Case report - Vascular thoracic

Repair of the symptomatic aberrant aortic arch aneurysm without hypothermic circulatory arrest

Ramin E. Beygui*, Fardad Esmailian, David A. Rigberg, Hillel Laks

*Department of Cardiothoracic Surgery, UCLA School of Medicine, 10833 Leconte Avenue, Los Angeles, CA 90095-1741, USA
Department of Vascular Surgery, UCLA School of Medicine, 10833 Leconte Avenue, Los Angeles, CA 90095-1741, USA

Received 27 March 2004; received in revised form 14 June 2004; accepted 16 June 2004

Abstract

Manifestation of anomalies of the aortic arch in adulthood has been reported in the literature. The symptoms stem from the compression of the trachea and esophagus or peripheral arterial ischemia associated with coarctation of the aberrant arch. The aberrant arch aneurysms are subject to complications of rupture or dissection. This may be the first reported case of a large complex aneurysm of an anomalous aortic arch resected on cardiopulmonary bypass without any period of hypothermic circulatory arrest or distal ischemia.

Keywords: Aortic arch aneurysm; Aortic arch anomaly; Hypothermic circulatory arrest

1. Introduction

Manifestation of anomalies of the aortic arch in adulthood has been reported in the literature [1]. The symptoms stem from the compression of the trachea, esophagus, recurrent laryngeal nerve or peripheral arterial ischemia associated with coarctation of the aberrant arch. The aberrant arch aneurysms are subject to complications of rupture or dissection. The risk for complications increases with the degree of dilatation [2]. This may be the first reported case of a large complex aneurysm of an anomalous aortic arch resected on cardiopulmonary bypass (CPB) without any period of hypothermic circulatory arrest or distal ischemia.

2. Case report

A 45-year-old woman presented with intermittent symptoms of back pain, dyspnea, and dysphagia. Physical examination revealed decreased femoral pulses. The chest radiograph showed a tortuous thoracic aorta. There was a lobulated shaped mass seen in the right middle mediastinum causing anterior and left lateral deviation of the trachea. A CT angiogram of the chest was obtained (Siemens Sensation 16 multi-slice) with volumetric imaging performed through the lungs and upper abdomen with contiguous axial sections reconstructed at 1.5 mm. This revealed an aberrant aortic arch that extended superiorly and then to the right and then inferiorly (Fig. 1). The aberrant arch then traversed to the left hemithorax posterior to the trachea and the esophagus to connect with the descending thoracic aorta. The arch had dumbbell-shaped aneurysms of 7.5 and 6 cm each in the proximal and mid portion of the transverse arch. The first branch of the ascending aorta was the left common carotid followed by separate origins of the right common carotid and right subclavian arteries. The left subclavian artery arose from the hood of the descending thoracic aorta in the left hemithorax.

After endotrachial intubation in the operating room, fiberoptic bronchoscopy revealed external compression of the trachea without other pathology. The operation was performed through a median sternotomy. A right radial
and a right femoral arterial catheters were placed for monitoring upper and lower body blood pressures. The right radial systolic pressure was 20–30 mmHg higher than the right femoral pressure. The incision was extended slightly over the sternal head of the sternocleidomastoid to control the proximal right common carotid, right subclavian, and the downward turn of the aberrant arch. The right vagus recurring around the aberrant arch was identified and protected. The main and branch pulmonary arteries were dissected and mobilized exposing the left posterior pericardium. By entering the left posterior pericardium inferior to the left pulmonary artery, trachea, esophagus, and the aberrant transverse arch were exposed. The junction of the transverse arch with the descending thoracic aorta in the left hemithorax was dissected and controlled. The last brachiocephalic branch, the left subclavian, was controlled through the left posterior pericardium. After administration of systemic heparin and cannulation of the ascending aorta, superior and inferior vena cava and placement of antegrade cardioplegia catheter, CPB was initiated. The patient was initially cooled reaching a nasopharyngeal temperature of 28 °C. The aberrant transverse aortic arch was isolated between cross clamps placed distal to the right subclavian in superior right anterior mediastinum and proximal to the descending thoracic aorta in the left inferior posterior mediastinum. Immediately, the descending thoracic aorta was cannulated using a number 6 endotrachial connected the CPB circuit. The cannulation of the descending thoracic aorta using the cuffed endotrachial tube became feasible after transection of the aorta beyond the aberrant arch. Thus, the left common carotid, right common carotid, and right subclavian were perfused through the ascending aortic cannula. Concurrently, the left subclavian and descending thoracic aorta were perfused using the cuffed endotrachial tube in an antegrade fashion. The right radial and right femoral arterial pressures were carefully monitored to ensure adequacy of perfusion of the upper and lower body. The transverse arch was then reconstructed anteriorly using a 26-mm coated polyester (Hemashield Gold). A single dose of antegrade cold blood cardioplegia was given during completion of the proximal anastomosis between the ascending aorta and the graft. The systemic re-warming was completed and the patient was separated from CPB without any inotropic support. The entire dumbbell shaped aneurysm was then excised relieving the posterior compression of the trachea and the esophagus. Upon completion of the operation the right radial and femoral arterial pressures were identical. Fiberoptic bronchoscopy was performed in the operating room that showed resolution of tracheal compression without any other pathology. The patient was extubated the same day in the intensive care unit and was discharged home on the 6th post-operative day. Post-operative CT-angiogram indicated complete excision of the dumbbell-shaped aneurysm (Fig. 2). Anteriorly reconstruction of arch, from ascending aorta to descending thoracic aorta, caused no compression of the main or left pulmonary arteries.

