

Totally Occluded Coarctation of the Aorta in a Young Adult

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Abstract

We report the case of a 20-year-old female patient presenting with a totally occluded coarctation of the aorta. The patient was admitted for the evaluation of resistant grade III high blood pressure. The physical examination revealed a blood pressure gradient between the upper and lower limbs, absent femoral pulses, a diffuse continuous murmur over the chest, and hyperpulsatility of the carotid arteries and sternal notch. A transthoracic echocardiography outlined a narrowing in the caliber of the isthmus of the aorta without acceleration of flow or gradient. Thoracic CT angiography revealed a complete aortic coarctation with interruption of continuity between segment 3 and the descending aorta.

Keywords

Coarctation of Aorta, Complete Aortic Occlusion, High Blood Pressure, Adult

1. Introduction

Coarctation of the aorta (CoA) is a congenital heart condition that can go unnoticed until adulthood. Ten to 15% of aortic coarctations are diagnosed during adolescence or adulthood [1]. The totally occluded form, characterized by an interruption in the continuity of the aorta, is rare, with only a few cases described [2] [3]. Its clinical presentation does not differ significantly from that of tight CoA. It is suspected by the presence of high blood pressure (HBP) associated with diminished femoral pulses, contrasting with echocardiographic finding [2]. Cross-sectional imaging, including CT scans and magnetic resonance imaging, is

essential for confirming the diagnosis. We present the case of a 20-year-old young woman with coarctation of aorta with complete aorticocclusion.

2. Clinical Presentation

It was about a 20-year-old female, referred for the evaluation of resistant grade III high blood pressure. She had been under follow-up for two years and was on a quadritherapy regimen, which included an angiotensin-converting enzyme inhibitor, a calcium channel blocker, a diuretic, and a beta-blocker. She reported no specific symptoms. Coarctation of the aorta was suspected due to systolic-diastolic hypertension of 193/127 mmHg in the left arm and 198/118 mmHg in the right arm, along with absent femoral pulses. Additionally, there was a systolic blood pressure gradient between the upper and lower limbs, measuring 97 mmHg on the left and 88 mmHg on the right, as well as carotid hyperpulsatility and sternal notch findings. Auscultation revealed a diffuse continuous murmur over the chest.

The electrocardiogram revealed sinus arrhythmia and left ventricle hypertrophy (**Figure 1**). Chest X-ray showed a normal heart and notches at the lower edge of some ribs (**Figure 2**).

During transthoracic echocardiography (**Figure 3**), the exploration of the aortic isthmus revealed a reduction in its caliber without acceleration of flow or gradient on Doppler. The left ventricle was neither dilated nor hypertrophied, with a normal ejection fraction. There was no valvulopathy. Additionally, an ostium secundum-like small-sized interatrial communication with a left-to-right shunt was noted.

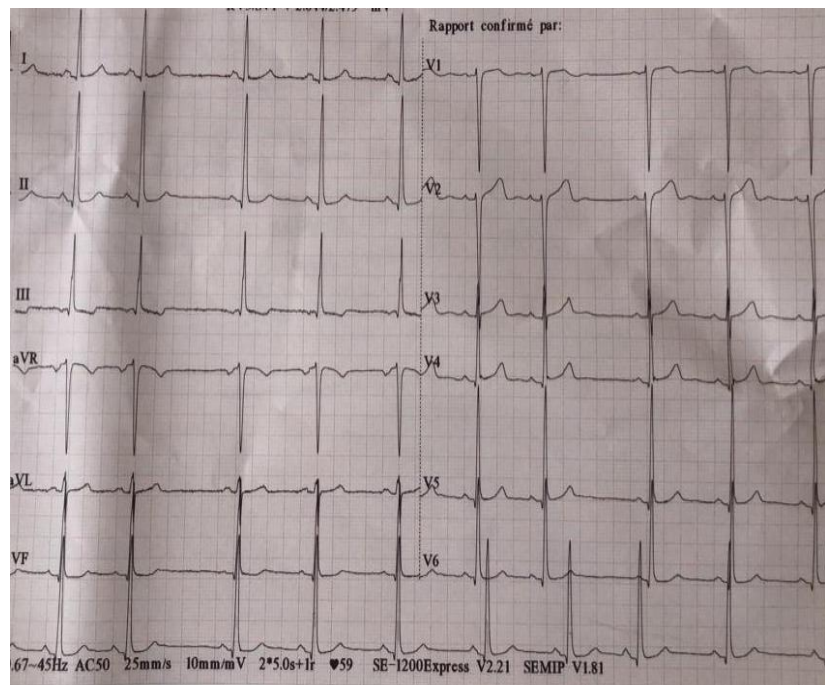


Figure 1. Electrocardiogram showing a sinus arrhythmia and left ventricle hypertrophy.

