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AN UNSUAL CASE OF SECONDARY HYPERTENSION IN AN ADOLESCENT FEMALE.

General Medicine

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ABSTRACT

Coarctation of the aorta (CoA) is a congenital narrowing of the aorta that often goes undiagnosed until adolescence or adulthood, leading to secondary hypertension. We report a case of seventeen year old female with CoA, presented with complaints of claudication in lower limbs and persistent headache. Coarctation was noted distal to the left SCA on 3D CT angiogram. Resection of the coarcted segment and end to end anastomosis with Dacron graft led to normalisation of blood pressure and resolution of symptoms on follow up. This case emphasizes the importance of considering CoA in the differential diagnosis of adolescents with secondary hypertension, it's early diagnosis and repair.

KEYWORDS

Hypertension, Headache, Claudication, radio-femoral delay, end-to-end anastomosis.

INTRODUCTION:

Coarctation of aorta is a congenital cardiovascular anomaly characterised by narrowing of the aortic lumen resulting in hypertension and left ventricular hypertrophy[1,2]. It accounts for 5 to 8% of all congenital heart defects and is more common in males[3,4,5].It may be isolated or associated with bicuspid aortic valve, mitral valve prolapse and ventricular septal defects[6]. The classic COA is located in thoracic aorta distal to origin of left Subclavian artery at about the level of ductal structures. In an adult COA may be long or fusiform, with irregular lumen which may be inflammatory or autoimmune in origin and may also be a variant of Takayasu arteritis.

Though it is often diagnosed in early childhood, a significant number of cases remain undiagnosed until adolescence or adulthood. In rare instances, CoA is diagnosed in older adolescents where clinical signs may be more subtle and overlooked for years. This delayed presentation often results in complications such as hypertension, left ventricular hypertrophy. In severe cases it might lead to heart failure or cerebrovascular accidents[7]. Since the introduction of surgical correction in the early 1940s, surgical repair remains the treatment of choice in symptomatic individuals and has been proven to significantly reduce arterial hypertension on follow up[8,9]. Surgical options include resection with end to end anastomosis, subclavian flap aortoplasty, by pass graft repair and patch aortoplasty with the techniques being occasionally combined or modified to fit the patient's individual anatomy[10]. Balloon angioplasty and stent implantation are viable options but are usually employed in adults or those with re-coarctation.

CASE REPORT:

A 17 year old female, recently diagnosed hypertensive on dual antihypertensive therapy, presented to Cardiology outpatient department with progressive holocranial headaches and claudication in lower limbs for several months .There is no family history of hypertension. Physical examination revealed visible suprasternal pulsations with radio-femoral delay, feeble femoral and popliteal pulsations and absent dorsalis pedis pulse in bilateral lower limbs. There was no history of pedal edema, discoloration of skin or nail changes. The BP in right and left upper limbs was 170/100 each and in both lower limbs 100mmHg SBP.On auscultation, ejection systolic murmur was audible in left interscapular area. Fundus examination revealed increased arteriolar thickening with tortuosity and anterior venous nipping pertaining to Grade 2 Hypertensive retinopathy changes and no flame shaped hemorrhages or cotton wool spots. Rest of the systemic examination was unremarkable.

Left ventricular hypertrophy was noted on 12 lead Echocardiogram. Chest radiograph revealed high positions aortic arch with a discreet '٣' sign, inferior rib notching and cardiomegaly. Echocardiography showed signs of coarctation with peak systolic gradient of 80 mm hg and left ventricular hypertrophy .CT angiogram of aorta and renal vessels showed *coarctation of aorta distal to origin of left subclavian artery and* normal opacification with contrast and no signs of stenosis within the renal vessels respectively. There was no evidence of aortic or branch (subclavian, carotid, renal) vessel wall thickening, stenosis, occlusion or aneurysmal dilatation ,effectively ruling out Takayasu arteritis. Her hematological and biochemical values were within their normal ranges (Sr creatinine 1.1).

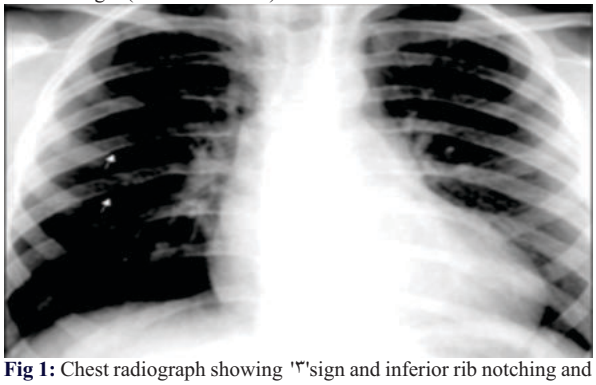

Fig 1: Chest radiograph showing '٣'sign and inferior rib notching and cardiomegaly

