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An unusual cause of hypertensive emergency

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A 42-year-old woman was referred to our consulting room after a first episode of hypertensive emergency (blood pressure 197/105 mmHg) with headache. She had suffered from high blood pressure for the last 18 years but currently without medication. Physical examination showed difference between superior (180/90 mmHg) and lower limb blood pressure (140/80 mmHg) and no difference between both superior limbs. A systolic murmur irradiated to the interscapular and abdominal regions was present. Despite normal echocardioscopy, under suspicion of aortic coarctation, a routine transthoracic echocardiogram (TTE) was made. The TTE showed mild hypertrophy of the left ventricle and no valvular disease. From suprasternal view, aortic arch was mildly dilated with no signs of aortic coarctation (Figure 1*A*). From subcostal view the abdominal aorta showed colour aliasing (Figure 1*B*) and a significant systolic gradient with diastolic tail (Figure 1*C*). A cardiac computed tomography was performed (Figure 2), showing a progressive decrease in aortic diameter from distal descending aorta to the thoracoabdominal junction where there was a minimum diameter of 7.2 × 8.3 mm. Left renal artery had a short stenosis from moderate to severe 1.5 cm from its origin, suggesting middle aortic syndrome (MAS).

Middle aortic syndrome can be acquired, such as Takayasu's disease and temporal arteritis, congenital or associated with genetic syndromes such as neurofibromatosis or Williams syndrome.[1,2] Hypertension proximal to the aortic stenosis is a characteristic finding of MAS. Other symptoms may be headache, dyspnoea, abdominal angina, or lower limb claudication.[1] The challenging diagnosis by TTE raises the importance of clinical suspicion to perform further image studies of the aorta. Surgical treatment should be offered when refractory hypertension, symptoms, or end-organ failure is present.[3] The absence of elevated inflammatory markers (C-reactive protein 1.44 mg/L) or constitutional symptoms and negative auto-antibodies suggested congenital MAS. Since renal function was normal (serum creatinine 0.56 mg/dL) and hypertension was controlled with Candesartan/hydrochlorothiazide 16 mg/12.5 mg, we adopted a conservative approach.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

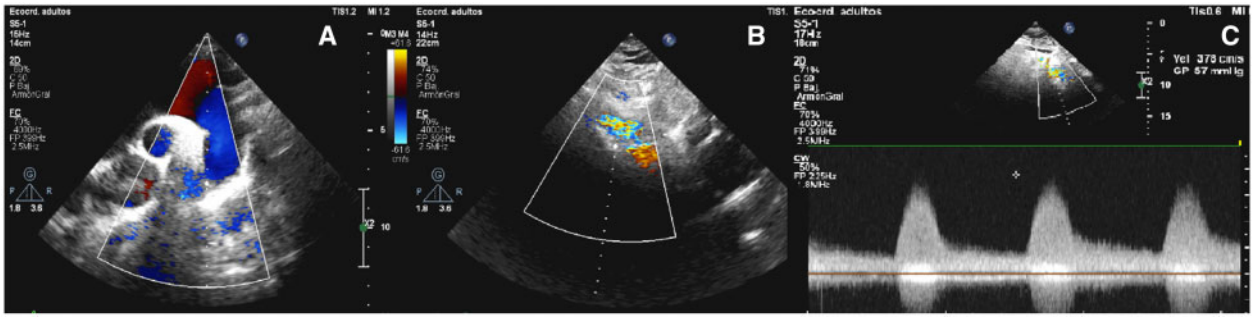

Figure 1 (*A*) Normal aortic arch; (*B*) abdominal aorta with flow acceleration; and (*C*) systolic gradient in abdominal aorta with diastolic tail.

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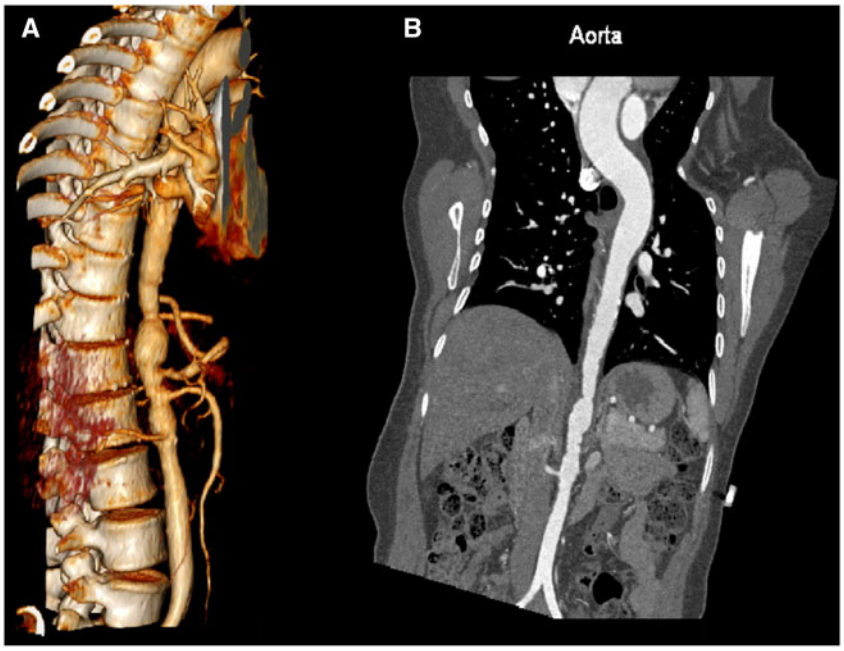

Figure 2 (*A*) Aortic computed tomography with three-dimensional reconstruction of descending and abdominal aorta; (*B*) two-dimensional view.

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