Infantile (Preductal) Coarctation of Aorta Presenting as Cerebellar Infarct – A Rare Presentation

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Abstract
Coarctation of aorta is a common congenital heart defect. The diagnosis may be missed unless a high index of suspicion is maintained, and is often delayed until the patient develops congestive heart failure (CHF), (common in infants) or hypertension (common in older children). It seldom goes undiagnosed till adulthood and frequently leads to complications as a result of long-standing high blood pressure. Intracranial haemorrhage, premature coronary artery disease, aortic aneurysms and rupture have all been reported. But it is rare to see a patient with preductal (infantile) coarctation survive childhood and presents with an infarct in adulthood. We herein present a case report of a young woman who came with vomiting and giddiness and was diagnosed as a case of cerebellar infarct due to a concealed preductal (infantile) coarctation of aorta.

Introduction
Coarctation of the aorta (CoA) is a relatively common defect that accounts for 5-8% of all congenital heart defects and is found at autopsy in up to 1:1550 patients.1 It is approximately three times more common in males.2 The traditional classification (Figure 1) into infantile (preductal) and adult (postductal) types is now regarded as too simplistic, since many patients with preductal lesions do not present until adulthood.3 A spectrum of lesions is now recognized, and it is only those with the most severe obstruction (e.g. aortic arch atresia or interruption) or associated cardiac defects who invariably present in infancy. Most other cases are now identified at routine medical examination. Otherwise, age rather than the site of obstruction, of cardiac failure or occasionally cerebral vascular accident (CVA), aortic dissection, or endocarditis.4

Case Report
A 32-year-old woman presented with symptoms of headache, nausea, vomiting and giddiness while walking. Her long-standing hypertension, which had recently become more difficult to control, was being treated with triple-drug therapy (amlodipine, losartan, and clonidine). She was a known case of chronic infarct in left frontoparietal region diagnosed by CT scan in 2010. Her evaluation revealed a radio-systolic murmur was heard in the aortic area, which was transmitted to both carotid arteries. Weak femoral pulses were palpated bilaterally. Laboratory evaluation revealed blood count and biochemistry results within normal limits. Chest X-ray showed inferior rib notching bilaterally from 3rd to 5th ribs (Figure 2). Ultrasonography of abdomen was within normal limits. 2D Echocardiography revealed eccentric left ventricular hypertrophy. Non-contrast CT brain revealed chronic infarct in left frontoparietal region of 16 × 10 mm with hydrocephalus. MRI brain revealed acute infarct in left cerebellar hemisphere with old infarction in left frontoparietal region.

CT angiography showed diffuse narrowing of arch of aorta just distal to brachiocephalic artery with more discrete narrowing of aorta distal to origin of left subclavian artery s/o femoral delay and a difference of 60 mm Hg in blood pressure measured in the right and left arms (190/100 vs 130/90 mmHg, respectively). On physical examination, cerebellar signs were present on the left side. On auscultation, heart sounds were normal but a harsh systolic murmur was heard in the aortic area, which was transmitted to both carotid arteries. Weak femoral pulses were palpated bilaterally. Laboratory evaluation revealed blood count and biochemistry results within normal limits. Chest X-ray showed inferior rib notching bilaterally from 3rd to 5th ribs (Figure 2). Ultrasonography of abdomen was within normal limits. 2D Echocardiography revealed eccentric left ventricular hypertrophy. Non-contrast CT brain revealed chronic infarct in left frontoparietal region of 16 × 10 mm with hydrocephalus. MRI brain revealed acute infarct in left cerebellar hemisphere with old infarction in left frontoparietal region.

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Fig. 1: Types of aortic coarctation: (a) Atresia of ascending aorta; (b) pre-ductal infantile coarctation; (c) post-ductal infantile coarctation; (d) adult coarctation distal to left subclavian artery; (e) adult coarctation proximal to left subclavian artery; (f) adult coarctation at the left subclavian artery

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Chest X-ray showed inferior rib notching bilaterally from 3rd to 5th ribs

Discussion

Coarctation of the aorta, a congenital narrowing of the aorta that usually occurs near the site of insertion of the ductus arteriosus, is classified anatomically as preductal (infantile) or postductal (adult). Aortic coarctation has an incidence of 0.2 to 0.6 per 100 live births and accounts for 5% of all cases of congenital heart disease. The clinical presentation varies from cardiovascular collapse in infancy to asymptomatic hypertension in older children and in adults. Adults who have coarctation tend to present with proximal systemic hypertension that manifests as headaches, epistaxis, or cerebral aneurysm rupture. Ischaemic strokes have not been reported in literature. Arteriosclerosis occurring due to uncontrolled hypertension may be responsible for recurrent ischaemic strokes in our patient. Left untreated, such patients have a lower than average survival rate. Usually the patients with preductal coarctation do not survive beyond infancy. But well developed collaterals may be the reason for improved survival in our case.

Because there is essentially no medical treatment for older children or adults with coarctation, they are usually referred for surgical repair.

Conclusion

The presentation of infantile aortic coarctation for the first time in adulthood is unusual. We are reporting this case for its rarity as the patient had severe symptoms and uncontrolled hypertension and recurrent infarct caused by preductal (infantile) coarctation of aorta.

References