



CASE REPORT

Interrupted aortic arch: A misdiagnosed cause of hypertension



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Abstract We present the case of a 47-year-old man with hypertension for over 20 years, referred to our hospital due to mild aortic dilatation detected on a transthoracic echocardiogram. On physical examination weak lower limb pulses and a blood pressure differential of >50 mmHg between arms and legs were detected. Complete interruption of the aortic arch below the left subclavian artery was diagnosed by computed tomography angiography. With this case we aim to draw attention to aortic coarctation and interrupted aortic arch as potential causes of hypertension and to highlight the importance of the physical examination in the diagnosis of secondary causes of hypertension.

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PALAVRAS-CHAVE

Hipertensão;
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Interrupção do arco aórtico: uma causa de hipertensão mal diagnosticada

Resumo Apresentamos o caso de um homem de 47 anos, com hipertensão arterial desde há mais de 20 anos, referenciado ao nosso hospital por dilatação ligeira da aorta ascendente detetada em ecocardiograma transtorácico. No exame físico constatou-se que os pulsos femorais eram pouco amplos e existia um diferencial de pressões superior a 50 mmHg entre os membros superiores e inferiores. Por angiotomografia computadorizada do tórax diagnosticou-se uma interrupção completa do arco aórtico abaixo da artéria subclávia esquerda. Com este caso pretendemos relembrar a coartação da aorta e interrupção do arco aórtico como potenciais

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causas de hipertensão e realçar a importância do exame físico no diagnóstico de causas secundárias de hipertensão.

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Introduction

Hypertension is an increasingly important medical and public health issue, with an overall prevalence of 30–45% of the general population.¹ Appropriate diagnosis and treatment are important in reducing hypertension-related morbidity, mortality and costs. A detailed medical history and a rigorous physical examination have a pivotal role in the accurate diagnosis of hypertension, particularly for the detection of underlying secondary causes.²

Aortic coarctation and interrupted aortic arch (IAA) are examples of rare potential secondary causes of hypertension in adults. These entities can be easily diagnosed on physical examination, by palpation of the brachial and femoral arterial pulses and blood pressure measurement in the four limbs. However, there are reports showing that the diagnosis is often missed and a significant number of asymptomatic subjects are probably not detected until adulthood.³

Case report

We present the case of a 47-year-old male patient with a history of untreated high blood pressure since the age of 25. At 40 years old, he suffered an intracranial hemorrhage due to rupture of a right anterior choroidal artery aneurysm and underwent aneurysm clipping. He was then referred to an outpatient hypertension clinic for management and etiological study. Laboratory and imaging studies were performed with no apparent secondary causes of hypertension detected. With a presumed diagnosis of essential hypertension, the patient remained asymptomatic and with well-controlled blood pressure in the following years, taking irbesartan 300 mg plus hydrochlorothiazide 12.5 mg/day and amlodipine 5 mg/day.

At the age of 47, he was referred to our hospital's outpatient cardiology clinic due to mild dilatation of the aortic root and ascending aorta detected on a transthoracic echocardiogram (Figure 1A). The aortic valve was tricuspid with mild central regurgitation, and the left ventricle presented normal dimensions, mild concentric hypertrophy and preserved systolic function (Figure 1B–D). Due to a poor suprasternal view, the aortic arch could not be observed. Blood pressure was 135/80 mmHg in both arms. Lower limb pulses were weakly palpable and blood pressure was significantly lower (80/60 mmHg), so aortic coarctation was considered. No cardiac, dorsal or abdominal murmurs were audible. On the 12-lead electrocardiogram he had incomplete right bundle branch block and left ventricular

hypertrophy. Chest radiography showed normal cardiac silhouette and bilateral rib notching (Figure 2).

Multislice computed tomography angiography revealed an unexpected finding: complete interruption of the aortic arch 15 mm distal to the origin of the left subclavian artery (LSA) (Figure 3A and B). The ascending aorta was slightly dilated (42 mm) and the descending thoracic and abdominal aorta were hypoplastic (diameters of 16 mm and 11 mm, respectively). A massive collateral network, predominantly through the internal mammary and intercostal arteries, ensured circulation to the hypoplastic distal aorta (Figure 3C and D).

The patient was asymptomatic, with controlled hypertension and no symptoms of claudication, despite the distal aortic hypoplasia and pressure gradient between upper and lower limbs. The case was considered for surgical repair but, in view of the absence of symptoms, good blood pressure control and effective collateral circulation, a strategy of delayed intervention and close clinical surveillance was chosen.

Discussion

IAA is a rare congenital malformation that occurs in 3/1 000 000 live births, accounting for 1% of all congenital heart disease.⁴ It is defined as complete discontinuity of the aortic lumen between the ascending and descending portions of the thoracic aorta. IAA is commonly associated with additional cardiovascular anatomic defects, such as patent ductus arteriosus, ventricular septal defect, bicuspid aortic valve, subaortic stenosis, truncus arteriosus, aortopulmonary window and double outlet right ventricle. Rarely, IAA is an isolated finding without other associated congenital anomalies. A classification system introduced by Celoria and Patton in 1959⁵ divides IAA into three types: type A – interruption distal to the LSA; type B – interruption between the left common carotid artery and the LSA; and type C – interruption between the brachiocephalic trunk and the left common carotid arteries. In infants, type B is the most common (53%), whereas in adults type A has been more often reported.

Almost all cases of IAA are diagnosed in early childhood or the neonatal period, and clinical presentation involves severe congestive heart failure. If left untreated, 90% of affected infants die in the first year of life,⁶ the majority in the first few days due to the spontaneous closure of the ductus arteriosus. Consequently, IAA is only very rarely diagnosed in adult patients, and few cases of persons having reached adulthood with IAA have been reported

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