CASE REPORT

Coarctation of the aorta with left pulmonary artery stenosis: a rare association diagnosed with ECG-gated multislice dual-source CT angiography

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SUMMARY

We report an extremely rare case of coarctation of the aorta with associated left pulmonary artery stenosis. This violates the traditional fetal flow pattern theory. Two-dimensional echocardiography, although being the initial imaging modality for diagnosing coarctation of the aorta, is limited in the demonstration of branch pulmonary arteries. Retrospective ECG-gated multidetector CT angiography is a non-invasive imaging technique for comprehensive assessment of the site, length and severity of the stenosed aortic segment. It is also helpful in differentiation of an interrupted aortic arch from severe coarctation. Associated pulmonary arterial and venous system anomalies can be evaluated, which dictates the management of aortic coarctation.

BACKGROUND

Coarctation of the aorta is a common congenital cardiovascular anomaly. We report an exceptionally uncommon case of aortic coarctation with associated left pulmonary artery stenosis. The association of coarctation of the aorta with right-sided obstructive lesions is a very rare entity, with only one case reported in the literature.^{1 2}

CASE PRESENTATION

A 2-month-old infant was presented with wheezing and poor feeding. There was no history of congenital heart disease in the family.

INVESTIGATIONS

Clinical examination revealed no facial dysmorphism. Bilateral femoral pulses were feeble and a radial-femoral pulse delay was noted. There was evidence of upper limb systolic hypertension. A right parasternal heave was present. On auscultation, an ejection systolic murmur was heard over the pulmonary area. ECG and chest X-ray were normal. Two-dimensional echocardiography revealed turbulent flow in the descending thoracic aorta, distal to the origin of the left subclavian artery. The right ventricular outflow tract and pulmonary valve were reported normal on echocardiography. Retrospective ECG-gated multislice (2×128 slice using z-focal flying spot technique), dual-source multidetector CT (Somatom Definition Flash, Siemens, Germany) angiography was performed. Non-ionic contrast material was injected with bolus tracking over the ascending aorta. Parameters for scanning included slice thickness 0.6 mm and rotation time 0.28 s. The heart rate

was 80 bpm during the study. Axial images were reconstructed at various time points along the R-R interval. Reconstructed images were then processed (Syngo Via software) using standard algorithms. Axial images, three-dimensional volume-rendered (VR), multiplanar reconstruction and maximum intensity projection images were integrated together. CT cardiac angiography showed short segment severe aortic narrowing in the descending aorta approximately 14 mm distal to the origin of the left subclavian artery. Post-stenotic dilation was noted (figure 1A). The right ventricular outflow tract showed ballooning at the valvular level with no obvious supravalvular stenosis (figure 1B). The main pulmonary and right pulmonary artery appeared normal. Mild focal narrowing was noted at the origin of the left pulmonary artery (figure 2). No evidence of collateralisation or patent ductus arteriosus was noted.

TREATMENT

Treatment of aortic coarctation depends on various clinical, echocardiographic and imaging findings. Significant hypertension or congestive heart failure is an indication for intervention. Surgical relief of the aortic obstruction and catheter interventional techniques (balloon angioplasty and stents) are available alternatives. Balloon angioplasty was performed in this child with native coarctation. There was a reduction in the mean systolic pressure gradient from 21 mm Hg to 8 mm Hg. There were no immediate complications and the patient was stable. Balloon angioplasty was then performed for left pulmonary artery stenosis in the second sitting. No postprocedural complications were noted.

OUTCOME AND FOLLOW-UP

The patient clinically improved with reduction in upper limb systolic blood pressure and normal volume of femoral pulses. Oral intake improved significantly with normal weight gain. The patient is on regular follow-up in cardiology clinic.

DISCUSSION

Coarctation of the aorta is a congenital vascular anomaly characterised by narrowing of the aorta, typically at the aortic isthmus. However, a level of obstruction is also known to occur in the aortic arch or abdominal aorta. Various cardiac and non-cardiac anomalies can be associated with coarctation of the aorta, and is seen in up to 50% of cases.^{3–5}

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Figure 1 (A) VRT image showing severe coarctation (arrow) of descending thoracic aorta with post-stenotic dilation. (B) VRT image showing ballooning at pulmonary valve level (arrow) with no obvious supravalvular stenosis (arrowhead).

Figure 2 (A and B) Axial maximum intensity projection (A) and VRT (B posterior view) images showing mild focal narrowing of left pulmonary artery (arrow) with ballooning at valvular level (arrowhead in A) LPA, left pulmonary artery; RPA, right pulmonary artery.

The junction of the isthmus and the descending thoracic aorta is determined by an indentation over the posterior wall of the aorta, opposite the site of the ductus arteriosus. In conditions such as tetralogy of Fallot, where the pulmonary blood flow is reduced, there is presence of increased aortic blood flow with consequent aortic dilation. Hence, in pulmonary artery stenosis, coarctation of the aorta is not expected. Conversely, in intracardiac anomalies where blood is diverted away from the aorta to the pulmonary artery, isthmal narrowing or posterior indentation may be exaggerated; this results in isolated juxtaductal coarctation.⁶

In a study by Shinebourne and Elseed, 162 patients with coarctation of the aorta were analysed, of whom 83 showed

increased pulmonary blood flow resulting from intracardiac lesions, and 21 showed left-sided anomalies. In this study, no patient with coarctation revealed reduced pulmonary blood flow or right-sided obstructive pathology. From their study they concluded that coarctation is prevented when flow in the main pulmonary artery is reduced in the fetus. This hypothesis is the fetal flow pattern theory. However, in our patient, coarctation of the aorta was associated with right-sided obstructive lesion, at the branch pulmonary artery level (left pulmonary artery stenosis). This association has not been reported previously and is contrary to the traditional fetal flow theory.⁶

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Multidetector CT angiography is an indispensable tool for comprehensive evaluation of aortic coarctation and associated pulmonary arterial and venous system anomalies.
- ▶ Right-sided obstructive lesions are not necessarily prevented in aortic coarctation, as previously thought. Hence, a careful evaluation of the pulmonary artery and its branches is vital, as this dictates the further surgical management.
- ▶ The traditional fetal flow pattern theory describes the haemodynamic relationship between the aortic and pulmonary circulation; wherein this hypothesis explains the prevention of aortic coarctation in right-sided obstructive lesions.

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