition with coarctation of the aorta, the management strategy may involve aortic valve replacement to alleviate symptoms and prevent further deterioration of cardiac function. However, the presence of aortic coarctation introduces additional considerations, as the narrowed aortic segment may impact surgical planning and outcomes[3,4].

In this backdrop, we present the case of a 54-year-old man who was initially diagnosed with aortic valve stenosis and subsequently found to have coarctation of the aorta. Through this case, we aim to underscore the importance of considering congenital heart diseases, even in older adults, and emphasize the need for a comprehensive diagnostic evaluation and a multidisciplinary approach to effectively manage these complex conditions.

Case Presentation

We present the case of a 54-year-old man with a history of intermittent dyspnea classified as level II-III according to the New York Heart Association (NYHA) classification over the past six months. He was admitted to our hospital due to chest discomfort, accompanied by acute dyspnea classified as level III-IV on the NYHA scale. On clinical examination, diminished pulses were noted, with blood pressure measuring 110/79 mmHg in the upper limbs and 100/70 mmHg in the lower limbs, indicating a 10 mmHg difference. Auscultation over the aortic area revealed a harsh meso-telesystolic crescendo-decrescendo murmur rated as 4/6, radiating to the carotids, along with abolition of the second heart sound (B2) and signs suggestive of congestive left heart failure. Pulses were palpable in both the upper and lower extremities.

Electrocardiography (EKG) displayed sinus rhythm at a rate of 91 beats per minute, with a normal axis and evidence of left ventricular hypertrophy, as indicated by a Corenell index of 35 mm. Chest radiography demonstrated interstitial edema, with the disappearance of the aortic knob and rib notching. Echocardiography revealed normal systolic function, with an ejection fraction of 60%. The aortic valve exhibited significant calcification with three cusps, and hemodynamic assessment revealed a peak transvalvular pressure gradient of up to 129/88 mmHg and a mean transvalvular pressure gradient. The valve area was notably reduced to 0.6 cm^2 (0.33 cm^2/m^2 of body surface area). Sub-sternal imaging did not reveal an enlarged ascending aorta or subclavian artery. Cardiac catheterization demonstrated normal

coronary arteries. Computed tomography (CT) imaging revealed post-ductal occlusion of the descending aorta with a dilated segment post-coarctation, with no significant collateral formation observed.

After consultation with the heart team, it was decided to address the aortic stenosis through surgical intervention. Aortic valve replacement was performed using a bileaflet mechanical valve sized 21 mm. Arterial return was facilitated by cannulation of both the ascending aorta and the right iliac artery, while a single two-stage venous cannula was inserted into the right atrium. Extracorporeal circulation was initiated, with a cross-clamping time of 86 minutes and a total bypass time of 120 minutes. The patient's postoperative course was uneventful, and he was discharged from the hospital on the eighth postoperative day.

Surgical repair of the coarctation was planned as a subsequent stage of treatment; however, the patient declined. One month following the aortic valve replacement, the patient reported a significant clinical improvement, with resolution of chest pain and dyspnea. Echocardiographic assessment revealed a decrease in both peak and mean transvalvular gradients (6.59/2.52 mmHg), indicating successful resolution of the aortic valve stenosis.

Diagnostic Assessment

The diagnostic journey of our patient involved a meticulous and thorough assessment integrating clinical findings with a range of imaging modalities. Initially, the clinical evaluation revealed symptoms consistent with aortic stenosis, including chest discomfort, dyspnea, and diminished pulses, prompting further investigation. Transthoracic echocardiography emerged as a cornerstone in confirming the diagnosis of aortic stenosis, offering detailed insights into the structure and function of the aortic valve, particularly highlighting the severity of stenosis. Meanwhile, electrocardiography (ECG) provided valuable information regarding cardiac rhythm and evidence of left ventricular hypertrophy, though direct confirmation of aortic stenosis was not achieved.

The diagnostic process further expanded with the utilization of a CT aortogram, which proved instrumental in anatomically characterizing the aorta and its adjacent structures. This imaging modality unveiled post-ductal occlusion of the descending aorta and a dilated segment post-coarctation, contributing vital information to the diagnostic puzzle. Additionally, coronary angiography, initially intended for coronary artery assessment, unexpectedly revealed the coarctation of the aorta. As the guide wire encountered resistance crossing the focal stenosis in the descending aorta, the presence of coarctation became evident, emphasizing the importance of comprehensive imaging evaluation in complex cardiac cases.

