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Atypical Coarctation of the Aorta Revealed by Arterial Hypertension in a 22-Year-Old Young Man

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Abstract

Non-isthmic coarctation of the aorta is a rare congenital malformation in adults. Arterial hypertension is a frequent circumstance of discovery. We reported the case of a 22-year-old Guinean man who had been followed for 5 years for hypertension. Clinically, he presented with hypertension of the upper limbs with a systolic pressure gradient of 100 mmHg. The diagnosis was confirmed by thoracic angioscan, which showed a 65.8% coarctation of the abdominal aorta. He was on triple antihypertensive therapy combining Atenolol 100 mg, Amlodipine 10 mg and Perindopril 10 mg. He is awaiting interventional treatment. His blood pressure is stable at around 140/90 mmHg.

Keywords

Abdominal Aortic Coarctation, Arterial Hypertension, Young Subject, Congenital Malformation

1. Introduction

Typical coarctation of the aorta is a congenital malformation characterized by a localized narrowing, most commonly found at the isthmus; the region bounded by the origin of the left subclavian artery on one side and the insertion of the arterial ligament on the other [1]. However, although rare, it may be found in other locations. It is most often localized, but the narrowing may sometimes be more extensive.

Coarctation of the aorta is a frequent congenital malformation representing

6% to 8% of congenital cardiac malformations with an incidence of 3 to 4 per 10,000 births per year and a predominance of males with a sex ratio of 3:2 [1] [2] [3]. Isthmic coarctation of the aorta is the most common coarctation, whereas coarctation of the descending aorta and/or abdominal aorta is rare, accounting for only 0.5% to 2% of all coarctations of the aorta [4] [5].

The narrowing of the aorta is responsible for arterial hypertension upstream, hypo-perfusion downstream, the reduction or even abolition of pulses in the lower limbs and a systolic blood pressure gradient between the upper and lower limbs [6].

Mingorance *et al.* [7] reported a case of coarctation of the aorta in a 41-year-old woman with hypertension.

In Guinea, we have not found any published study on coarctation of the aorta in adults.

We report a case of atypical coarctation of the aorta revealed by hypertension in a young adult.

2. Observation

This is a 22-year-old student with asymptomatic hypertension who has been on Atenolol 100 mg for 5 years. He had no other cardiovascular risk factors and no known malformations.

The physical examination revealed hypertension in the upper limbs: 190/90 mmHg (right arm) and 190/80 mmHg (left arm).

Lower limb systolic blood pressure was 90 mmHg, giving a systolic pressure gradient of 100 mmHg.

The pulses in the upper limbs were strong, while those in the lower limbs were weak.

Auscultation revealed a diastolic regurgitant murmur of intensity 2/6 at the aortic focus. A systolic murmur of intensity 3/6 in the epigastrium along the abdominal aorta was found.

The rest of the clinical examination was normal.

The electrocardiogram showed sinus rhythm, left ventricular hypertrophy and 1st-degree atrioventricular block (**Figure 1**).

A frontal chest X-ray showed cardiomegaly with a cardiothoracic index of 0.59 and a few costal notches (**Figure 2**).

Transthoracic echocardiography showed moderate dilatation of the left ventricle, moderate rheumatic aortic insufficiency on a 3-cusp valve with no coarctation on the aortic isthmus, but the reduced diameter of the abdominal aorta with pulsed Doppler aliasing; continuous Doppler recorded a velocity of 5.86 m/s, as shown in **Figure 3**.

The thoracic angioscan showed a coarctation of the abdominal aorta of 65.8% (**Figure 4**).

In terms of treatment, he was on a strict diet, taking Atenolol 100 mg daily, Amlodipine 10 mg daily and Perindopril 10 mg daily.

He is awaiting interventional catheterisation.

His blood pressure remained stable at around 140/90 mmHg.

