Post - Ductal Coarctation of Aorta : A Late Presentation in Adulthood

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Introduction

Coarctation of the aorta (CoA) is the narrowing of the aorta commonly located at the site of insertion of ductus arteriosus, distal to the origin of the left subclavian artery . It is a congenital defect accounting for 4-6% of all congenital heart diseases [1] .The lesion disrupts the normal outflow of blood from the left ventricle, consequently giving rise to compensatory mechanisms like ventricular myocardial hypertrophy to surmount this obstruction and maintain the normal systolic function and ejection fraction; and collateral blood flow (involving the intercostal, internal mammary, and scapular vessels) [2,3]. Other cardiac defects such as ventricular septal defect, bicuspid aortic valve, and mitral valve abnormalities occasionally occur in conjunction with coarctation of aorta.

The most prominent clinical feature in this condition is systemic hypertension associated with discrepancy in the arterial pulses of the upper and lower limbs. Majority of cases are diagnosed and treated in the neonatal period, whereas some asymptomatic patients may remain undiagnosed until adulthood [4,5]. The classic clinical manifestation of such cases later in life is severe hypertension.

Case Report

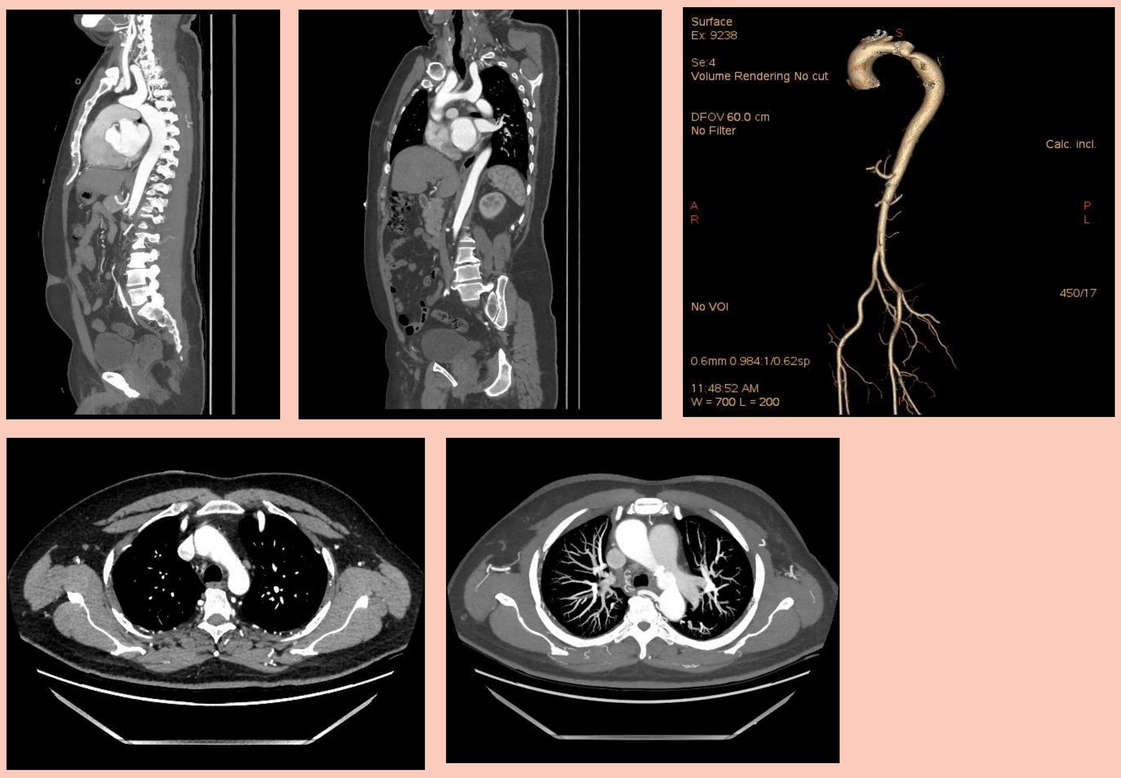
A 60 year old man with a known case of essential hypertension for 20 years on medication (Atenolol 100 mg & Enalapril 5 mg) was admitted as a case of hypertensive emergency with a 1 day history of dizziness and central chest pain which was intermittent and pressure- like in character. It was initially exertional after 15-20 minutes of walking, however it progressed to even occurring at rest, associated with generalized weakness but no focal neurological deficits. He had no other symptoms of leg swelling, orthopnea, PND, or shortness of breath. Upon further enquiry, it was brought to attention that he had a previous admission with similar symptoms 2 years ago which was not followed up.

On examination he was found to have BP of upper limbs >200/70 mmHg and lower limbs 120/70 mmHg (right upper limb 197/69, right lower limb 122/85, left upper limb 220/71, left lower limb 124/78) and ejection systolic murmur on both sides of the precordium, more prominent along the left parasternal border, radiating to the carotids and back. A vascular exam revealed feeble lower limb pulses and a positive finding of a radio-femoral delay.