Despite the incidental nature of the coarctation discovery, its identification was pivotal, as it added a layer of complexity to the diagnostic and therapeutic approach. Thankfully, the hospital's well-equipped facilities facilitated this comprehensive evaluation, mitigating diagnostic challenges and ensuring prompt management. In summary, the collaborative integration of clinical examination and various imaging modalities enabled an accurate diagnosis, guiding tailored management strategies for addressing both aortic stenosis and coarctation of the aorta in our patient's complex cardiac presentation.

Therapeutic Intervention

Following the diagnosis of aortic valve stenosis and coarctation of the aorta, our patient underwent surgical intervention aimed at addressing the aortic valve pathology and subsequent anticoagulant therapy for postoperative management.

1The patient underwent surgical replacement of the diseased aortic valve with a metallic prosthetic valve. This procedure, performed under general anesthesia, involved excision of the calcified native valve and implantation of the prosthetic valve. A bileaflet mechanical valve, sized appropriately for the patient's anatomy, was selected to ensure

optimal hemodynamic function. The surgical approach included cannulation of the ascending aorta and right iliac artery for arterial return, with a single two-stage venous cannula inserted into the right atrium to facilitate venous drainage. Extracorporeal circulation was initiated, allowing for cardiac bypass during valve replacement. The duration of cross-clamping and bypass was carefully monitored to minimize perioperative complications. Following successful valve replacement, the patient's postoperative course was closely monitored for any signs of complications, with appropriate supportive measures implemented as needed.

Postoperatively, the patient was initiated on anticoagulant therapy to prevent thromboembolic complications associated with the metallic prosthetic valve. Vitamin K antagonist agents, such as warfarin, were chosen as the primary anticoagulant therapy. Warfarin inhibits the synthesis of vitamin K-dependent clotting factors, thereby reducing the risk of thrombus formation on the prosthetic valve surface. The therapy was carefully monitored through periodic assessment of international normalized ratio (INR) levels to ensure it remained within the therapeutic range. Regular monitoring and adjustment of warfarin dosage were essential to achieve and maintain the target INR, typically between 2.5 and 3.5, depending on individual patient factors and prosthesis type.

Following discharge from the hospital, the patient underwent regular follow-up appointments to assess the efficacy and safety of anticoagulant therapy, monitor prosthetic valve function, and evaluate overall cardiac status. Periodic INR monitoring allowed for timely adjustment of warfarin dosage to maintain therapeutic anticoagulation while minimizing the risk of bleeding or thromboembolic events. Additionally, echocardiographic surveillance enabled ongoing assessment of valve function and detection of any potential complications, facilitating prompt intervention as needed.

Follow-up and Outcomes

During each follow-up visit, the patient underwent comprehensive assessments, including clinical evaluation and echocardiography. Clinical assessment involved monitoring for any new symptoms or complications related to the surgical intervention or anticoagulation therapy. Echocardiography played a crucial role in assessing the function of the prosthetic aortic valve, detecting any abnormalities or dysfunction, and evaluating overall cardiac performance.

The patient's adherence to anticoagulation therapy, particularly with warfarin, was closely monitored during follow-up visits. International normalized ratio (INR) levels were regularly measured to ensure that the patient maintained therapeutic anticoagulation within the target range. Any deviations from the target INR range were promptly addressed through adjustments in warfarin dosage, guided by clinical guidelines and recommendations.

Throughout the follow-up period, the patient experienced no adverse outcomes related to the surgical intervention or anticoagulation therapy. The absence of complications such as thromboembolic events, bleeding episodes, or prosthetic valve dysfunction was a testament to the effectiveness of the treatment approach and the patient's diligent adherence to therapy.

Echocardiography served as a valuable tool for assessing the outcomes of the surgical intervention and monitoring the long-term function of the prosthetic aortic valve. Serial echocardiographic examinations provided insights into valve hemodynamics, prosthetic valve integrity, and overall cardiac function. Any changes or abnormalities detected during echocardiographic assessments were promptly addressed, ensuring timely intervention and optimization of therapeutic outcomes.