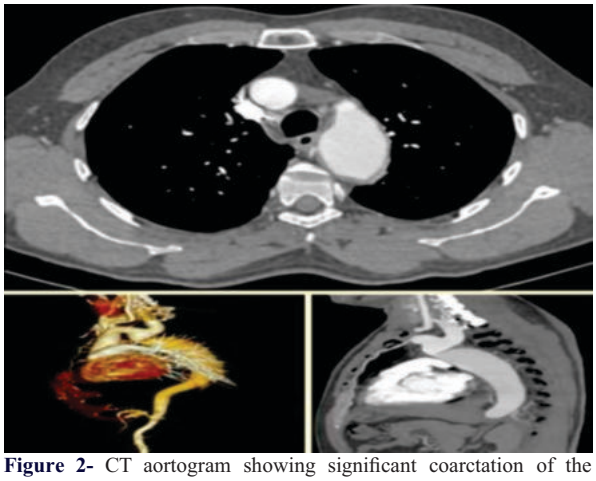

Figure 2- CT aortogram showing significant coarctation of the thoracic aorta beyond the origin of the left subclavian artery (arrow) with post stenotic dilatation.

She was started on dual antihypertensive therapy . She underwent resection of coarcted segment with end to end anastomosis and aortoplasty with Dacron patch. Surgery was uneventful and was discharged on a beta blocker. At 6 month follow up visit the clinical examination showed preserved lower limb pulses and patient needed lower dose of antihypertensive therapy.

In subsequent follow ups her blood pressure came down to normal and she was weaned off of the antihypertensives. She got married 4 years later and went on to conceive again. The pregnancy was uneventful and there was no evidence of high blood pressures or Preeclampsia and at term she delivered a healthy baby by Caesarean section.

DISCUSSION:

Coarctation of the aorta in adults accounts for 0.2% of all hypertension cases in adults[11].It is often diagnosed in infancy or early childhood, but in some cases, it may remain undetected until adolescence or adulthood. According to the *Practice Guideline for Screening and Management of High Blood Pressure in Children and Adolescents*[12], secondary hypertension in adolescent population may often be attributed to renal diseases, followed by cardiac diseases including CoA and rarely endocrine diseases such as pheochromocytoma, primary aldosteronism and Cushing's. As of now, there are still many CoA patients who are diagnosed in adolescence and adulthood and may require surgical treatment[13,14]. Therefore, it is reasonable to suspect that CoA is probably a major cause of hypertension in adolescence, because of the current scenario of low diagnostic accuracy.

In this case, the patient's symptoms of headache and claudication were initially under evaluated. However, the discrepancy in blood pressure between the upper and lower limbs and the presence of weak lower limb pulses raised suspicion for an underlying vascular abnormality, prompting further investigation. The most common presentation of CoA in adolescents is secondary hypertension, which results from the increased afterload due to the narrowed aorta. Headache and claudication pain can occur as a result of reduced perfusion to the lower extremities and the brain. This case highlights the importance of considering CoA in the differential diagnosis of adolescents with unexplained hypertension and peripheral vascular symptoms.

Studies suggest that CoA can be safely repaired until early adolescence, preferably before 5 years of age[15,16]. As the disease progresses, complications such as heart failure, dissection of aorta or rupture , coronary disease, hypertensive crisis, hemorrhagic cerebrovascular accident can occur[7]. Without correction, the mean life expectancy of patients with CoA is around 34 years, and 90% die of vascular complications before the age of 50 [17],emphasizing the need for early diagnosis and treatment of this condition. Management of previously undiagnosed COA adults varies based primarily on the stability of the patient and severity of the lesion. *The American College of Cardiology and the American Heart Association* recommended intervention in adults with COA in the following settings: when a peak-to-peak coarctation gradient ٢٠ ≤ mm Hg or peak-to-peak coarctation gradient < 20 mm Hg with imaging evidence of significant coarctation and evidence of collateral flow Surgical intervention remains the gold standard treatment for CoA.

Currently, different surgical methods and interventions are available but this should be individualised for each patient and for each type of coarctation (native coarctation or recoarctation after surgical or interventional therapy). The end-to-end anastomosis procedure, which involves removing the narrowed section of the aorta and rejoining the healthy ends, has been shown to yield excellent results in terms of blood pressure normalization and symptom resolution. Studies suggest that normalization of blood pressure was achieved in majority of the patients 5-10 years. Upto three quarters of all patients with repaired CoA may develop hypertension by 20-30 years after surgery with the risk being relatively high when operated at a later age (10-20 years age)[18]. In this case, the patient experienced significant improvement in both her blood pressure and quality of life following surgery and was advised regular follow ups.

CONCLUSION:

This case illustrates the importance of considering coarctation of the aorta in the differential diagnosis of adolescents presenting with secondary hypertension. Early diagnosis and prompt surgical intervention can lead to normalization of blood pressure and resolution of symptoms, preventing long-term cardiovascular complications. Given the current situation of hypertension in adolescents being highly underdiagnosed, clinicians should maintain a high level of suspicion for CoA in patients with unexplained hypertension and look for discrepancies in blood pressure measurements in primary hospital. Long term follow up is necessary In view of late hypertension.

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