His lab values were unremarkable. ECG revealed ST- elevation of 5 mm in the anterior pericardial leads with evidence of left ventricular hypertrophy. The relevant imaging was requested with the chest X-ray showing mild cardiomegaly with left ventricular configuration of left heart border and a widened mediastinum. Based on this result a preliminary diagnosis of aortic dissection was made. Furthermore , departmental echo was arranged which revealed an ejection fraction of 60-65% , moderate left ventricular hypertrophy with an intact interatrial septum with no evidence of a shunt on doppler studies and lastly coarctation of aorta measuring the peak gradient at 113 mmHg.

In order to confirm the diagnosis of coarctation of aorta a CT aortogram was performed (in addition to an Invasive Aortography which confirmed diagnosis of tight coarctation of descending thoracic aorta just distal to the insertion site of the left subclavian artery),with the following findings:

- Short segment abrupt significant narrowing of the descending thoracic aorta just distal to the insertion site of the left subclavian artery (post ductal coarctation of the aorta)
- Associated cardiomegaly with left ventricular hypertrophy. Compensatory collateral mechanism in form of prominence of anterior mammary arteries and intercostal arteries collaterals bilaterally.
- Contrast defect in posterior branch of the right pulmonary artery to the upper lobe extending for around 3.5 cm long segment consistent with pulmonary embolism.
- The stenosed segment of the aorta is noted in the descending aorta just distal to the arch for 2 mm long segment and it is 4.8 mm in width. The part of aorta just proximal to the coarctation is up to 23.9 mm in width and distal to it is 23.8 mm in width.



He was admitted as a case of hypertensive emergency and loaded with Aspirin 300 followed by Aspirin75 mg daily and Atorvastatin 80 mg, enoxaparin 40 mg Amlodipine-valsartan-hydrochlorothiazide 1 tablet twice daily.

Patient was planned for surgical correction of coarctation of aorta and follow-up in the OPD. Prior to the scheduled date for surgery, he presented to the ER with chest pain and high BP 175/63 which was attributed to his condition. {Left hand :183/76mmhg Right hand : 199/75 mmhg Left leg : 120/68 mmhg Right leg : 120/75 mmhg}

Correction of the coarctation site was done successfully with the stent, without any residual stenosis. Day 1 post-stenting the patient started feeling dizzy, weak and mildly confused with delayed speech and was found to have an acute left PCA territory infarct with associated cytotoxic oedema at the left occipital region. Patient's anti-coagulation medications consisting of Enoxaparin and Rivaroxaban were withheld with regards to further clearance and was only given Clopidogrel.

As the patient had the risk of predisposition to thromboembolic events and ischemic infarct with severe stenosed coarctation of the aorta, the possibility of acquired von Willebrand disease was considered and hence the decision was made to conduct Factor V leiden and thrombophilia screen once the course of anticoagulation was completed, to assess for long term need for anticoagulation. He was referred to Hematology after blood work up showed Factor V heterozygous.

Antihypertensive medication started to be weaned slowly according to hypertension response

Consultant review was done and the patient was asked to follow up in the Adult Congenital Clinic in 2 months

He is doing well and currently asymptomatic.

Discussion

Coarctation of Aorta (CoA), is a congenital heart disease, rarely diagnosed in adulthood. Its presentation in adults is commonly with hypertension, or with complications such as epistaxis, heart failure, angina, or aortic dissection etc. Physical examination along with a high index of suspicion in patients with hypertension is key in its diagnosis. CoA causes an elevated blood pressure in the upper limbs and a decreased blood pressure in the lower limbs, along with diminished femoral pulses. Additional physical signs include an ejection systolic murmur with radiation to the back, or a continuous murmur suggestive of arterial collaterals in those with long standing coarctation [6]

Many imaging modalities can be used to establish the diagnosis such as transthoracic echocardiography, cardiac magnetic resonance imaging or computed tomographic angiography. The gold standard test, however, is cardiac catheterization and angiography. In our case, the initial diagnosis was established based on TEE and CT angiography [7]

Treatment of CoA in adults can be either surgical or via transcatheter interventions which include balloon angioplasty with or without stenting. Intervention is indicated when a peak-to-peak coarctation gradient is ≥ 20 mg or peak-to-peak coarctation gradient is < 20 mg with imaging evidence of significant coarctation and radiographic evidence of collateral flow [8]. The choice of intervention should be determined on an individual basis and depends on many factors including patient's comorbidities and presence of other congenital anomalies. Nonetheless studies have shown that both balloon angioplasty and surgical correction are equally effective in acutely reducing the pressure gradient [9,10]

In untreated cases, the average life expectancy is till the age of 35 years. Death may occur from left ventricular failure, cerebrovascular events, endocarditis, or aortic dissection. Therefore, timely recognition and treatment is important in improving the overall survival of individuals [6]

Our case highlights a relatively rare cause of hypertension in adults. In an older adult who presents with hypertension such as in this case, it is important to measure blood pressure in all four limbs and consider coarctation as a differential.

Conclusion

All patients with either interventional catheterization or surgical repair of coarctation of the aorta should have close follow-up and aggressive management of blood pressure and other risk factors for cardiovascular disease. This should include at least yearly cardiology evaluations. Consultation with a cardiologist with special expertise in ACHD should be obtained on initial contact to determine risk factors specific for the patient's anatomy and the presence of associated lesions. Evaluation of the repair site by MRI/CT should be repeated at intervals of 5 years or less, depending on the specific anatomic findings before and after repair. Consideration should be given to cumulative lifetime radiation exposure with multiple CT examinations.

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