Case report - Aortic and aneurysmal

Diagnosis and surgical treatment of an aneurysm on a cervical aortic arch associated with an anomalous origin of the left main coronary artery

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Abstract

Cervical aortic arch (CAA) is a rare congenital anomaly. An aneurysm developed on a CAA is even rarer and a life threatening condition. We report the diagnosis and surgical treatment of an aneurysm on a CAA associated with an anomalous origin of the left main coronary artery. The surgical procedure consisted in the resection of the aneurysm, a direct aorto aortic anastomosis and a coronary artery bypass to the left anterior descending (LAD) artery with a good result at 11 months. This first case reported of an anomaly of a coronary artery origin associated with an aneurysm on a CAA, underlines the interest of a preoperative complete anatomical and functional diagnosis, to define an optimal intraoperative strategy.

Keywords: Congenital; Thoracic aorta; Aneurysm; Anomalous coronary artery

1. Introduction

Cervical aortic arch (CAA) is a rare congenital anomaly first described by Reid [1] in 1914. An aneurysm developed on a CAA is even rarer and a life threatening condition. We report the diagnosis and surgical treatment of an aneurysm on a CAA associated with an anomalous origin of the left main coronary artery.

2. Case report

A 48-year-old female was addressed, complaining about a pulsatile mass on the left side of her neck. The patient’s history started at the age of three years by the diagnosis and the follow-up of a CAA confirmed on an aortography (Fig. 1a), without aneurysmal formation at this time.

On physical examination, there was a left supra clavicular pulsatile mass with a thrill. Blood pressure was 110/70, all peripheral pulses were palpable and there was no pressure gradient. Three-dimensional computed tomography (CT) scan (Fig. 1b) showed a saccular and tortuous aneurysm on the left side of the CAA, with a maximum diameter of 59 mm, just beyond the left common carotid artery, including the origin of the left subclavian artery. Furthermore, there was no brachiocephalic trunk and the right subclavian artery was an arteria lusoria.

A preoperative myocardial scintigraphy showed ischemia in the anterior left ventricular wall. Then, a coronary multi-detector CT-scan (Fig. 2) revealed the presence of an abnormal left main trunk (LMT) arising from the anterior wall of the ascending aorta, immediately below the right coronary artery (RCA). This LMT took an abnormal route around the aorta, giving birth to a circumflex coronary artery and an inter aorto-pulmonary LAD. Based on these findings, surgical intervention was indicated.

Surgery was performed through a median sternotomy with left cervicotomy. A normothermic cardiopulmonary bypass was established between the left common femoral artery and the right atrium. After clamping the descending thoracic aorta and the aortic arch downstream the left common carotid artery, the aneurysm was removed and an aortic end-to-end anastomosis was performed. The left subclavian artery was reimplanted on the aorta with a graft. Then, a coronary artery bypass was achieved on the left anterior descending artery with a venous graft, under complete aortic cross clamping. The postoperative course was uneventful, and 11 months later the patient is asymptomatic without anatomical anomaly (Fig. 1c).

3. Discussion

A CAA is a rare congenital anomaly, complicated with an aneurysmal evolution in about 20% of cases [2]. Its origin is discussed but is probably due to an abnormal persistence of the third branchial arch, while the normal left aortic arch depends on the persistence of the fourth branchial arch [3].

A chromosomal 22q11 micro deletion was recently reported in several cases of aneurysms on CAAs. This 22q11 micro...
deletion is related to the CATCH 22 syndrome described by Wilson et al. in 1993 [4], characterized by congenital conotruncal cardiac anomalies, abnormal facies, thymic hypoplasia, cleft palate and hypocalcemia. Such a micro deletion was not identified in this case.

This pathology concerns in majority young females, often asymptomatic, but patients may present symptoms due to vascular compression of the esophagus or trachea, such as dysphagia, dyspnea, dizziness or frequent pulmonary infections. Our patient was asymptomatic despite this pulsatile left supra clavicular mass.

The ‘gold standard’ exam for diagnosis was the aortic angiography, although nowadays, magnetic resonance angiogram (MRA) or CT-scan have shown their efficiency for the preoperative check-up (for topographical diagnosis or coronary arteries anomalies, as in the reported case) [5]. Angiography can still be indicated and associated to coronarography.

An endovascular approach was not considered here because of the tortuosity of the aorta and the necessity for a coronary artery bypass. Although thoracotomy is frequently used for aneurysm repair, median sternotomy is more suitable for access to aortic arch and supra aortic vessels. The surgical technique consists in the removal of the aneurysmal tissues and, in few cases, an end-to-end aortic anastomosis or, in most cases, an aortic replacement with an artificial graft. In some cases, as ours, circulatory arrest and hypothermia can be avoided if anatomical features allow an aortic clamping, preserving cerebral perfusion by the heart and a lower body perfusion by the cardiopulmonary bypass. In this very rare case of association of an anomaly of the origin and route of the left main coronary artery (unique to our knowledge), we had to complete the procedure by a coronary artery bypass under aortic cross-clamping and cardioplegia. We preferred a venous graft because of the uncertain evolution of the left subclavian artery (reimplanted in the aorta with a prosthetic graft), and because of a potential competitive flow, due to the dynamic compression of LAD different to that of a permanent coronary stenosis. As the patient was asymptomatic even for efforts, long-term patency of the coronary bypass has not been investigated yet.

This first case reported of an anomaly of a coronary artery origin associated with an aneurysm on a CAA, underlines the interest of a preoperative complete anatomical and functional diagnosis, to define an optimal intraoperative strategy.

References