Refractory hypertension and the lower limbs ischaemia as an aortic coarctation symptom

Katarzyna Drożdż[1, 2], Angelika Chachaj[1, 2], Paweł Gać[3, 4], Roland Fiszer[5], Jacek Białkowski[5], Andrzej Szuba[1, 2]

[1]Division of Angiology, Faculty of Health Science, Wroclaw Medical University, Wroclaw, Poland
[2]Department of Internal Medicine, 4th Military Teaching Hospital, Wroclaw, Poland
[3]Department of Radiology and Diagnostic Imaging, 4th Military Hospital, Wroclaw, Poland
[4]Department of Hygiene, Wroclaw Medical University, Wroclaw, Poland
[5]Department of Congenital Heart Diseases and Paediatric Cardiology, Silesian Centre for Heart Diseases, Medical University of Silesia, Zabrze, Poland

Abstract

Coarctation of the aorta (CoA) is a very rare cause of secondary hypertension, accounting for 0.1% of cases. The coarctation can present at any age. Hypertension is the most common presenting symptom. All patients with newly diagnosed hypertension must have a physical examination with assessment of the brachial and femoral pulses and measurement of brachial and popliteal blood pressures. A thorough physical examination is a crucial first step in diagnosing aortic coarctation.

Key words: hypertension; aortic coarctation; ABI; lower limb ischaemia

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Introduction

Coarctation of the aorta (CoA) is a very rare cause of secondary hypertension, accounting for 0.1% of cases [1]. The coarctation can present at any age, from childhood to adulthood. If not recognized in childhood, CoA has a high mortality rate, with an average life expectancy of only 35 years [2]. The untreated CoA in adults include hypertension, premature coronary artery disease, sudden cardiac death, heart failure, stroke, endocarditis, rupture, dissection, aneurysm and cardiovascular disease [3–7]. Hypertension is the most common presenting symptom. All patients with newly diagnosed hypertension, including children and young adults, must have a physical examination with assessment of the brachial and femoral pulses and measurement of brachial and popliteal blood pressures [2]. A thorough physical examination is a crucial first step in diagnosing aortic coarctation.

We present a case of 42-year-old woman with long–term resistant hypertension, aortic coarctation and ischaemia of lower limbs, successfully treated with percutaneous stent implantation.

A case report

Forty-two-year-old woman was admitted to the Department of the Internal Medicine because of the refractory hypertension. Hypertension was already diagnosed in the childhood. The combination antihypertensive treatment was all the time ineffective; currently the patient was treated with: betaxolol,

Address for correspondence: Paweł Gać M.D. Ph.D.
Department of Radiology and Diagnostic Imaging, 4th Military Hospital
Weigla 5, 50–981 Wroclaw, Poland, tel/fax: (+48) 261 660 480
e-mail: pawelgac@interia.pl

