VIA MEDICA

CASE REPORT

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Silent interrupted aortic arch in an elderly patient

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Abstract

Patients with complete interruption of the aortic arch (IAA) very rarely reach late adulthood without having undergone surgical intervention. Only a few cases of IAA in adults have been reported in the medical literature. In this case report, we present a late diagnosis of interrupted aortic arch in a 68 year-old male. Our patient was relatively asymptomatic until he presented with fatigue after walking quickly. A guidewire could not be passed to the aortic arch via the femoral approach; descending thoracic aortography revealed complete occlusion of the descending thoracic aorta. Cardiac catheterization via the right brachial artery confirmed the diagnosis of a complete interruption of the aortic arch distal to the left subclavian artery and showed distinct collateral circulation predominantly via the internal mammary arteries. Also, magnetic resonance angiography showed cuttings that reveal the interruption in the aortic arch and the prominent collateral vessels to the descending aorta. This case report was also interesting in that pressure measurements at a proximal point of the interrupted aortic arch were not hypertensive. Using both catheters, placed proximally and distally to the point of the interruption, by simultaneous pressure measurement, it was measured as 120/75 mm Hg at the proximal point, 60/40 mm Hg at the distal point. (Cardiol J 2011; 18, 6: 695–697)

Key words: congenital anomalies, interrupted aortic arch, elderly patients, vascular imaging

Introduction

Interrupted aortic arch (IAA) is an extremely rare congenital malformation. It occurs in three in every million live births, and accounts for 1% of all congenital heart disease. IAA is associated with additional cardiovascular anatomical defects in up to 98% of cases. Rarely, IAA is an isolated finding without another associated cardiac defect [1]. This anomaly is an extreme form of aortic coarctation, characterized by total luminal and anatomical interruption between the ascending and descending thoracic aorta [2]. Only a few cases of IAA in adults have been reported in the literature [4–9]. Substantial collateral circulation must be presented to maintain blood flow to tissues below the aortic interruption, and thus to enable survival. Arterial hypertension is a typical co-morbidity. Our patient was relatively asymptomatic until he presented with slight fatigue after having walked quickly, and IAA was found incidentally during cardiac catheterization. This case report is also interesting in that pressure measurements at a proximal point of the interrupted aortic arch were not hypertensive.

Case report

A 68 year-old man was referred to our hospital because of slight fatigue after walking quickly. He had been well until the previous 12 months. The patient had no history of hypertension. On physical examination, blood pressure was 125/80 mm Hg in both arms. A grade 2/6 systolic murmur was heard

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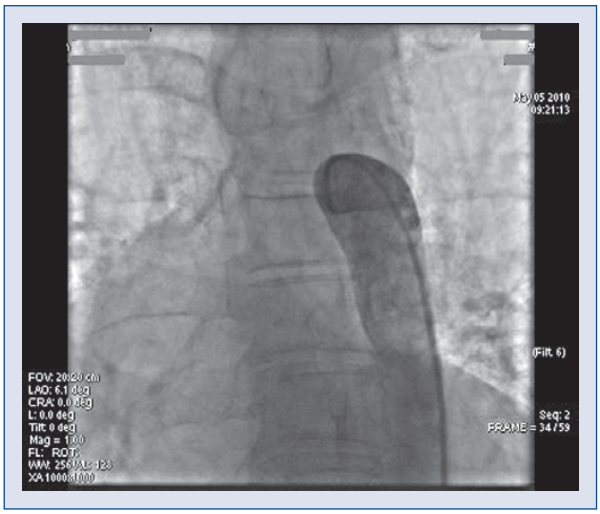


Figure 1. The descending thoracic aortogram shows complete interruption of the aorta in the upper thorax.

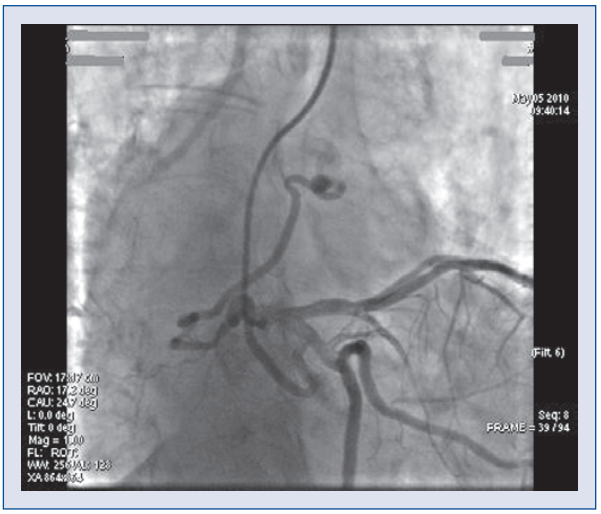


Figure 3. Left coronary angiography showed coronary arteries of atheromatosis without significant stenoses and coronary artery fistula that supplied the arcus aorta.

