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Severe Coarctation of Aorta in a 24-year Old Male: A Case Report

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ABSTRACT

Coarctation of aorta (CoA) is a congenital vascular malformation usually diagnosed and corrected early in life but the diagnosis is missed or delayed unless there is a high index of suspicion. Long-term survival is exceptional in patients with untreated aortic coarctation. In our case report, we present a late diagnosis of aortic coarctation in a 24-year-old male who was relatively asymptomatic until he presented with progressive exertional dyspnoea, fatigue, headache and poorly controlled hypertension despite getting a combination of antihypertensive agents in his 2nd decade of life. The patient was managed by surgery of aorta. The coarcted segment was mobilized, resected totally followed by insertion of interposition tube graft of prosthetic material (PTFE) 20 mm in diameter with ligation and transfixation of patent ductus arteriosus (PDA). After 4-year follow-up visit, the patient is in good clinical condition.

Key Words: Coarctation of aorta, Congenital malformation, PTFE.

INTRODUCTION

Coarctation of aorta (CoA) may be defined as a congenital narrowing of the upper descending thoracic aorta that comprises localized medial thickening with a ridge of intimal hyperplasia adjacent to the site of attachment of the ductus arteriosus with tissue extension from it and sufficiently severe that there is a pressure gradient across the area.[1] The aortic lumen may be atretic, but in coarctation the aortic walls above and below are in continuity. Uncommonly, coarctation occurs more proximally between the left common carotid and subclavian arteries. Occasionally, coarctation of aorta may occur in the lower thoracic and abdominal aorta. When associated with or without patent ductus arteriosus (PDA) is called primary coarctation. Other commonly associated cardiac anomalies are ventricular septal defect (VSD), bicuspid aortic valve, aortic stenosis and various mitral valve disorders but may be in association with transposition of great arteries (TGA), double outlet right ventricle (DORV) and Turners syndrome. Some patients with coarctation of aorta may have cerebral aneurysm (Berry aneurysm). Externally the aorta appears to be sharply indented or constricted and internally the obstructing diaphragm or shelf consisting of an infolding of the aortic media on the posterior wall, located preductally, postductally but is usually juxtaductally.[2] The ductal tissue forms a sling that completely surrounds the juxtaductal aorta, which may progressively proliferate after birth and cause restenosis after the repair of coarctation in neonates and young infants.[3]

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Coarctation, as well as isthmic hypoplasia, is more common than usual when ascending aortic flow is diminished during fetal life by lesions such as aortic stenosis or atresia and mitral stenosis or incompetence.[2] Conversely, the prevalence of coarctation is severely reduced and the size of isthmus increased, when pulmonary flow and thus right to left ductal flow is decreased by lesions such as pulmonary stenosis or atresia, tetralogy of Fallot and tricuspid atresia.[4] Coarctation is very uncommon when the aortic arch is right sided, presumably because of alteration of ductal and isthmal flow patterns in this situation.[2] The collateral circulation between aorta proximal to the coarctation and that distal to it is one of the striking feature of coarctation. When fully developed, parascapular pulsation and rib notching-the classical signs of malformation is observed as the patients age advances. Absence of rib notching in right chest suggests an anomalous origin of the right subclavian artery and in the left chest a stenosis of the left subclavian artery origin.[1]

When the ductus arteriosus remains widely patent and a severe coarctation proximal to it (preductal coarctation), there may be a right to left shunt into the descending aorta and classically, cyanosis of the toes and sometimes the left hand while the right hand and lips remain pink (differential cyanosis). The femoral pulse under these circumstances is normal and there is no ductus murmur.[1]

Case Report:

A 24-year-old man was referred to our hospital because of increasing fatigue and exertional dyspnoea, headache and hypertension. He had been well until 11 months previously. The patient had a medical history of hypertension. His hypertension was poorly controlled despite a combination of antihypertensive agents (beta-blocker, calcium channel blocker and angiotensin receptor blocker). Physical examination showed blood pressure 145/85 mmHg in both arms, a heart rate of 92 beats/minute. Femoral pulses were palpable bilaterally but weak and delayed compared to the radial pulses. A systolic murmur was heard on the left precordium in infraclavicular region in front and interscapular area on the back. A cardiac silhouette at the upper limits of normal and notching of the ribs were observed on the chest radiography. His echocardiogram showed coarctation of aorta with a patent ductus arteriosus, mild aortic regurgitation, trivial mitral insufficiency, concentric left ventricular (LV) hypertrophy with good LV systolic function (62%). CT aortography showed a mildly dilated aortic root, and juxtaductal coarctation of aorta below the subclavian artery with post stenotic dilatation of descending aorta with numerous grossly dilated collaterals (Fig. 1 and 2). The operation was done via left posterolateral thoracotomy in 4th intercostal space. Since, the collaterals were well recognized before surgery, the procedure was done carefully without major bleeding and any adverse event. Ligation with transfixation of PDA