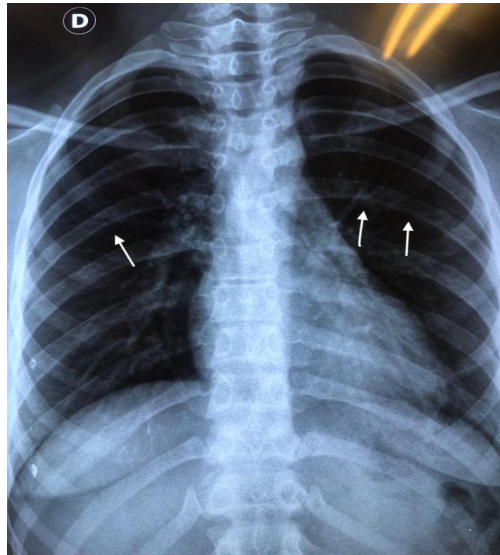


Figure 2. Frontal chest X-Ray showing irregular lower edge of some ribs: rib notching (arrows).

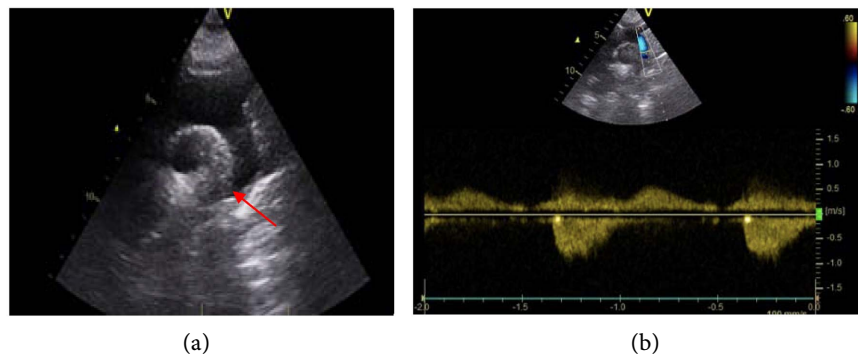


Figure 3. Echocardiography 2D outlining the coarctation of the aorta (a) (red arrow) and on Doppler a systolic flow of 60 cm/s with the consistency of a diastolic pattern (b).

Regarding this suggestive presentation of aortic coarctation, contrasting with the absence of flow acceleration and the non-significant gradient on cardiac Doppler echo, a CT angiography of the aorta was performed. It revealed a complete interruption of aortic continuity between segment 3 and the descending segment, along with a significant collateral network originating from the intercostal and mammary arteries (**Figure 4**). Percutaneous treatment involving the placement of a covered stent is being considered for the correction of this coarctation.

3. Discussion

Coarctation of the aorta is a focal narrowing of the aorta's caliber, with isthmic localization observed in 95% of cases [4]. It constitutes 5% to 8% of congenital heart diseases, with a male predominance [5]. Diagnosis can occur either in the context of neonatal acute heart failure or incidentally in adulthood when severe and treatment-resistant hypertension is present, as seen in our patient.

