Clinical Image

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Abscess of the axilla in hypertensive adult female

Clinical Presentation

A 26-year-old female was referred from the primary care center to our medical clinic for management of hypertension. She presented to the primary care center with an abscess of the axilla and was found to be hypertensive. She did not have any other symptoms. Clinical examination showed a moderately built young lady. Blood Pressure was 166/93 in both upper limbs. Upper limbs pulses were felt normally. However, lower limb pulses including femorals were not felt. There was no femoral or renal bruit. There was 2/6 systolic murmur heard below the left clavicle, which radiated to the left infrascapular region. We present this case to highlight the importance of systematic clinical examination while evaluating patients with hypertension.

Figure 1 - Computerized tomography scan with contrast.
Clinical Image

Questions

1. What abnormality is seen?
2. Which segment of aorta is affected?
3. What are the associated cardiovascular abnormalities associated with this condition?
4. What is the most common clinical presentation?

Answers

1. Coarctation of the aorta, adult type.
2. Post-ductal or middle aortic form. It is due to thickening of the aortic media and typically occurs just distal to the ligament arrirosium (remnant of the ductus arteriosus).
3. Coarctation of aorta is usually accompanied by another cardiac lesion. Approximately one-third of patients have other complex cardiac lesions including single ventricle variants, atroventricular canal defect or transposition of great vessels. Eighteen percent had ventricular septal defect. Other associated lesions include bicuspid aortic valve (50%), atrial septal defect or patent ductus arteriosus, mitral regurgitation, aortic stenosis, aortic regurgitation, and mitral stenosis.
4. Clinical manifestations vary in different age groups. New born may remain asymptomatic if there is persistent patent ductus arteriosus or if the coarctation is not severe. Neonate with severe coarctation may present with heart failure and/or shock when patent ductus arteriosus (PDA) closes. In previously undiagnosed adults, the classic presentation is hypertension. Most patients are asymptomatic unless severe hypertension is present.

Discussion

Coarctation of aorta was first described by Morgagni in 1760. Coarctation of the aorta is narrowing of the descending aorta typically located at the insertion of the ductus just distal to the left subclavian artery. Coarctation of aorta accounts for 4-6% of all congenital heart diseases and has a prevalence of approximately 4 per 10,000 live births. Most patients have discrete narrowing of the descending aorta at the insertion of ductus arteriosus. Long segmental defects, tubular hypoplasia, and rarely coarctation located in the abdominal aorta can also occur. There is slightly higher incidence in males than in females (51 versus 49%). Most cases are sporadic.

Two main theories for the development of congenital coarctation of the aorta are:
1. Reduced ante grade intrauterine blood flow causing underdevelopment of fetal aortic arch.
2. Migration or extension of ductal tissue into the wall of fetal thoracic aorta.

A genetic predisposition is suggested by reports of coarctation occurring in family members and by its association with Turner syndrome.

Acquired cases can occur due to inflammatory diseases of aorta such as Takayasu arteritis or rarely severe atherosclerosis. In the case of Takayasu arteritis, the mid thoracic or abdominal aorta is often the site of involvement. Coarctation of aorta is usually accompanied by another cardiac lesion. In one large series approximately one-third of patients had other complex cardiac lesions including single ventricle variants, atroventricular canal defect, or transposition of great vessels. Eighteen percent had ventricular septal defect. Other associated lesions include bicuspid aortic valve (50%), atrial septal defect, or patent ductus arteriosus, mitral regurgitation, aortic stenosis, aortic regurgitation, and mitral stenosis.
Clinical manifestations vary in different age groups. Newborns may remain asymptomatic if there is persistent patent ductus arteriosus or if the coarctation is not severe. A clinical diagnosis is suspected if there is absent or delayed femoral pulse when compared with brachial pulse. A neonate with severe coarctation may present with heart failure and/or shock when the PDA closes.

Diagnosis is often delayed in older infants and children because physical findings are subtle and most patients are asymptomatic. In young children, coarctation of the aorta may present with hypertension and/or murmurs resulting from collaterals or associated heart defects. Heart failure rarely occurs beyond the neonatal period.

In previously undiagnosed adults, the classic presentation is hypertension. Most patients are asymptomatic unless severe hypertension is present. Therefore, we stress the importance of thorough clinical examination in evaluating patients with hypertension.

The average survival age of individuals with unoperated coarctation is approximately 35 years of age, with 75% mortality by 46 years of age. Common complications in unoperated patients or in those operated on late childhood or adulthood is systemic hypertension, accelerated coronary heart disease, stroke, hypertension, and heart failure. The classical findings of coarctation of the aorta are systolic hypertension in the upper extremities, radio-femoral delay, and low or unobtainable blood pressure in the lower extremities. Non-cardiac complications of coarctation include intracranial hemorrhage and subarachnoid hemorrhage. In addition dilated collaterals in the spinal canal can compress the spinal cord or can rupture.

Prenatal diagnosis is often difficult as only 10% of cardiac output flows through the defect. Post natal diagnosis is suspected by systolic hypertension in upper extremities, delayed femoral pulse and low or absent blood pressure in the lower extremities. Echocardiography can establish the diagnosis and severity of coarctation of the aorta. Magnetic resonance and CT angiography clearly demonstrate the location and severity of coarctation of the aorta.

Management of patients with coarctation depends on age, presentation, and severity of lesion. In neonates with severe coarctation, treatment consists of medical therapy with a continuous infusion of prostaglandin E1 to keep the ductus arteriosus open. Dopamine and/or dobutamine is administered in those with heart failure. Once the patient is stabilized, surgical repair can be performed. Palliative balloon angioplasty may be considered to stabilize the critically patient. In older infants and young children, surgical correction has been the primary treatment. There has been increased use of balloon angioplasty if the lesion is amenable. In case of older children and adults, transcatheter intervention with stenting has become the standard treatment. Surgical treatment may be necessary in patients with suboptimal anatomy with vessel tortuosity and transverse arch hypoplasia.

Complications following either surgery or percutaneous intervention include recoarctation, aortic aneurysm, aortic dissection and systemic hypertension.

References