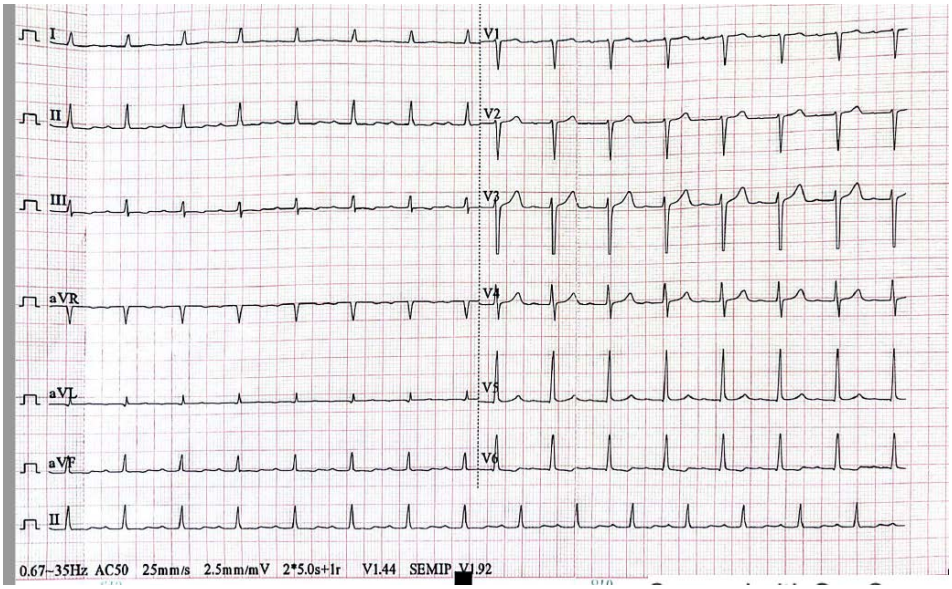


Figure 1. The electrocardiogram showed sinus rhythm, left ventricular hypertrophy and 1st-degree atrioventricular block.

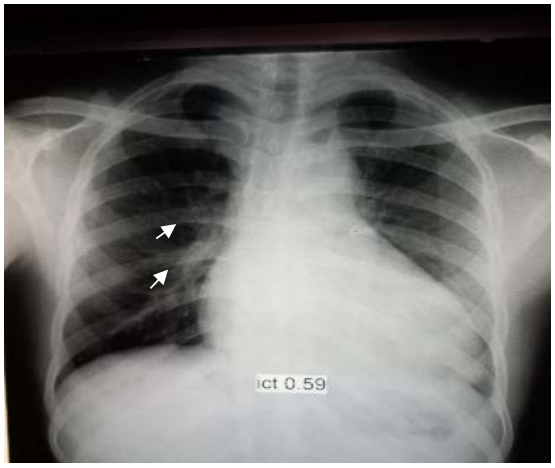
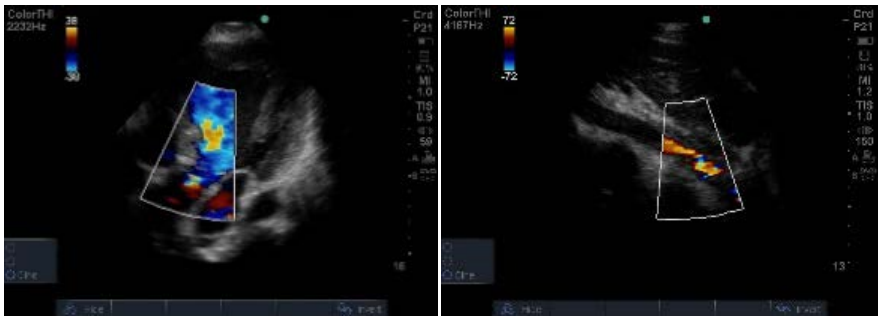
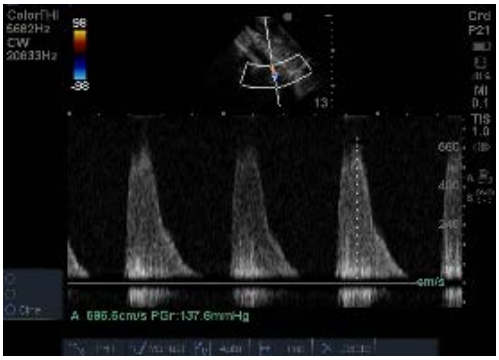


Figure 2. Frontal chest X-ray showing moderate cardiomegaly and costal notches on the right.



