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ORIGINAL ARTICLE

Aortic coarctation misdiagnosed as a descending thoracic aorta aneurysm

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KEYWORDS
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Abstract Aortic coarctation (AC) represents ~7% of congenital cardiac diseases and is usually diagnosed in childhood or early adult life, depending on the severity of obstruction and associated malformations. Left untreated fewer than 20% of patients survive to age 50. We describe a case of thoracic AC, diagnosed at age 61, in a woman with known hypertension since age 45. At age 56 the patient was admitted with a subarachnoid hemorrhage and, during cerebral angiography, a thoracic aortic aneurysm was detected. Four years later the patient was referred to the outpatient hypertension clinic due to uncontrolled hypertension and cardiac failure. The echocardiogram disclosed left ventricular hypertrophy and aggressive treatment failed to control her hypertension. At age 61, due to lower limb muscular fatigue, arterial Doppler ultrasound was performed that revealed symmetrically decreased ankle/brachial pressure index, suggesting aortic stenosis. MRI angiography enabled a diagnosis of AC with a large poststenotic dilation which had been interpreted as an aortic aneurysm in successive CT scans. The authors highlight the unusually late clinical presentation and misdiagnosis despite extensive radiologic investigation. The subarachnoid hemorrhage was probably a disease manifestation, since berry aneurysms are among the noncardiac malformations associated with AC.

PALAVRAS-CHAVE
Coartação da aorta;
Hipertensão;
Aneurisma da Aorta

Coartação da aorta diagnosticada como um aneurisma da aorta torácica descendente

Resumo A coartação da aorta (CA) representa ~7% das cardiopatias congénitas sendo habitualmente diagnosticada na infância ou no início da vida adulta, dependendo da gravidade da obstrução e das malformações associadas. Não tratados, menos de 20% dos doentes sobrevivem até aos 50 anos. Descreve-se um caso de CA torácica, diagnosticada aos 61 anos, numa mulher com HTA conhecida desde os 45. Aos 56 anos foi internada por hemorragia subaracnoideia e, aquando da realização de angiografia cerebral, foi detetado aneurisma da aorta torácica. Quatro anos mais tarde foi referenciada à Consulta de Hipertensão por HTA não controlada e insuficiência cardíaca. O ecocardiograma mostrava hipertrofia ventricular esquerda e

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a HTA mantinha-se não controlada apesar de polimedicada. Aos 61 anos, por queixas de fadiga muscular intensa dos membros inferiores, efetuou Doppler Arterial que identificou índices de pressão sistólica diminuídos, bilateralmente e de forma simétrica, sugerindo estenose da aorta abdominal. A angio-RMN permitiu o diagnóstico de CA torácica com importante dilatação pós-estenótica, que foi interpretada em TAC seriadas como correspondendo a aneurisma da aorta descendente. Destaca-se a apresentação clínica tardia e diagnóstico erróneo, apesar de extensa investigação radiológica. Salienta-se ainda que a hemorragia subaracnoideia foi, provavelmente, uma manifestação da doença já que os aneurismas do polígono de Willis são uma das malformações extracardíacas associadas à CA.

Introduction

Aortic coarctation is a relatively common abnormality that occurs in approximately 6–8% of patients with congenital heart disease.[1] It can occur in isolation – simple coarctation – or in association with other cardiac and/or noncardiac lesions – complex coarctation.[2] The most common intracardiac lesions (present in approximately 50% of cases) are patent ductus arteriosus (closely related to the proposed pathogenesis of AC[3]), ventricular and atrial septal defects, and bicuspid aortic valve. Chromosome abnormalities and prematurity are the most common noncardiac conditions associated with AC. Extracardiac vascular abnormalities are also common, including anomalies of the brachiocephalic artery and its branches – stenosis or aneurysm – and aneurysms of the circle of Willis (berry aneurysms).[2]

The clinical features depend largely on the nature of the associated cardiac lesions and severity of the narrowing. Diagnosis is usually made during childhood or early adult life; AC is almost always evident on physical examination and classic radiologic findings. However, left untreated, as the collateral circulation develops the typical clinical signs may disappear. The young adult with uncorrected coarctation is usually asymptomatic. The most frequent manifestation is hypertension (especially systolic hypertension) and, later on, congestive heart failure and coronary artery disease.

In an autopsy series of uncorrected coarctation, 50% of patients had died by age 30 and 90% by age 60.[4] The most common causes of death are aortic rupture or dissection, cerebral hemorrhage due to aneurysm rupture and congestive heart failure. The former usually occurs before age 30, while congestive heart failure is a late consequence of long-standing hypertension and accelerated coronary artery disease.

Case report

In 2001, a 56-year-old Caucasian woman, obese, with hypertension since age 45 and no other relevant medical history, was admitted to the emergency room of our hospital with a subarachnoid hemorrhage. Cerebral angiography was performed and incidentally disclosed the presence of a thoracic aortic aneurysm (7 cm long, 6 cm maximum diameter), below the aortic arch. After discharge the patient was referred to the vascular surgery outpatient clinic for follow-up.

Over the course of three years, several computed tomography (CT) scans were performed, which showed no progression of the aneurysm (Figure 1).

The hypertension was controlled with a diuretic and the ECG was normal, with no evidence of left ventricular hypertrophy (LVH).

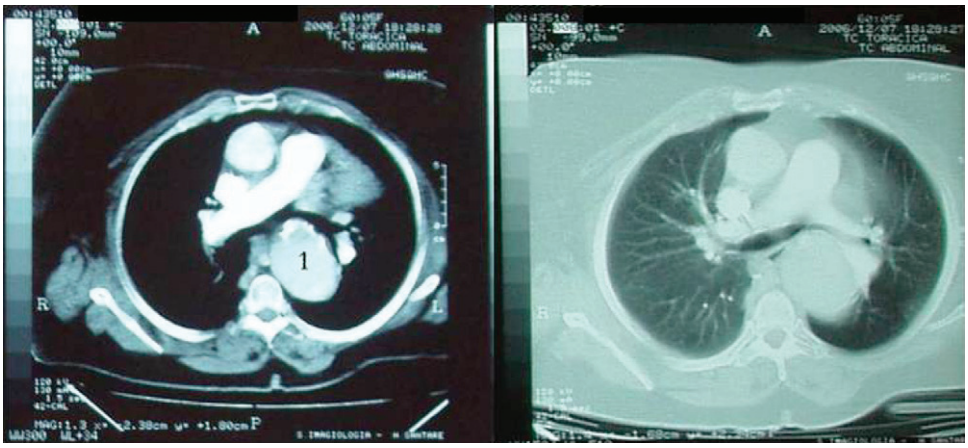

Figure 1 Computed tomography showing an aortic aneurysm (1).