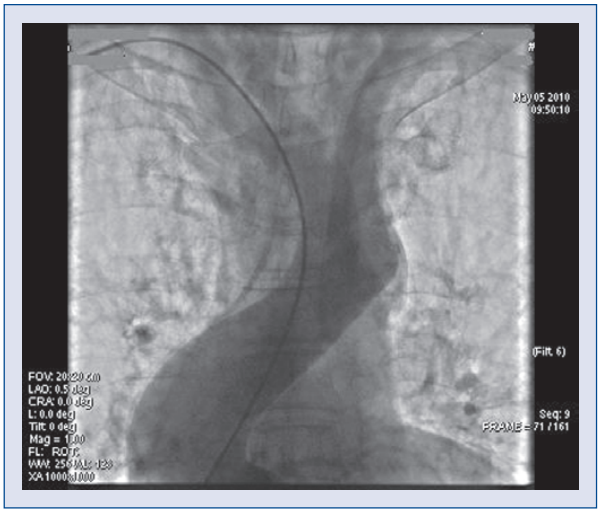


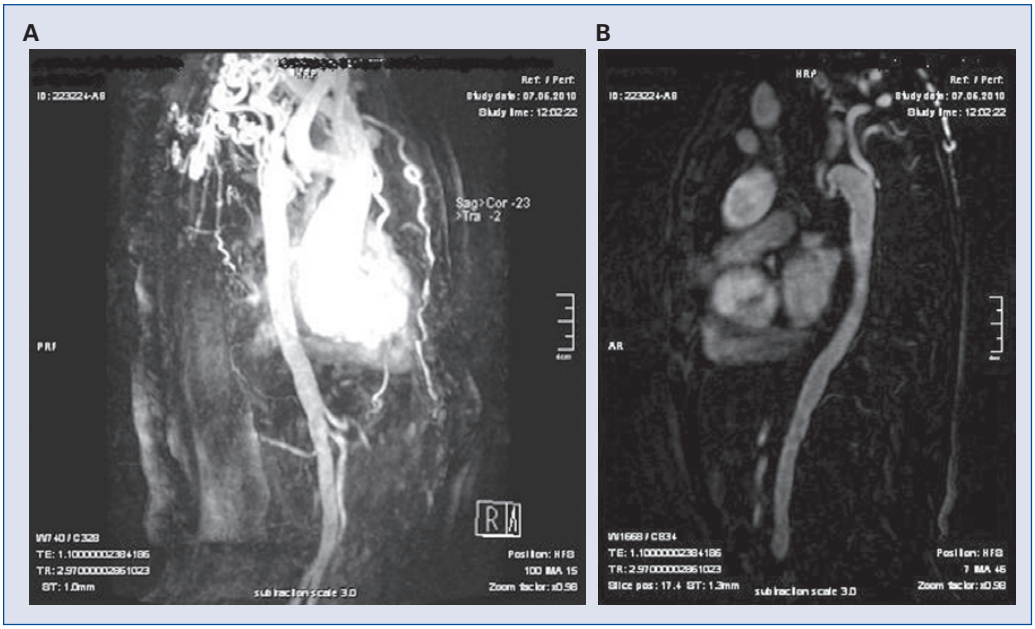
Figure 2. Ascending aortogram shows the aortic arch with complete interruption from the descending thoracic aorta.

in the vicinity of the aorta. Electrocardiography revealed sinus rhythm and left bundle branch block. His echocardiogram showed dilatation of ascending aorta (50 mm), moderate systolic dysfunction of the left ventricle (ejection fraction 45%), moderate aortic and mitral regurgitation and concentric left ventricular hypertrophy. An attempt to perform coronary arteriography failed because a guidewire could not be passed to the aortic arch via the femoral approach; descending thoracic aortography revealed complete occlusion of the descending thoracic aorta (Fig. 1). Cardiac catheterization via the right brachial artery confirmed the diagnosis of a complete interruption of the aortic arch distal to the left subclavian artery and showed distinct collateral circulation, predominantly via the internal mammary arteries (Fig. 2). Coronary angiography showed coronary arteries of atheromatosis without significant stenoses and coronary artery fistula that supplied the arcus aorta (Fig. 3).

Using both catheters, placed proximally and distally to the point of the interruption, by simultaneous pressure measurement, we measured 120/ /75 mm Hg at the proximal point, 60/40 mm Hg at the distal point. The patient underwent magnetic resonance angiography (MRA). Figures 4A and 4B show selected MRA cuttings that reveal the interruption in the aortic arch and the prominent collateral vessels to the descending aorta. While he was being followed up clinically, we performed 24 hour ambulatory blood pressure monitoring which showed a mean pressure of 128/83 mm Hg. The patient was referred to the cardiothoracic surgeon to evaluate his candidacy for surgical or percutaneous therapy, but the patient rejected surgery and conservative therapy with frequent monitoring seemed justified, considering the existence of good blood pressure and the extensive collateral vascularization.

Discussion

Patients with complete interruption of the aortic arch very rarely reach late adulthood without having undergone surgical intervention. When aortic arch interruption in an adult patient is reported, a disease that joins these two factors is under discussion [1–3].



Figures 4A, B. Magnetic resonance angiographic reconstruction shows the interrupted aortic arch in the sagittal plane with extensive well-developed collaterals.

This disease displays the absence of communication between the two segments of thoracic aorta and, consequently, of the blood flow; thus, most cases are expected to be fatal. But sometimes vessels develop extensive collateral networks to maintain hemodynamic stability and this causes an absence of symptoms.

Because of our patient's age, we assumed that the pathogenesis of this interruption of the aortic arch probably was progression and finally occlusion of an aortic coarctation. It was impossible to ascertain whether it was a true congenital atresia, because there was no histological examination.

Acknowledgements

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