CASE REPORT

COARCTATION OF AORTA

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Four cases of coarctation of aorta are reported. Presentation, site, severity of coarctation and associated lesions were different in each case.

INTRODUCTION

Coarctation of aorta is a narrowing of some part of the aorta. Most commonly this occurs in the region of the ductus arteriosus but occasionally it may involve the lower thoracic or abdominal aorta.

Coarctation of aorta is an uncommon condition and it has been estimated that less than 0.5 % of hypertensive patients have coarctation of aorta as the etiology.

We saw four cases of coarctation of aorta in Medical-B Ward within a short period of three months. This illustrates the importance of comprehensive workup of every hypertensive patient, specially presenting in young age group.

Our cases have demonstrated the wide spectrum of presentation of this condition quite interestingly.

CASE NO. 1

Mohammad Farid, a 16-year-old boy was admitted in the ward for investigations. He was not well for about one year prior to this presentation.

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Corresponding Author: Col(R) Dr SHARIF-UZ- ZAMAN. He complained of paroxysmal palpitations which exaggerated during exercise and were associated with chest pain and breathlessness.

One month prior to admission the patient developed migratory polyarthralgia and fever, which was associated with rigors and sweating and was intermittent.

His one brother and sister had a history of rheumatic fever but not involving the heart.

He was ill and toxic. He had sinus tachycardia of 110 /min. His pulses in upper limbs were collapsing in character and had radio-femoral delay. His BP was 140/10Qmm of Hg in upper limbs and 100/60mm of Hg in lower limbs. He was running high grade fever with no cyanosis or clubbing. Apex beat was visible in the left 5th intercostal space in anterior axillary line. Posteriorly pulsating vessels were palpable over 4th, 5th and 7th intercostal spaces. A systolic thrill was palpable over precordium. A systolic nurmur was heard over the apex, right border of sternum in 3rd and 4th intercostal spaces, at the back and also at aortic areas radiating to the neck. A bruit was heard over the dilated intercostal vessels as well. Spleen was palpable below the left subcostal margin about 3cm and was soft.

His Hb was 9Gm%. TLC was 7000/cram with normal DLC. ESR was 70mm in 1st hour and ASO titre was raised. Urine microscopy showed a few RBCs. Three blood samples were sent for C/S prior to starting him on treatment, which were later reported sterile. On X-Ray chest, cardiac shadow was enlarged. Ribs were notched on inferior margins. ECG showed left ventricular hypertrophy and strain. On echocardiography no vegetations were seen. There was stenotic bicuspid aortic valve and left ventricle was hypertrophied. He

responded very well to antihypertensive and antibiotics. His BP was controlled, fever subsided and subsequently surgery was advised.

CASE NO. 2

Mr. Naveed, a 19-year-old student of Commerce College, Abbottabad was admitted in Medical-B Ward on 30th September, 1992 with headache, dizziness, occasional palpitations and cramps in both legs. He had these symptoms for the last one month. Both his parents were known hypertensive and were on treatment.

On physical examination, he was a healthy looking young boy without any obvious distress. His pulse was 96/min. BP 240/130mm of Hg in upper limbs and unrecordable in lower limbs. Both femoral pulses were weak and popliteal, posterior tibials and dorsalis pedis arteries were not palpable on both sides. A pansystolic murmur was beard over the apex radiating to axilla. Another systolic bruit could be heard at back and infrascapular area. Arterial pulsations were palpable at the back. His fundi showed Grade-II hypertensive changes.

ECG showed left ventricular hypertrophy. Biochemical parameters were normal. X-Ray chest showed notching of the left 4th, 5th, 6th ribs at the inferior margins. There was no notching of the ribs on right side. Echocardiography showed LVH.

His blood pressure was controlled with calcium antagonists and he was advised aortogram and surgery.

CASE NO. 3

FNA, a 15-year-old school girl was admitted in surgical ward for recurrent abdominal pain and backache. On review of history it was found that she was not well for the last three months. She used to get severe abdominal pain after meals, backache and cramps in both legs. She also had headache, giddiness and nausea.

On physical examination she was pale and ill looking. Her BP was 240/160mm of Hg in upper limbs. There was a systolic murmur at left sternal edge. Another bruit was heard in epigastrium, over umbilical area down to right iliac fossa and femoral arteries. A systolic bruit was also audible at interscapular area. BP in lower limbs was 180/120mm of Hg. Her fundal examination showed AV nipping on both sides and hemorrhages in right eye.

