Case Report

Coarctation of aorta presenting as hypertensive encephalopathy in a young female

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Abstract

Coarctation of Aorta (CoA) is a congenital cardiac defect which can present in adults. The usual presentation is hypertension. Though it has subtle clinical signs but they can go undetected if not sought for. One of the most catastrophic presentations can be severe hypertension with its complications. We report a case of a 40 year old female who presented to us with hypertensive encephalopathy and finally diagnosed as CoA

Key words: Coarctation of Aorta, CoA, Hypertension, Hypertensive encephalopathy

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Introduction

CoA is a congenital cardiac malformation, where there is a constriction in the aorta distal to the left subclavian artery. Sometimes the constriction occurs proximal to the subclavian artery when it is called preductal CoA. The incidence of CoA is 4 in 10,000 live births. Five to eight percent of children with congenital heart disease have CoA [1].

The defect manifests in early life and is usually treated appropriately but sometimes when CoA presents in an adult, it is mainly due to re-coarctation following catheter or surgical therapy and rarely can be a case of native coarctation presenting first time in adult life. If not treated, the mean life expectancy is 35 years. Ninety percent of patients die before the age of 50 years. It can complicate as systemic hypertension, coronary heart disease, congestive cardiac failure, aortic dissection and stroke [2].

The different methods of treatment are medical therapy and surgical therapy including percutaneous angioplasty with or without stent placement.

Case Report

A 40 year old female presented to us with 2 days history of severe headache, 2 episodes of vomiting and drowsiness since 8 hours. There was history of seizure 2 hours before admission. On examination, the patient was drowsy and a bit irritable (GCS: 9), pulse- 118/min regular. Dorsalis pedis, posterior tibial and popletial pulses were not palpable. Femoral pulses were weak, no carotid bruit or thrill was detected. Radio femoral delay was present. Blood pressure was 210/ 120 mmHg in right arm, 206/112 mmHg in left arm and 170/98 mmHg in left lower limb. There was no cyanosis, jugular venous pressure was normal and pedal edema was absent. Cardiovascular examination revealed normal S1 and S2. Fourth heart sound and ejection systolic murmur in the lower left sternal border were heard. Respiratory and abdominal examinations were normal. CNS- higher function: drowsy, no cranial nerve deficit, moving all four limbs, planter's bilateral extensor and no terminal neck stiffness.

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Fundus examination revealed Grade III hypertensive retinopathy. In view of the above clinical examination, possibility of hypertensive urgency due to CoA was kept. For evaluation of seizure and headache, urgent neuroimaging was done and subarachnoid hemorrhage and stroke were ruled out.

The patient was treated with intravenous nitroglycerine (20mcg/min) and injection hydralazine (20 mg intramuscular stat) and blood pressure was controlled to 160/90 mmHg in the next 7 hours. The patient was put on tablet metoprolol 50 mg twice a day once the injectable anti-hypertensive drugs were tapered off.

Her complete blood count, kidney function test, liver function test and blood sugar levels were normal. X-ray chest showed cardiomegaly and visible biarcute appearance of aortic arch ("3" sign). ECG showed left ventricular hypertrophy with strain pattern. 2D-Echocardiography showed a peak systolic gradient of 72 mmHg and low antigrade diastolic flow in the descending thoracic aorta in a classical saw tooth pattern which was highly specific for aortic obstruction.

Peripheral angiography was done by placing a sheath in the femoral artery and dye was injected which revealed coarctation distal to left subclavian artery with post stenotic dilatation of descending aorta. (Fig 1)

Considering the presence of high gradient coarctation with severe hypertension, the patient underwent balloon angioplasty with stent placement.

Immediately after the stent placement, the lower limb pulses were easily palpable. The blood pressure came back to normal decreasing the dose of antihypertensive within 6 days. The patient was discharged after 7 days and is awaiting follow up after 3 months. She was given tablet clopidogrel 75 mg and tablet Aspirin 150 mg for 3 months at the time of discharge.

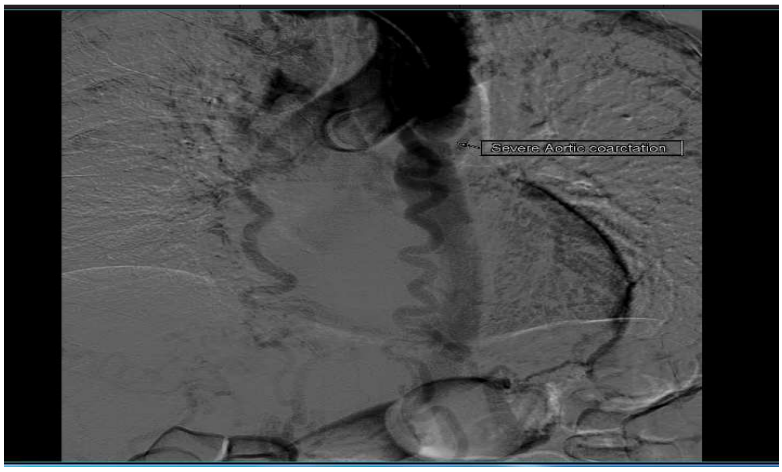

Fig 1: CoA distal to left subclavian artery (small black arrow) with post stenotic dilatation of descending aorta

Discussion

CoA is characterized by discreet narrowing of thoracic aorta distal to left subclavian artery. The defect imposes significant afterload on the Left Ventricle resulting in wall stress, compensatory left ventricular hypertrophy, left ventricular dysfunction and collateral formation.

CoA in adults usually presents as systemic hypertension and discrepancy between upper limb and lower limb blood pressure as in our case. The peculiarity of our case is hypertensive encephalopathy which is a medical catastrophe.

The 2008 American Heart Association/American College of Cardiology guidelines for intervention in CoA includes peak to peak coarctation gradient >= 20 mmHg or peak to peak coarctation gradient < 20 mmHg in presence of anatomic imaging revealing significant coarctation. The European society of cardiology recommends early treatment in all patients with a non-invasive pressure difference of more than 20 mmHg in upper and lower limbs regardless of symptoms but with upper limb hypertension of > 140/90 mmHg and significant left ventricular hypertrophy [3].

Surgical repair of coarctation is achieved by resection with end to end anastomosis [4,5]. Aneurysm formation is a common complication of surgical therapy. Sometimes aortic dissection may occur late after surgical repair. Morbidity includes paradoxical hypertension, hoarseness of voice, diaphragmatic palsy and subclavian steel syndrome.

Balloon angioplasty was introduced in 1982 and is currently done with or without stent deployment. It is the preferred treatment modality in native coarctation in adults or re-coarctation after surgery [6]. There is an increase incidence of aneurysm and restenosis after balloon angioplasty.

Stenting of CoA was introduced in the 1990 using bare metal stents. Acute mortality with this procedure is 0-3%[7]. Acute aortic dissection may be seen in 13% of cases [8]. Biodegradable stents are an area of research [9,10].

The best method of repair of CoA is based on several factors. Though stent implantation carries the lowest morbidity, repeated interventions may be required as compared to surgery. Endovascular therapy is currently the treatment of choice, when there is ventricular dysfunction and other comorbidities like diabetes and ischemic heart disease are present [7].

Drugs that are used to control the hypertension are beta blockers, angiotensin converting enzyme inhibitors and angiotensin receptor blockers [3]. Survival of the patients with CoA has dramatically improved after surgical repair became available.

Conclusion

CoA is a congenital cardiac malformation that can go undiagnosed. Hypertension in a young individual should raise a suspicion of CoA and it should be sought for. Nowadays different surgical and interventional treatment are available but these modalities should be individualized for each patient.

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