Discussion

Coarctation of the aorta, a congenital vascular malformation characterized by narrowing of the thoracic aorta after the left subclavian artery,

represents a significant subset of congenital heart diseases, accounting for approximately 5-8% of cases. Often, it coexists with other congenital heart anomalies, such as atrial septal defect, ventricular septal defect, patent ductus arteriosus, or bicuspid aortic valve, with the latter being the most commonly associated condition, found in an estimated 50 to 85% of cases. Studies have shown a high prevalence of aortic valve disease, especially in patients with aortic coarctation, further emphasizing the importance of comprehensive cardiac evaluation in these individuals[5].

While typically diagnosed during childhood, approximately 10% of patients remain asymptomatic until adulthood, underscoring the need for continued vigilance in assessing cardiac health throughout the lifespan. Symptoms, when present, vary in severity and may include headache, shortness of breath, abdominal angina, and claudication, along with signs of hypertension and characteristic findings on clinical examination such as blood pressure gradient between upper and lower limbs, weak or absent pulses in the lower extremities, and systolic para sternal heart murmur[6,7].

Unfortunately, diagnosis often occurs only after complications arise, such as aneurysm rupture, aortic dissection, accelerated coronary artery disease, stroke, or congestive heart failure, highlighting the importance of early detection and intervention. The increased incidence of aortic valve problems associated with coarctation of the aorta is likely attributable to hypertension and the mechanical stress imposed by the coarctation gradient on the aortic valve and aortic wall, particularly in the presence of a bicuspid valve[8,9].

Echocardiography remains a cornerstone in diagnosing and assessing the severity of coarctation, as well as evaluating associated cardiac anomalies. However, cardiovascular magnetic resonance and computed tomography are preferred non-invasive techniques for comprehensive evaluation of the entire aorta in adults[10].

Surgical treatment of coarctation of the aorta has evolved significantly, with endovascular approaches emerging as the preferred and safest option. When coarctation coexists with significant aortic stenosis, surgical challenges arise, necessitating careful consideration of the optimal treatment strategy. Various surgical techniques, including simultaneous correction of coarctation and aortic valve replacement, or staged approaches, offer viable options, each with its advantages and potential complications.

In our patient's case, asymptomatic until recent onset dyspnea and angina attributed to aortic valve degeneration, a two-step correction approach was adopted. A successful aortic valve replacement was performed initially, followed by the patient's refusal of surgical treatment for coarctation. During the six-month follow-up period, the patient remained asymptomatic, underscoring the effectiveness of the chosen treatment strategy and the importance of ongoing monitoring for optimal long-term outcomes.

Conclusion

Managing coarctation of the aorta in conjunction with aortic valve disease presents a significant clinical challenge. The optimal approach—whether a one-step or two-step procedure—is contingent upon various factors, including the patient's individual condition, the expertise of the medical team, and the availability of advanced technical resources. It is imperative to carefully weigh these considerations when formulating a treatment plan, ensuring that the chosen approach maximizes the likelihood of successful outcomes while minimizing procedural risks. Ultimately, a multidisciplinary approach, guided by the collective expertise of the heart team, is essential in navigating the complexities of coarctation of the aorta and aortic valve disease, with the goal of providing optimal care tailored to the unique needs of each patient.

Learning points

Consideration of Coarctation of Aorta in Adults: While coarctation of the aorta is less common in adults compared to children, it should be included in the differential diagnosis, especially in individuals

presenting with resistant hypertension. Clinicians should maintain a high index of suspicion for coarctation of the aorta, particularly in patients with uncontrolled blood pressure despite appropriate medical management.

Association with Congenital Cardiac Pathologies: Coarctation of the aorta frequently coexists with other congenital cardiac pathologies, such as atrial septal defect, ventricular septal defect, aortic valve disease (particularly bicuspid aortic valve), and patent ductus arteriosus. Comprehensive evaluation, including echocardiography, is essential to assess for the presence of these associated anomalies, as they may impact treatment decisions and outcomes.

Challenges in Management: Managing coarctation of the aorta in conjunction with aortic valve disease poses significant challenges. The decision-making process should involve consultation with a multidisciplinary cardiac team, including cardiologists, cardiac surgeons, and other specialists, as well as discussions with the patient and their family members. This collaborative approach ensures that treatment plans are tailored to the individual patient's needs and preferences while considering the complexities of managing both conditions simultaneously.