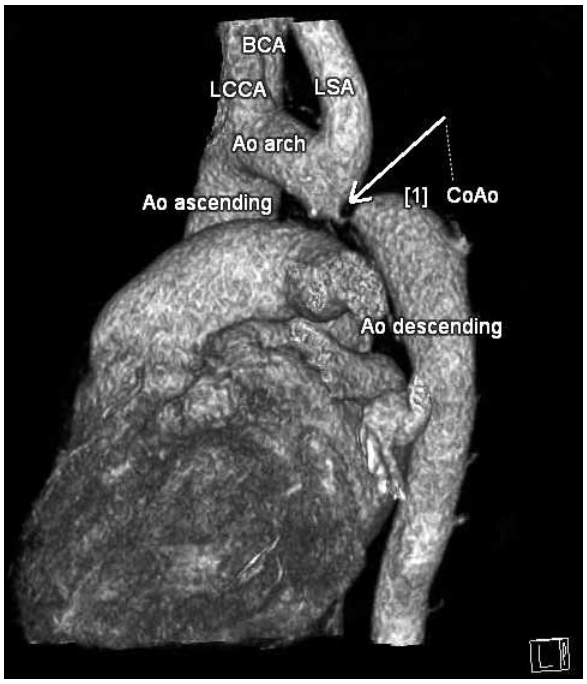

Figure 1. Tight stenosis distal to the left subclavian artery in angio-CT

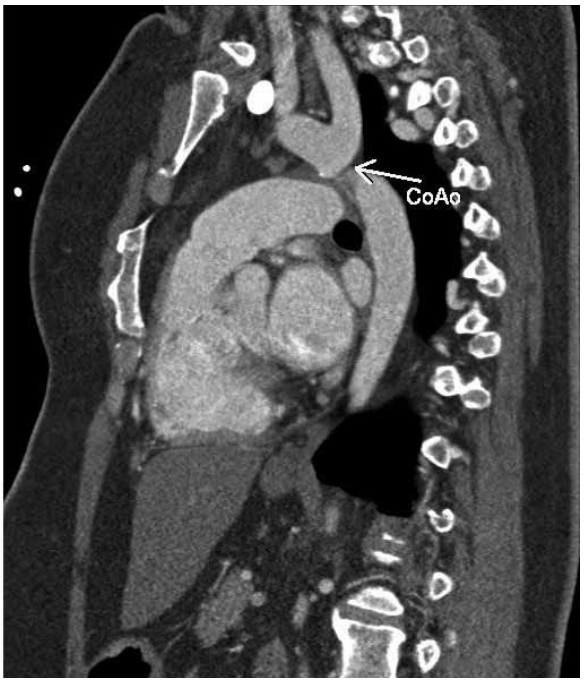

Figure 2. Tight stenosis distal to the left subclavian artery in angio-CT

amlodipine, and clonidine. The mean arterial blood pressure values were 180/100 mm Hg, with the highest value of 260/90 mm Hg. The patient was pregnant twice and delivered two children naturally. A few months before admission to the hospital, she complained of the lower limbs pain with intermittent claudication distance above 200 meters. No clinical evidence of heart or kidney failure was observed. At the time of the current admission, the physical examination revealed: obesity (BMI = 31), hirsutism, presence of the loud systolic murmur over the interscapular region, week pulse on lower limbs, blood pressure: 220/90 mm Hg on the right arm and 210/80 mm Hg on the left arm. The blood tests showed hypercholesterolaemia and impaired fasting glucose, with no abnormalities in the hormonal tests. The ABPM (ambulatory blood pressure monitoring) revealed the average blood pressure 172/79 mm Hg, the maximum 221/101 mm Hg, the minimum 133/51 mm Hg, with a normal night decline. The assessment of the hypertension-related organ damage showed second degree hypertensive retinopathy and concentric left ventricular hypertrophy with interventricular septum thickness of 20 mm. The chest x-ray revealed rib notching. The ABI (ankle-brachial index) was 0.66 on the right and 0.65 on the left side.

The results of diagnostic test arouse a suspicion of coarctation of the aorta. Computed tomography (CT) angiography confirmed tight stenosis distal to the left subclavian artery and the presence of extensive collateral circulation (Fig. 1–4). The stenosis was about 0.5 cm long, with cross sectional diameter up to 0.5 cm.

Magnetic resonance angiography of the head excluded aneurysm. The transcatheter angioplasty with placement of Covered Cheatham-Platinum Stents was performed (Fig. 5, 6). The peak systolic pressure gradient before and after stent implantation was 78 mm Hg and 14 mm Hg, respectively.

Four months after the procedure, the blood pressure remains correct, without any antihypertensive therapy. There is no more pain of the lower limbs with well-defined pulse rate and correct ABI values. The patient remains under follow-up.

Discussion

We demonstrate an extremely rare presentation of adult aortic coarctation in woman surviving into late adulthood with hypertension and lower limb ischaemia.

