CASE REPORT

Discovery of coarctation of the aorta following renal doppler sonography

Sidney, Ching Liang Ong, FRCR¹, Li Shyan, Ch'ng, FRCR², Aida Binti Abdul Aziz, MMed(Rad)²

¹ Department of Radiology, International Medical University, Seremban, Negeri Sembilan, Malaysia, ²Department of Radiology, Hospital Sungai Buloh, Sungai Buloh, Selangor, Malaysia

SUMMARY

Coarctation of the aorta (CoA) is present in 0.4% of live births and in 7% of patients with congenital heart disease. While there may be florid presentations of congestive heart failure in the neonatal period, the diagnosis during adulthood is often delayed. We encountered a 20-year-old woman who was discovered to be hypertensive on routine check-up. Following bilateral abnormal renal doppler sonography, MR angiogram revealed a short-segment stenosis of the descending thoracic aorta. Review of her chest radiograph showed a small aortic knuckle. This case highlights an unconventional algorithm in diagnosing aortic coarctation in adulthood.

INTRODUCTION

Young-onset hypertension is arbitrarily defined as hypertension in individuals below 40 years of age.¹ Hypertension involving young individuals may be secondary in nature and renovascular causes have to be investigated as they are potentially treatable.

Common causes of renovascular hypertension are fibromuscular dysplasia (mid-segment renal artery stenosis) and atherosclerotic disease (proximal/ ostial stenosis). The first line of imaging is doppler sonography assessment of the renal arteries. Ultrasound is an excellent screening tool as it is noninvasive, affordable, repeatable and accurate.² This study involves direct visualisation of the vessels or via interpretation of the spectral waveforms. Following abnormal ultrasound studies, the patient needs to undergo another scan (CT angiogram, MR angiogram or catheter angiogram) to further characterise the abnormality.

CASE REPORT

An asymptomatic 20-year-old woman was found to have high blood pressure on routine medical assessment – blood pressure reading of 168/94mmHg at initial presentation. There was a recorded systolic blood pressure as high as 188mmHg during follow-up. Clinical examination was unremarkable. No radioradial or radiofemoral delay. No significant difference in blood pressure readings obtained from bilateral upper and lower limbs.

She was then referred for doppler ultrasound of the kidneys to look for renal artery stenosis. Pulsus parvus et tardus

waveforms were present bilaterally with average acceleration time (AT) of 0.2s (normal AT should be <0.07s) [Fig 1]. Similar waveforms were also seen at the abdominal aorta, at the level of renal arteries. A diagnosis of bilateral renal artery stenosis was established. Subsequent MR Angiogram (MRA) abdomen showed normal appearance of the renal arteries and abdominal aorta. However, there is a severe, shortsegment stenosis of the descending thoracic aorta, 3.5cm distal to the origin of the left subclavian artery [Fig 2]. There are also bilateral enlarged intercostal collateral arteries [arrows, Fig 2]. Retrospective review of the chest radiograph showed smooth narrowing of the proximal descending aorta with small aortic knuckle. No demonstrable inferior rib notching.

Finally she was referred to a cardiac and vascular centre for further workup and treatment. Follow-up records at the tertiary referral centre showed that her condition was managed conservatively without further imaging. Her hypertension was under control with oral Amlodipine 10mg od.

DISCUSSION

Traditionally, CoA is classified into preductal, juxtaductal or postductal in relation to the ductus arteriosus.3 However, this classification does not take into account the degree of narrowing, associated anomalies and relation with arch vessels.³ Recently, CoA is classified into 'simple' when isolated and 'complex' when there are accompanying cardiac anomalies.³

More severe forms of CoA will present during infantile period as congestive heart failure while milder forms typically present in late childhood or adulthood with upper limb hypertension, heart murmurs, lower limb hypoperfusion and rarely complications from hypertensive emergencies.^{3,4} Our patient had no significant clinical clues apart from high blood pressure readings.

Renal Doppler sonography is used to assess the renal arteries via visualization of main renal arteries (direct) or analysis of segmental arterial spectral waveforms (indirect). The direct method is time consuming (mean time of 69 minutes compared to 14 minutes for distal evaluation) and is technically difficult for patients with thick abdomen.² The indirect evaluation entails identifying the waveform pattern,

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Fig. 1: Doppler ultrasound images of the right upper pole and left upper pole segmental arterial waveforms with acceleration time (AT) of 0.2s.



Fig. 2: MRA aorta showing normal renal arteries, aortic coarctation and enlarged intercostal collateral arteries (arrows).

acceleration time and resistive index. Presence of pulsus parvus et tardus waveforms which refer to slow rise to a small peak systolic velocity suggests proximal stenosis. The presence of this type of waveforms in both kidneys may suggest bilateral fibromuscular dysplasia, bilateral atherosclerotic ostial narrowing of main renal arteries or as in our case, a coarctation at the descending thoracic aorta. Other causes of stenosis along the aortic blood flow such as aortic valve stenosis or aortitis (Takayasu) may also result in this finding. Hence, it is imperative to image the entire length of aorta following bilateral abnormal renal Doppler sonography.

There is no significant diagnostic difference between MRA abdomen (with and without contrast) and CTA abdomen (with contrast).⁵ However, MRA abdomen has the advantage of zero radiation risk to the patient. This modality may be more desirable in young patients with no contraindication to MRI. In our case, MRA showed normal appearance of the renal arteries. Coarctation of the descending thoracic aorta was only identified after assessing the entire length of aorta. Review of her chest radiograph showed a small aortic knuckle with no other tell-tale signs of CoA such as inferior rib notching or cardiomegaly. The more commonly associated "figure-of-3" appearance of the aortic knuckle could not be confidently established. This finding underlines a potential blind spot in the interpretation of a chest radiograph. Without the MRA, the diagnosis of CoA based on this patient's chest radiograph alone may be challenging.

CONCLUSION

This case emphasises the importance of being vigilant of a more proximal aortic stenosis in the event of bilateral abnormal renal doppler sonography. There must also be careful radiographic assessment of the aortic knuckle as this may potentially lead to expedited diagnosis of CoA.

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