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Congenital coarctation of the aorta: Role of peripheral vascular ultrasound in young hypertensive patients.

Introduction

Coarctation of the aorta (CoA) is a congenital heart disease that occurs in 3–4 per 10 000 live births with a male predominance. It is defined as a localized narrowing of the aortic lumen. Although it's the most common cause of non-endocrine secondary hypertension, CoA can still present a diagnostic challenge in those presenting beyond the neonatal period. In fact, CoA is missed in about 85 % of patients referred to a hospital for hypertension or murmurs (JI. Hoffman. Cardiovasc J Afr 2018; 29: 252–255).

Because of the concealment of CoA, we present an interesting case of a young male patient with CoA who was initially diagnosed by peripheral vascular ultrasound instead of physical examination or aortic imaging, thus providing a working diagnosis for physicians.

Case Description

A 20-year-old male college student with mildly elevated BP for 1 year was referred to our hospital due to ineffective anti-hypertensive medication and recent mild chest tightness. A basic physical examination showed a slightly elevated BP of about 140/100 mmHg on the left arm. Renal ultrasonography was first prescribed to exclude secondary hypertension caused by kidney or adrenal diseases. Two-dimensional ultrasound showed normal renal morphology and no adrenal tumors. However, pulse wave Doppler imaging of the renal arteries showed a significantly abnormal spectrum – the "tardus-parvus waveform", which is characterized by a decreased peak systolic velocity, delayed systolic acceleration, and rounding of the systolic peak (▶ **Fig. 1c**). Combined with the hypertension symptom, we speculated that this young man may have had congenital vascular stenosis. Pulse wave Doppler ultrasound examinations of the abdominal aorta, 4-limb arteries, and carotid artery were then conducted. The findings showed that all of the arteries of the lower body (▶ **Fig. 1c–f**) exhibited the "tardus-parvus waveform", while the blood flow spectrum of all of the arteries of the upper body was normal (▶ **Fig. 1a, b**). These positive findings further confirmed the assumption of congenital vascular stenosis. Subsequent transthoracic echocardiography (TTE) showed significant coarctation of the descending aorta, with a maximum flow velocity of about 478 cm/s, and an intracardiac abnormality of the bicus pid aortic valve. Subsequent cardiac computed tomographic angiography confirmed the presence of aortic coarctation (▶ **Fig. 1g**). The patient was thus transferred for interventional treatment. The patient recovered after stent implantation of the stenotic aorta and the BP returned to normal thereafter.