Coarctation of the aorta is one of the most common congenital cardiovascular defects, with an estimated birth incidence of 0.28 to 0.64 per 1000 live births, more common in males [8]. The heart defects that accompany the coarctation include: bicuspid aortic valve, ventricular septal defect, patent ductus arteriosus, transposition of the great

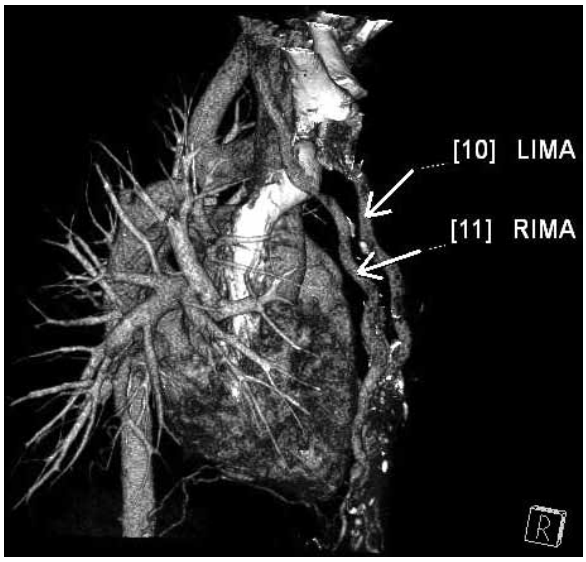

Figure 3. Tight stenosis distal to the left subclavian artery in angio-CT

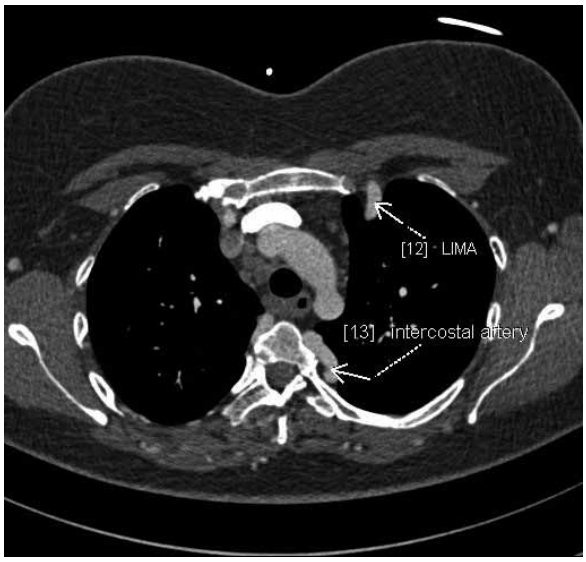

Figure 4. Tight stenosis distal to the left subclavian artery in angio-CT

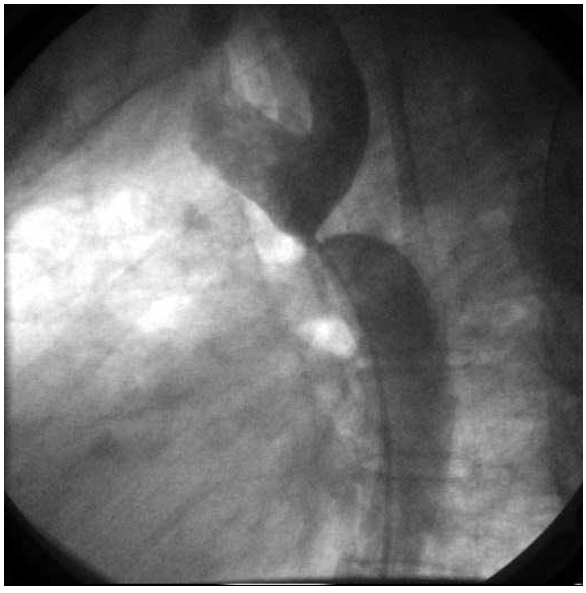

Figure 5. Coarctation of the aorta before the stent placement

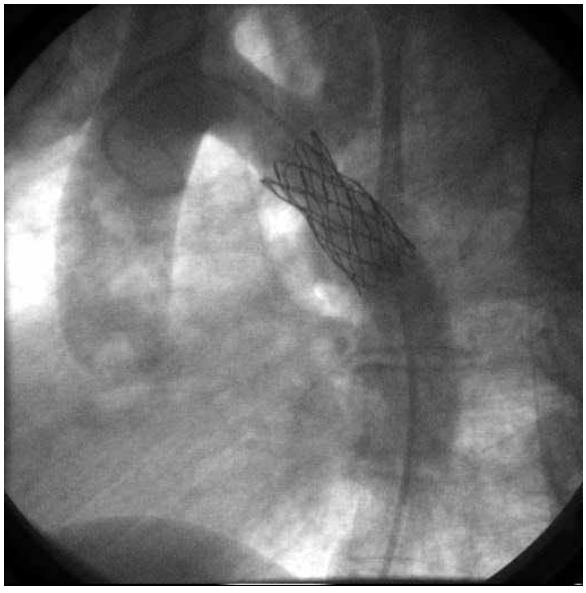

Figure 6. Coarctation of the aorta after the stent placement

arteries, atrioventricular canal defects, or left-sided obstructive heart defects with hypoplastic left heart syndrome [9–11].

The head CT or MRI must be performed to exclude the intracranial berry aneurysms, which are found in up to 15% of adult patients [12].

The age at diagnosis and symptoms of CoA depend on the severity of the lesion and associated diagnoses. The symptoms such as heart failure, acidosis, and shock following closure of the ductus arteriosus are most common in neonates with ductal-dependent or "critical coarctation" [13, 14]. If the symptoms are not so intense, about 30% of neonates with coarctation are discharge after delivery undiagnosed [15].

The hypertension is the most common presenting symptom for patients who enter adulthood undiagnosed [16]. Physical examination findings suggestive of coarctation are: diminished and/or delayed lower extremity pulses and a systolic pressure gradient between the upper and lower extremities [16].

The patient described in this report survived for more than 7 years beyond the expected age of death for untreated coarctation. She also failed to present with other consequences of persistent hypertension including congestive heart failure, atherosclerosis,

endocarditis, aneurysms and kidney insufficiency, even during pregnancy and natural births.