(a) (b)



(c)

Figure 3. Echocardiography showing (a) moderate aortic insufficiency, (b) narrowing of the abdominal aorta with colour aliasing, (c) velocity acceleration on continuous Doppler scan.

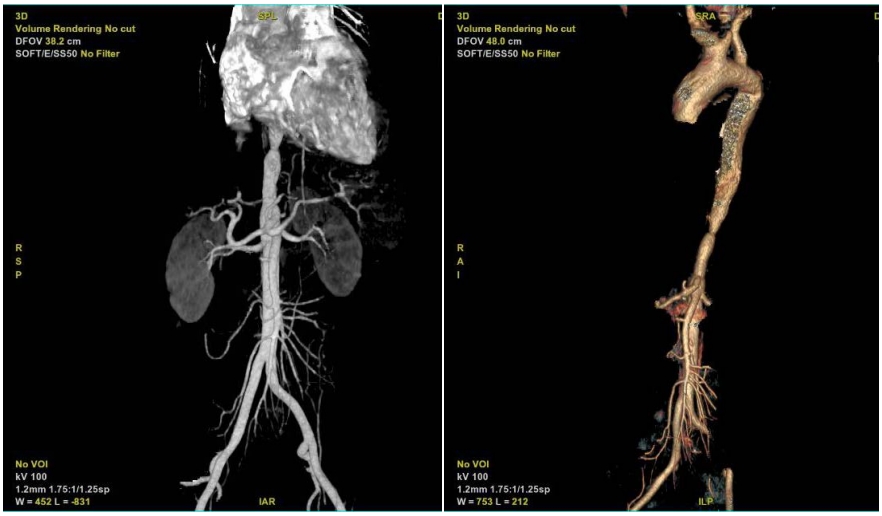


Figure 4. Angioscan with 3D reconstruction showing coarctation of the abdominal aorta.

3. Discussion

Coarctation of the descending and/or abdominal aorta is a rare vascular anomaly, accounting for only 0.5% to 2% of all coarctations of the aorta [4] [5]. This form, discovered in adulthood, is rarely described.

In terms of pathogenesis, many abdominal aortic stenoses appear to be linked to events occurring around the 25th day of foetal development. At this time, the two embryonic dorsal aortas fuse and lose their intermediate wall to form a single vessel. The exaggerated fusion of the two embryonic dorsal aortas or their failure to fuse with the subsequent obliteration of one of these vessels would predictably lead to aortic narrowing [8] [9]. Exaggerated fusion during the development of the two primitive dorsal aortas is supported in patients with reduced aortic diameters that have unique origins in the lumbar arteries [9] [10].

Congenital aortic coarctation can occur in isolation or as part of a spectrum of cardiac and extracardiac malformations such as aortic bicuspidism (60%), aortic arch hypoplasia (18%), interventricular septal defects (13%), mitral malformation

(8%), subaortic stenosis (6%) and elastopathy leading to aortic dissection [6]. Although bicuspidism is a frequent association [6], it was not observed in our patient, although moderate rheumatic aortic insufficiency was found.

Hypertension is by far the most common finding in adult aortic coarctation. This is arterial hypertension with a systolic pressure gradient between the limbs and the lower limbs. This gradient is significant when it is greater than 20 mmHg. This was the case with our patient, who had a pressure gradient of 100 mmHg. Inadequate blood supply to the lower limbs can also be manifested by weak or even imperceptible femoral pulses, or painful claudication on exertion [11].

The electrocardiogram typically shows signs of left ventricular hypertrophy or insufficiency [12], as was the case in our patient. In other cases, rapid supraventricular tachycardia with haemodynamic intolerance has been found [7].

Chest X-rays are of little diagnostic value but, if performed, can show mild cardiomegaly, costal erosions and, more rarely, the aortic notch of the coarctation [3]. Our patient presented with cardiomegaly and a few costal notches.