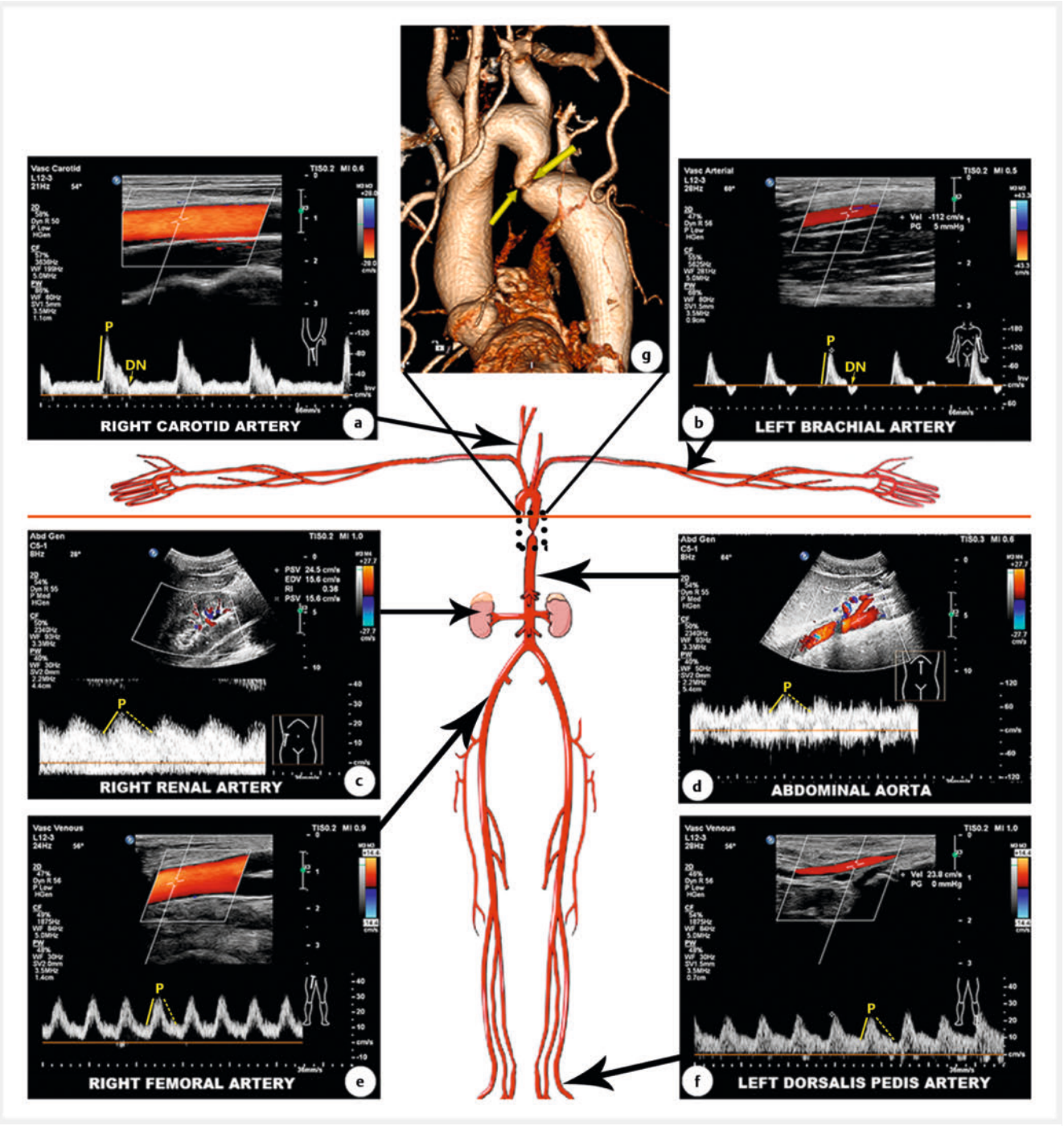
Discussion

CoA is one of the most common congenital cardiac anomalies. The incidence is higher among males than females, with a proportion of about 1.27–1.74:1 (M. Campbell et al. Lancet 1961; 1: 463–467). CoA refers to a narrowing of the proximal descending aorta. It can occur as an isolated lesion or can be associated with other cardiovascular abnormalities (JI. Hoffman et al. J Am Coll Cardiol 2002; 39: 1890–1900). Most coarctations fall into one of two groups: critical coarctation that causes symptoms within the first two months of life and subsequent death if not treated promptly, or asymptomatic coarctation that presents later, usually with hypertension in the upper limbs (JI. Hoffman. Cardiovasc J Afr 2018; 29: 252–255). In this case, the patient was a young male who had been asymptomatic for almost 20 years. The main reason for the medical consultation was mild hypertension, mild chest tightness, and ineffective medication. Echocardiography confirmed the associated abnormality of the bicuspid aortic valve.

According to the 2017 American Heart Association and American College of Cardiology guidelines (PK. Whelton et al. Circulation 2018; 138: e484–e594), patients who are newly diagnosed with hypertension should be screened for secondary causes, especially in those with suggestive signs, including an acute increase in previously well-controlled blood pressure, accelerated hypertension with evidence of end-organ damage, and a patient age at onset of younger than 30 years. In this case, the patient experienced hypertension, although mild, at an early age of only 20 years old. The possibility of secondary hypertension was considered and renal ultrasonography was first prescribed to exclude the causes of endocrine hypertension. However, a thorough and detailed physical examination was not performed, which may be due to the busy schedule of the physicians in the outpatient department or their over-reliance on modern examination technologies.

For patients with signs of a secondary cause, a focused and thorough physical examination can provide significant information. Classic physical examination findings of CoA could include discrepant 4-limb BP, weak femoral pulses, and/or cardiac murmur from collaterals or associated defects. Imran et al. (I. Asad et al. Pediatr Emerg Care 2021; 37: e1724–e1725) reported two cases of adolescents with undiagnosed CoA who were both initially misdiagnosed as essential hypertension until careful detailed physical examination later detected weak/absent femoral pulses and a discrepancy in the upper and lower limb blood pressures.

Although ultrasound may have restricted accessibility for the distal thoracic aorta in some patients, it is always the first-line imaging modality to evaluate the etiology and complications of hypertension in patients. This technology can be quickly and easily implemented in hospitals at all levels, including community hospitals. It can provide physicians with a real-time and interpretive display of vascular conditions (EA. Murphy et al. Cardiovasc Eng Technol 2017; 8: 255–272). Careful vascular ultrasound examination including the descending tho-



▶ **Fig. 1** Pulse wave Doppler spectra of peripheral arteries and 3D reconstruction image of aorta in a 20-year-old CoA patient. Vessels upstream from the stenosis including (**a**) the right carotid artery and (**b**) the left brachial artery show a normal blood flow spectrum: steep systolic acceleration (yellow solid line in panels a and b), sharp systolic peak (P), and significant dicrotic notch (DN). In peripheral vessels, the dicrotic notch is often reversed below the baseline due to higher distal resistance. Vessels downstream from the stenosis including (**c**) the right renal artery, (**d**) the abdominal aorta, (**e**) the right femoral artery, and (**f**) the left dorsalis pedis artery exhibit the typical "tardus-parvus waveform": delayed systolic acceleration (yellow solid line in panel c–f), rounded systolic peak (P) with decreased peak velocity, and slow deceleration (yellow dotted line in panel c–f). Visceral organs appear more rounded than peripheral vessels because of lower distal resistance. (**g**): 3D reconstruction image of aortic arch and descending aorta which shows significant stenosis of the descending aorta (indicated by yellow arrows). Abbreviations: 3D: Three dimensional; CoA: Coarctation of the aorta; P: Systolic peak; DN: Dicrotic notch.

racic aorta, abdominal aorta, renal artery, and 4-limb artery could provide a rapid overview of the patient's overall vascular condition. Furthermore, for atypical aortic coarctation like middle aortic coarctation, ultrasound could also be used for the differential diagnosis of congenital and acquired types by clearly delineating both the outer and inner diameter (KT. Delis et al. Perspect vasc surg endovasc ther 2005; 17: 187–203).

CTA has incomparable advantages for diagnosing CoA, especially for evaluating the extent and length of the narrowed segment and assessing the collaterals between the proximal and distal segments (J. Wang et al. BMC Cardiovasc Disord 2022; 22: 27). MRI is mainly indicated in young individuals who require repetitive studies and long-term follow-up (A. Evangelista. Heart 2014; 100: 909–991). However, a quick and carefully performed extensive Doppler exam of the vascular system can also improve the diagnostic rate of CoA, especially when an abnormal Doppler waveform in the peripheral arteries is detected (AL. Pinto et al. Port J Card 2023; 30: 89–91).

Conflict of Interest

The authors declare that they have no conflict of interest.

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