The only symptoms were hypertension, lower extremity blood pressures discrepant from those obtained in her upper extremities and systolic murmur over the interscapular region. Probably, if there wasn't a pain of the lower limbs with intermittent claudication distance above 200 meters, which became a new symptom for the patient, she would still remain undiagnosed.

This report encourages inclusion of aorta coarctation in a differential diagnosis for resistant hypertension, even in late adulthood. In addition, physical examination with assessment of the brachial and femoral pulses and measurement of brachial and popliteal blood pressures should be performed very carefully in all patients, especially those with hypertension. The European Society of Hypertension (ESH) and of the European Society of Cardiology (ESC) recommend the ABI assessment in patients with hypertension [17].

The current guidelines for the management of adults with aortic coarctation recommend intervention when the peak-to-peak coarctation gradient is 20 mm Hg or when there is a pressure difference > 20 mm Hg between the upper and lower limbs, with upper limb hypertension (> 140/90 mm Hg in adults), or significant left ventricular hypertrophy [2]. Surgical and interventional treatment of aortic coarctation is available. Surgery remains the treatment of choice in neonates with native aortic coarctation [18]. Balloon angioplasty with stent implantation is a viable therapeutic option for native and recurrent coarctation in adults with coarctation. It is less invasive and demonstrates lower complication rates. Stenting also carries a lower risk of re-coarctation and aneurysm formation [18–20]. Patients after aortic coarctation treatment require close monitoring over their lifetime for complications such as re-coarctation, aortic aneurysm, persistent hypertension and changes in any associated cardiac defects.

Conclusion

A thorough physical examination is a crucial first step in diagnosing aortic coarctation. The assessment of the brachial and femoral pulses with ABI measurements should be performed in all patients with newly diagnosed hypertension.

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