3. Discussion

Manifestation of anomalies of the aortic arch in adulthood has been reported in the literature [1]. The symptoms stem from the compression of the trachea, esophagus, re-current laryngeal nerve, or peripheral arterial ischemia associated with coarctation of the aberrant arch. The aberrant arch aneurysms are subject to complications of rupture or dissection. The risk for complications increases with the degree of dilatation [2].
In recent literature, a case of anomalous left aortic arch associated with aneurysm and coarctation of the distal arch in an infant [3] has been reported. The arch had been bypassed without CPB through a median sternotomy. Also, extra-anatomic bypass of the right aortic arch [4–6] for coarctation has recently been reported. The right-sided aortic arch is mostly asymptomatic and found incidentally in the adult, unless aneurysms develop. This usually occurs at the level of the take-off of an aberrant left subclavian artery and is known as a Kommerell’s diverticulum [7]. However, this is the first reported case of an anomalous aortic arch associated with a 7.5 and 6 cm complex aneurysms of the aberrant transverse arch taking a circuitous path in the retrotracheal space before traversing to the left hemithorax. The aneurysms needed to be excluded for their size to prevent rupture or dissection. The symptoms emanated from the compression of trachea and esophagus, thus requiring complete resection. The reconstruction of the arch anteriorly and complete resection of the posterior dumbbell-shaped aneurysm was accomplished through a median sternotomy. The conduct of CPB without any period of hypothermic circulatory arrest may prove to be a useful technique in many cases of aortic arch surgery. Double cannulation of the ascending aorta and the innominate artery for arch reconstruction in an infant was recently reported [8]. In that case the cannulation technique afforded regional perfusion of the brachiocephalic vessels. However, the perfusion to the descending thoracic aorta was interrupted during arch reconstruction. The antegrade cannulation of the descending thoracic aorta using a soft cuffed endotracheal tube, provided perfusion of the lower body concurrently with perfusion of the brachiocephalic vessels. The relative perfusion of the brachiocephalic vessels and descending thoracic aorta was easily monitored and adjusted by placement of a radial and a femoral arterial catheters. The continuous perfusion of the descending thoracic aorta may reduce the risk of spinal cord, mesenteric, and renal ischemia that is associated with hypothermic circulatory arrest. Avoiding hypothermic circulatory arrest also obviates the need for prolonged pump run for re-warming and potential prolonged intubation due to temporary neurologic dysfunction, and transfusion due to excessive coagulopathy [9,10]. A right thoracotomy approach with concurrent cannulation of the ascending aorta and femoral artery would have accomplished total body perfusion. However, the anatomic location of aneurysm in the retrotracheal space with the distal neck of the aneurysm in the left hemithorax did not afford itself to complete resection through a right thoracotomy. Finally, high-resolution CT-angiogram can clearly define the anatomy of the anomalous aortic arch and its branches [11]. The three-dimensional reconstruction of the images enhanced the anatomy of this unusual arch aneurysm such that the surgical approach and exposure was planned in advance and conducted without unexpected occurrence, obviating the need for contrast angiography.

**References**