Her urea, electrolytes, urine R/M and X-Ray chest were normal. ECG showed left ventricular hypertrophy.

She was sent for Doppler studies which revealed coarctation of aorta at diaphragmatic level. Additionally, there was stenosis of superior mesenteric artery, which explains the abdominal angina.

On aortogram she laid 4cm coarctation of aorta at T12-L1 level with 40% occlusion. Right renal artery was not outlined.

Her BP was controlled with beta blockers and calcium antagonists and she improved markedly. She was advised surgery.

CASE NO. 4

Mr. Aslam, 40 years old farmer, father of seven children from Mansehra was brought to Casualty Department of DHQ Teaching Hospital, Abbottabad on 15th March, 1993. He had accelerated hypertension and biventricular cardiac decompensation. His BP was 200/140mm of Hg in upper limbs and 1 10/80mm of Hg in lower limbs. He had a systolic murmur radiating to the neck. He had a marked radiofemoral delay. Prior to this admission he used to get attacks of breathlessness which got relieved without any medication.

ECG showed left ventricular hypertrophy and X-Ray chest showed rib notching. Echocardiography showed confined left ventricular hypertrophy and aortic valve stenosis.

His renal functions were normal and his BP was controlled with calcium antagonists and diuretics. He was advised to go for aortogram and surgery.

COMMENTS

The mode of presentation in these four cases was different.

Case one presented with SBE which is a recognised complication of coarctation of aorta.

In case two the coarctation was so severe that blood pressure was not recordable in lower limbs.

Case three presented with multiple stenotic lesions of tile arterial system involving the superior mesenteric and right renal artery in addition to coarctation of aorta. Rib notching was also absent.

Case four presented with accelerated hypertension.

DISCUSSION

Coarctation is a Latin word which means tightened, pressed together and contracted.

It was first described by J F Meckel in 1950. Lippincott showed that out of 1943 patients with congenital heart disease, 6.3% had coarctation of aorta. Fredrickson calculated 7.2% of patients having coarctation of aorta out of 2000 patients with congenital heart disease.

Males are 2 to 5 times more commonly affected than females. Coarctation of aorta is commonly associated with other cardiac malformations. Perloff estimated that upto 40% patients have other associated defects. Edward showed that 85% of patients might have bicuspid aortic valve. They commonly become stenotic and incompetent.

VSD is seen in 10% of post-ductal coarctation. PDA, mitral stenosis and mitral regurgitation, ASD, transposition of great arteries and endocardial fibroelastosis are reported.

Severity of lesion is variable from complete atresia to very slight narrowing. Proximal and distal aortic segments are distended and dilated. In addition to hypertension, epistaxis, dyspnoea, headache, signs of left ventricular failure and radiofemoral delay are quite characteristic. Time delay has been calculated to be one second between radial and femoral arteries. Systolic murmurs and bruit over collaterals is a common finding.

On ECG, LVH, LBBB and AF are the usual findings. LVH is usually out of proportion to the degree of hypertension specially if there is aortic stenosis. On X-Ray chest, rib notching at inferior costal margins is seen. Reverse 3 sign is seen in the region of aortic knuckle, large left subclavian artery makes the upper part of 3 and lower part is formed by post-stenotic dilatation of the aorta below the coarctation.

Some workers have classified coarctation of aorta into two sub-groups.

A- Adult type (post ductal coarctation, ductus arteriosus usually closed, prestenotic hypertension and LVH). Average age at death is 35 years. 60-70% of all patients die before their 40th year of life. The usual causes of death are: rupture of ascending aorta, bacterial endocarditis and CCF or intracranial haemorrhage due to berry aneurysms which is usually associated with this type.

B - Infantile type (Preductal coarctation, PDA, pulmonary hypertension and right to left shunt and RVH). More than 50% of patients die during first month and more than 80% die in first three months of life.

Surgical resection of coarctation and end to end anastomosis or graft of aortic segments is the treatment of choice. It is better to do it as early as possible as the prognosis is good if surgery is done early. Angioplasty has been tried with some success, though re-stenosis is a problem.

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