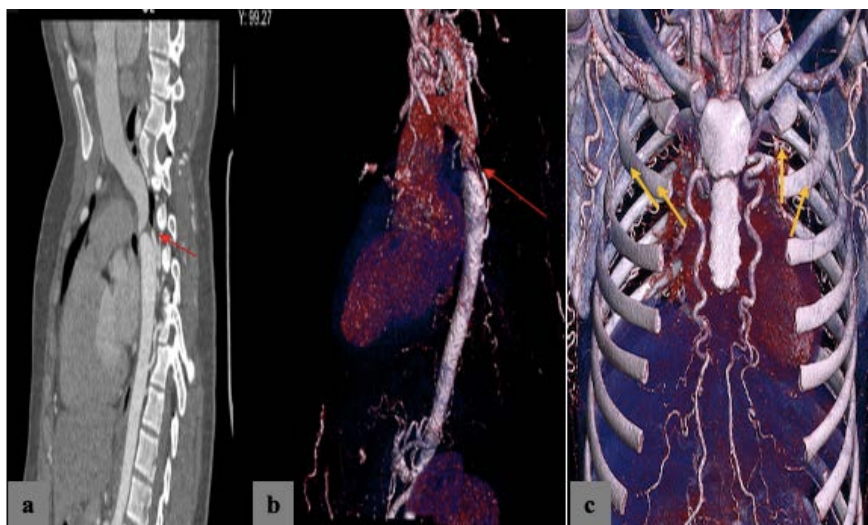


Figure 4. Thoracic CT angiography in sagittal reconstruction (a) and 3D VR ((b), (c)) revealing complete interruption of the continuity of the aorta between segment 3 and descending segment (red arrows) along with a significant collateral network originating from the intercostal and mammary arteries (yellow arrows).

Classically, the stenosis is localized, characterized by a diaphragm that creates an eccentric passage for blood flow. Totally interruption of aortic continuity is rare [6]. Typically, diagnosis occurs during adolescence or young adulthood, likely due to the progressive nature of the obstruction in the coarcted segment [2] [7].

The clinical presentation of the complete form does not significantly differ from that of tight CoA. Indeed, in our patient, high blood pressure in the upper limbs was associated with a significant systolic blood pressure gradient (>20 mmHg) and absent femoral pulses.

The characteristic murmur is a left interscapulovertebral systolic murmur or a continuous murmur from collaterals. Electrocardiogram findings vary based on the impact. The most common abnormality is left ventricular hypertrophy [8]. On chest radiography, a classic “number 3” appearance is observed, formed by the dilation of the left subclavian artery and the aorta near the stenosis, the stenosis itself, and post-stenotic dilation. Additionally, there may be erosion of the lower edge of the ribs (rib-notching) due to collateral development through intercostal and mammary arteries, as seen in our patient [4].

Given such a clinical picture, bidimensional echocardiography is the initial diagnostic test. It allows visualization of the coarctation via the suprasternal approach and assessment of its impact on the left ventricle. Using continuous-wave Doppler through the suprasternal approach, it searches for a negative systolic flow with diastolic extension in cases of severe coarctation. The trans-stenotic pressure gradient can be evaluated based on the maximum velocity peaks using the Bernoulli formula. In color Doppler, the mosaic flow pattern of the coarctation resembles an hourglass, resolving in the descending aorta where it becomes highly turbulent. Trans-thoracic echocardiography also contributes to detecting

other associated anomalies such as bicuspid aortic valve, aortic stenosis, or mitral stenosis [1] [9].

Based on a highly suggestive clinical scenario, bidimensional echocardiography revealing left ventricular hypertrophy, alleviated or non-pulsatile flow in the abdominal aorta associated with diastolic extension, and an unvisualized aortic isthmus should raise suspicion of a complete form of CoA, similarly to our case, or an atypical localization. Consequently, cross-sectional imaging, such as CT angiography or MRI, proves invaluable. Thoracic CT angiography precisely localizes the coarctation and provides a thorough analysis of circulation and associated vascular anomalies (such as aneurysms upstream or downstream from the coarctation). Angio-MRI is the method of choice for exploring aortic coarctation in adults. It yields information similar to that obtained from aortography and angioscanner and serves as the gold standard for post-treatment evaluation of CoAo [1] [4].

In the absence of therapeutic management, the life expectancy of patients with complete Coarctation of the Aorta (CoA) is shortened. Heart failure is the main cause of death, followed by dissection, endocarditis-related complications, and intracranial hemorrhage [3] [10].

Transluminal percutaneous angioplasty with a covered stent is currently preferred over surgery in several centers, particularly in adults. Procedure-related complications are rare and primarily related to femoral artery puncture. However, there is still a risk of dissection during the intervention [2] [3] [11].

CoA remains a permanent condition. Even after repair, data indicate increased cardiovascular morbidity and mortality. The long-term survival rate of adult patients who have undergone surgery is significantly lower than that of the general population. This is associated with earlier coronary artery disease and complications related to persistent hypertension [12].

4. Conclusion

Totally occluded coarctation of the aorta is very uncommon. It should be considered in cases of high blood pressure associated with absent or weak femoral pulses, especially in young individuals. Transluminal angioplasty with the placement of a covered stent is the preferred method for correction.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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