Two-Stage Management Approach: In many cases, a two-stage approach is employed for the management of coarctation of the aorta with concomitant aortic valve disease. This staged approach allows for sequential correction of each condition, minimizing perioperative risks and optimizing outcomes. The decision to pursue a two-stage approach should be guided by careful assessment of the patient's clinical status, anatomical considerations, and procedural risks, with input from the cardiac team.

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Figure/Video

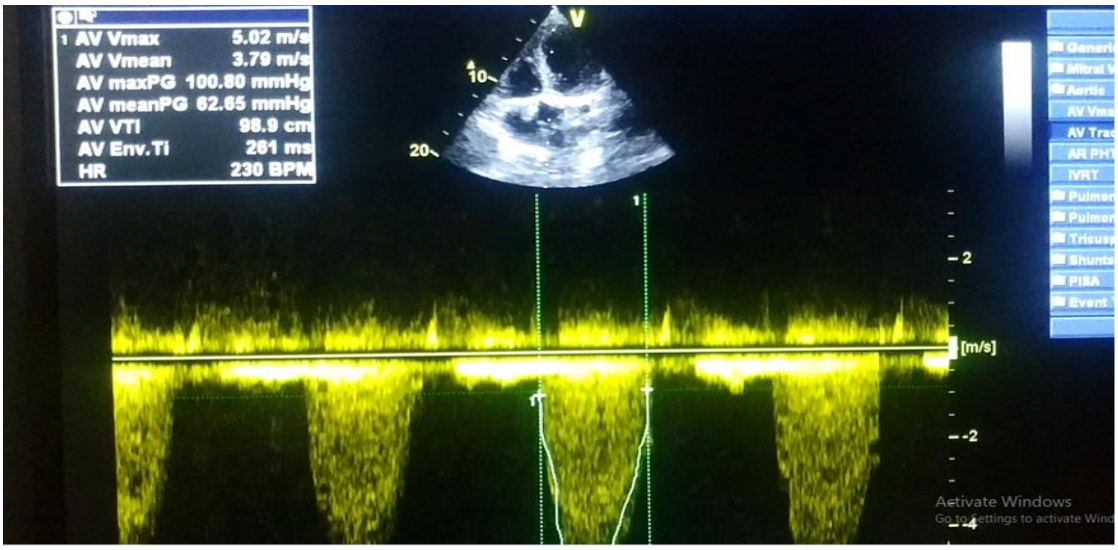

Figure 1: Continuous aortic Doppler showing the peak transvalvular pressure gradient and mean transvalvular pressure gradient respectively of 129/88 mmHg.

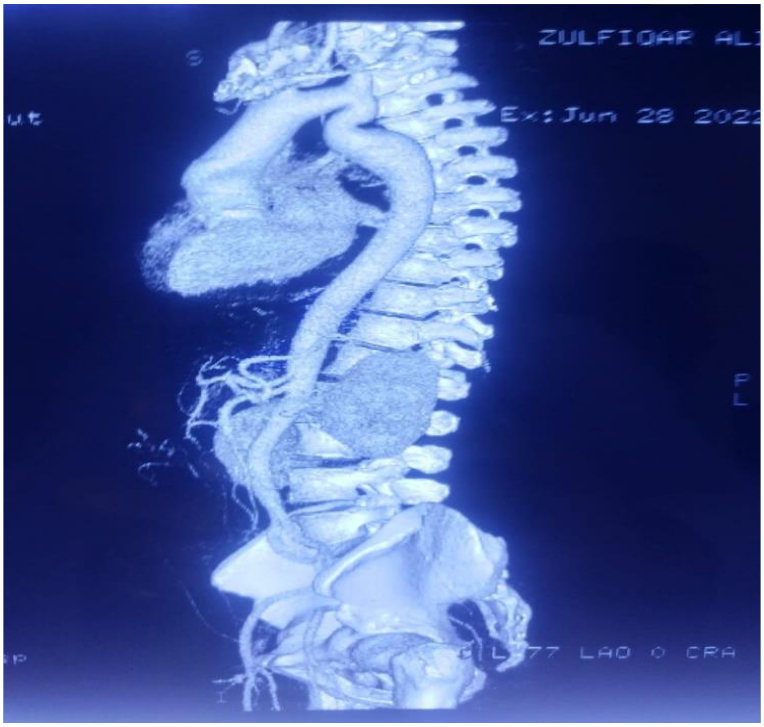

Figure 2: Aortic CT images showing a very tight narrowing of downstream of the descending aorta.