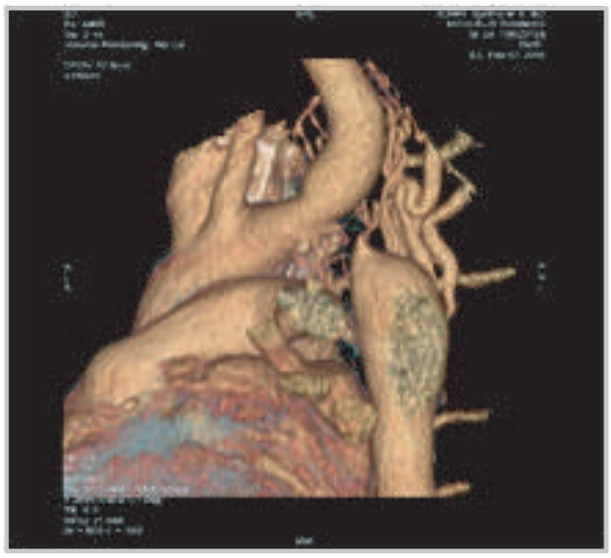

FIGURE 1: *CT aortography showing coarctation of aorta, post stenotic aortic dilatation, multiple dilated collaterals, and dilated subclavian artery and PDA.*

was done. The coarctated segment was mobilized, resected totally followed by insertion of interposition tube graft of prosthetic material (PTFE) 20 mm in diameter. The cross clamp time was 29 minutes and because the collaterals were left intact, any malperfusion syndrome has not occurred. Total hospital stay after procedure was only seven days. After the last 4-year follow-up visit, the patient is in good clinical condition.

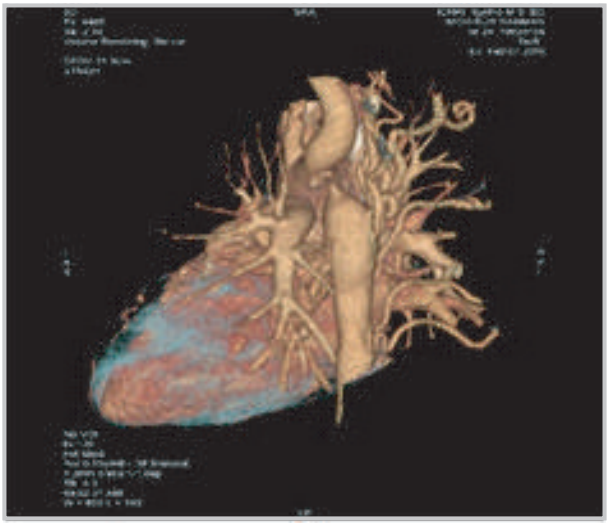

FIGURE 2: *CT aortography showing Co arctation of aorta, post stenotic aortic dilatation, multiple dilated collaterals, dilated subclavian artery and PDA.*

DISCUSSION

Coarctation of aorta is a vascular lesion accounting for 5 to 10% of all congenital cardiovascular defects,[5] more in males than females (2:1). The patient may be asymptomatic and remains undetected well until adulthood[6] or the diagnosis may be missed or delayed unless there is a high index of suspicion. It manifests as childhood hypertension, headache, congestive heart failure, and sometimes chest pain or lower extremity fatigue, weakness with or without pain and life threatening intracranial haemorrhage. Often diagnosis is made after hypertension is noted as an incidental finding during evaluation for other problems. Diagnosis is usually based on clinical suspicion and physical findings.[7] The latter include blood pressure difference between the upper and lower extremities, radio-femoral pulse delay, systolic murmur over the left precordium. Generally, patients with coarctation of aorta present with congestive heart failure in early life or with hypertension in older children and adults. The timing of clinical presentation and the severity of symptoms depend on the extent of patency of ductus arteriosus, the rapidity of closure of the ductus arteriosus, the level of pulmonary vascular resistance, presence of associated defects (e.g., VSD) and aortic arch anomalies. Presentation may be abrupt and acute with ductal closure. Patients may present in the first few weeks of life with poor feeding, tachypnea, lethargy and progress to overt CHF and shock.[8] After the neonatal period, they often do not develop overt CHF because of the presence of arterial collateral vessels. Prophylaxis treatment consists of aggressive hypertension therapy, endocarditis prophylaxis and corrective treatment either endovascularly (e.g., Stenting) or surgically including subclavian flap aortoplasty, PTFE patch aortoplasty, excision of coarcted segment and end to end anastomosis or with interposition graft. In our case, the patient was treated by PTFE interposition tube graft which is appropriate for an adult patient with ligation and transfixation of PDA. Prognosis and survival depends on the disease severity and patient's age at the time of correction. Most untreated patients with coarctation of the aorta will die before 50 years of age.[9] Death in untreated patients are usually due to heart failure, coronary artery disease, aortic rupture/dissection, concomitant aortic valve disease, infective endarteritis /endocarditis, or cerebral hemorrhage.[9,10]

CONCLUSION

Palpation of radio-femoral pulses in a hypertensive patient during routine examination is necessary to avoid a delay in the diagnosis of coarctation of aorta. Failure to diagnose the disease early in life may cause life-threatening cardiovascular problems at any time.

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