Two-dimensional echocardiography is often disappointing in adults. However, in echogenic patients and in the particular case of coarctation of the abdominal aorta, the subcutaneous incidence centered on the aorta allows color aliasing to be visualized, a high-velocity turbulent flow on continuous Doppler in the longitudinal section. The mean gradient across the coarctation can be used; it is significant when it is greater than 20 mmHg. Ultrasound can also be used to search for associated anomalies such as an anomaly of the aortic or mitral valve, aneurysmal dilatation of the ascending aorta or left ventricular hypertrophy in the case of hypertension [13].

Our patient presented with longitudinal incidence aliasing of the abdominal aorta on color Doppler with accelerated velocity on continuous Doppler.

Although MRI is generally preferred to reduce radiation exposure during repeated examinations, an angioscanner is more widely available [11]. In addition to making a positive diagnosis, it provides the necessary information on the location of the stenosis, its diameter, the length of the narrowed segment, the diameter of the upstream and downstream aortic segments, and collaterality, which will be a major factor in the indication and therapeutic strategy [12].

Our patient had undergone angioscanning which revealed aortic coarctation with a stenosis evaluated at 65.8%.

The first-line antihypertensive agents used in aortic coarctation are beta-blockers, ACE inhibitors/angiotensin II receptor blockers (ARBs) and calcium channel blockers (CCBs), often taken in combination. Our patient was on triple therapy combining a beta-blocker, an ACE inhibitor and a calcium channel blocker.

Endovascular treatment is the first-line procedure for non-extensive localized stenoses in adults with no other associated lesions. It reduces the risk of intra- and post-operative complications compared with conventional surgery, with satisfactory long-term results [4] [11]. The indication is significant aortic coarcta-

tion, which is defined by the combination of hypertension and a systolic pressure gradient of more than 20 mmHg, or a peak-to-peak gradient of more than 20 mmHg on catheterisation, or a mean gradient of more than 20mmHg on Doppler echocardiography [14].

When the left ventricular ejection fraction is impaired, a mean gradient of >10 mmHg across the coarctation by Doppler echocardiography is also an indication for endovascular intervention [14].

Our patient presented with hypertension associated with a systolic blood pressure gradient between the upper and lower limbs of 100 mmHg.

Without treatment, the prognosis for this disease is poor: 90% of patients die before the age of 58. The main causes of death are early coronary artery disease, left ventricular failure, aortic rupture or dissection, and ruptured intracranial aneurysms [12].

Cardiogenic shock c requiring high-dose inotropic agents, with multivisceral failure and associated paraparesis have been described in other studies [7].

Hypertension is common after aortic coarctation repair, particularly during exercise and when the repair is performed in adults. Ambulatory blood pressure monitoring and exercise testing are useful for diagnosis and management [12].

Aortic stenosis was estimated at 65.8%.

The first-line antihypertensive agents used in aortic coarctation are beta-blockers, ACE inhibitors/angiotensin II receptor blockers (ARBs) and calcium channel blockers (CCBs), often taken in combination. Our patient was on triple therapy combining a beta-blocker, an ACE inhibitor and a calcium channel blocker.

Endovascular treatment is the first-line procedure for non-extensive localized stenoses in adults with no other associated lesions. It reduces the risk of intra- and post-operative complications compared with conventional surgery, with satisfactory long-term results [4] [11]. The indication is significant aortic coarctation, which is defined by the combination of hypertension and a systolic pressure gradient of more than 20 mmHg, or a peak-to-peak gradient of more than 20 mmHg on catheterisation, or a mean gradient of more than 20 mmHg on Doppler echocardiography [14].

4. Conclusions

Coarctation of the abdominal aorta is rare and may be underestimated.

Our case highlights the need to systematically search for coarctation of the aorta in any young subject with arterial hypertension. Although pulse palpation and echocardiography are of great help, it is cross-sectional imaging that confirms the diagnosis. Medical treatment often involves triple or even quadruple antihypertensive therapy. Angioplasty, if available, remains the treatment of choice for the adult form.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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