Aortic arch aneurysm with aorto-pulmonary fistula: successful surgical repair

Abstract
Aortic arch aneurysm associated with an acute aorto-pulmonary fistula is rare and usually diagnosis is made during the postmortem. Few reports of successful surgical management have been reported and a high mortality rate remains. The authors report on a 78-year-old male patient with an acute aortic arch aneurysm communicating with the main pulmonary artery suffering from cardiac chest pain, hemodynamic instability and lung congestion. The patient underwent surgery to replace the distal ascending aorta, transverse aortic arch and proximal descending aorta as well as fistula closure. The operation was successful.


Resumo
Aneurisma de arco aórtico com fistula aorto-pulmonar aguda é uma afecção rara e, geralmente, diagnosticada postmortem. Poucos relatos de sucesso cirúrgico foram realizados e persiste uma alta taxa de mortalidade. Os autores relatam o caso de um paciente de 78 anos, com comunicação aguda entre aneurisma de arco aórtico e artéria pulmonar com precordialgia, instabilidade hemodinâmica e congestão pulmonar. A operação foi realizada com sucesso, sendo realizada a substituição da porção proximal da aorta ascendente, do arco aórtico e da porção distal da aorta ascendente, além do fechamento da fistula.

INTRODUCTION

Acquired aortopulmonary fistulae are rare [1-6] and the most frequent etiology is trauma. Fistulae between aortic arch aneurysms and the pulmonary arteries are generally only diagnosed in autopsies [1] and have been reported since 1812.

Rupture of thoracic aorta aneurysms generally result in death due to cardiac tamponing or hemorrhagic shock [1-3] and, in the few cases in which the patient survives, an aortopulmonary fistula can form leading to severe pulmonary hypertension and cardiac insufficiency. Successful surgical repair is rare with the first reported case performed by Giacobini and Cooley, in 1960 [2]. Large and English [3] published surgical management and results in 1988.

The case of a patient suffering from an aortic arch aneurysm with complications due to an aortopulmonary fistula, which was successfully treated by surgery, is reported.

CASE REPORT

A 78-year-old male patient presented with thoracic pain and epigastralgia together with an episode of hypertension in November 2002. Laboratorial examinations discarded acute myocardial infarction (AMI), but a radiological examination showed enlargement of the mediastinum (Figure 1).

After controlling blood pressure there was an improvement in his clinical state. The patient underwent a cinecoronariography which showed dual coronary artery lesions and after stabilization a computed chest tomography revealed an aortic arch aneurysm with a size of approximately 80 mm at its greatest diameter confirmed by magnetic resonance (Figure 2). Surgery was indicated, but the patient refused. In March 2003, the patient was readmitted to hospital with precordialgia and hemodynamic instability and again the diagnosis of acute myocardial infarction was discarded by laboratorial examinations. The physical examination demonstrated a continuous audible murmur in the entire precordial region and evidence of pulmonary congestion. Again, the patient was submitted to a magnetic angioresonance which showed the presence of a shunt between the aortic aneurysm and the pulmonary artery of roughly 7 mm in diameter and continuous aortopulmonary flow, in addition to signs of severe arterial hypertension. An echocardiogram demonstrated mitral insufficiency and an estimated pulmonary artery pressure of 90 mmHg.

The patient underwent percutaneous revascularization of the anterior interventricular artery and a Penta-type stent was implanted and he was referred to the surgical center for correction of the aortic disorder.

The operation was performed by means of median sternotomy using cardiopulmonary bypass via the right femoral artery, circulatory arrest and deep hypothermia of 20 °C. Brain protection was performed using retrograde perfusion via the superior vena cava with a flow rate of 300 mL/min and a pressure of 25 mmHg, as well as topical hypothermia and administration of endovenous Thionembutal. Myocardial protection was achieved by anterograde hypothermic sanguineous cardioplegic solution at 20-minute intervals. Significant dilatation and an intense atherosclerosis of the aorta were evidenced with many
thrombi, which were removed. The 10-mm orifice located in the inferior portion of the aortic arch that was connected to the pulmonary artery was occluded using 4-0 polypropylene thread. Substitution of the proximal portion of the descending aorta, the aortic arch and the distal portion of the ascending aorta was performed using Dacron nº 30 tubes with re-implantation of the cerebral vessels in a single orifice, suturing from the distal part to the proximal portion of the vessels.

The patient evolved with hemodynamic instability and ischemia of the left arm in the immediate postoperative period. He showed mental confusion and bronchopneumonia in addition to acute myocardial infarction in the anterior wall on the 9th postoperative day, which were satisfactorily treated. The patient was released from the intensive care unit on the 16th postoperative day, with good evolution in the cardiological and neurological states. He was well until one year after the operation, when he died of non-cardiac causes.

DISCUSSION

Aortopulmonary fistulae resulting from chronic aneurysms of the aorta are very rare and generally originate from rupture or dissection of the aorta aneurysm. They are most common with a diameter of greater than 60 mm and are frequently associated with systemic arterial hypertension [1,3,4].

An investigation of 4000 autopsies involving thoracic aorta aneurysms revealed that only 3.7% ruptured to the pulmonary artery [6]. One hundred and eight live cases were diagnosed and described in the international literature up to 1992 and, of these, only ten patients were successfully submitted to surgery [1,3]. Most of these patients were admitted to hospital with signs of cardiac insufficiency and severe pulmonary hypertension [3].


The treatment of these patients must include a previous diagnosis by laboratorial examinations and immediate clinical care with control of blood pressure and the reduction of right-left shunt using vasodilatory drugs, principally those that act directly on the pulmonary system [3]. Surgical treatment should be performed immediately.

The risk factors associated with early death after surgical repair are: old age, congestive cardiac insufficiency, angina, dissection, aortic arch management and co-morbidities [3]. The current case is an old patient with typical anginal pain without any dissection but with several co-morbidities, including: chronic obstructive pulmonary disease, systemic arterial hypertension, pneumonia, an increase of nitrogenous compounds in the preoperative period, peripheral vascular insufficiency in addition to coronary artery insufficiency. The patient only accepted the operation after the appearance of the fistula and severe cardiac insufficiency.

In the repair of aortic arch aneurysms, cerebral protection is very important, which is still controversial but seems to be a determining factor of surgical success, as well as the correct management of pulmonary complications, related to the aortopulmonary fistula [2]. In the present case, protection was established with topical cooling and deep systemic hypothermia (20ºC), cerebral retrograde perfusion through the superior vena cava, as well as other aforementioned measures. Nowadays our department prefers the anterograde cerebral perfusion associated with deep hypothermia as opposed to cerebral retrograde perfusion.

Numerous improvements to cardiopulmonary bypass, myocardial preservation, brain protection, anesthesia and surgical graft techniques have been associated with increases in the survival rate. However, despite of these advances, aortopulmonary fistulae present as difficult complications, related to the aortopulmonary fistula [2]. The postoperative treatment of these patients must be managed by an experienced multidisciplinary team.

Finally, thoracic aorta aneurysms associated with fistulae to the pulmonary artery require immediate care with cautious surgical preparation and in spite of being a condition that causes much morbidity-mortality, satisfactory surgical results can be obtained as demonstrated in this case.
BIBLIOGRAPHIC REFERENCES


