Spontaneous aortic arch rupture with pseudoaneurysm and constrictive-effusive pericarditis formation

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Abstract

Spontaneous aortic rupture, without any history of previous thoracic trauma, infection or acute thoracic pain is an extremely rare and potentially life-threatening event. Its diagnosis, in the absence of acute symptoms, is usually delayed and relies on secondary signs. While the etiology is atherosclerotic in most cases, the exact mechanisms of rupture have only recently been uncovered. Treatment may be surgical, endovascular or by a combined procedure, according to the anatomy, location and expertise of the medical team.

Keywords: Aortic arch; Rupture; Pericardium; Aneurysm

1. Introduction

Atraumatic, spontaneous aortic rupture is a rare but potentially lethal event. In the following report, we present a case of spontaneous aortic arch false aneurysm with an associated effusive-constrictive pericarditis that was treated surgically in our department. We discuss the etiology of this entity, the pathological processes involved and optimal treatment for these critically ill patients.

2. Clinical summary

A 53-year-old male patient was admitted to our department complaining of progressive breathlessness, fatigue, and dysphonia. His medical history was unremarkable except for hypertension and tobacco consumption. In particular, there was no recollection of recent thoracic trauma, infection or acute thoracic pain. The physical examination revealed bilateral, venous type, peripheral edema and a swollen left upper limb. The cardiovascular examination found diminished cardiac sounds but no murmurs. The chest X-ray exam found a left pleural effusion, normal heart size and a dilated upper mediastinum. The chest X-ray exam found a left pleural effusion, normal heart size and a dilated upper mediastinum. The chest X-ray exam found a left pleural effusion, normal heart size and a dilated upper mediastinum.

The patient was operated upon shortly thereafter. Peripheral femoral artery canulation was established prior to entering the chest, due to the intimate relationship between the aortic dilation and upper sternum. After midline sternotomy, we found an anterior aortic arch false aneurysm developed in the upper mediastinum and in close contact with the left pleura. The pericardium was fibrosed, thickened and contained a moderate amount of hemorrhagic fluid and organized clots (Fig. 1a). Venous canulation was performed via a double stage venous canula. Moderate hypothermia to 25 °C was used with cold cardiopulmonary arrest. At this temperature cardiopulmonary bypass (CPB) was stopped and the pseudoaneurysm opened. Antegrade cerebral perfusion (10 ml/kg min) was established by placing two coronary sinus canulas in the innominate and left common carotid arteries and delivered via a sideline of the cardiopulmonary circuit, switching back and forth between cold cerebral perfusion and cardioplegia every 15 min. Flow into the cardiopulmonary circuit consisted of cold blood and St. Thomas Hospital cardioplegic solution in a 4:1 ratio for 1 min every 20 min, when delivering cardioplegia and cold blood between doses, delivering antegrade cerebral perfusion. After excision of the defect margins into healthy tissue a Dacron, felt-reinforced patch repair was undertaken, using two 4-0 polypropylene stitches sutured in a continuous fashion (Fig. 1b). CPB was resumed after deairing maneuvers and warming was begun. Hypothermic arrest was 39 min and crossclamp time 46 min. CPB was gradually and uninterruptedly stopped under moderate inotropic support (Epinephrine 0.03 μg/kg min). The patient was extubated the following day and had, subsequently, an uneventful recovery. He is to date alive and well, six months after surgery. Histology of the resected aortic arch specimen revealed extensive atherosclerosis, with calcified subintimal plaques and a complete disruption of the aortic wall.

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Fig. 1. Contrast-enhanced CT scanning shows a 7-mm in size, anterior arch aneurysm with marked atherosclerosis of the aorta (a) and a constrictive-effusive pericarditis (b).

3. Comment

Since the first described spontaneous, non-traumatic aortic rupture case by Rodriguez and Rivera in 1961 [1], only 31 additional cases of this rare pathology have been reported [2]. This term is applied if the rupture is not due to thoracic trauma or any other previously identified aortic condition. The development of modern-era imaging studies has identified penetrating atherosclerotic aortic ulcer (PAU) as a possible cause of aortic rupture in an otherwise asymptomatic patient. PAU appears in patients with extensive atherosclerotic aortic lesions. We suspect, based upon histology, this mechanism to have acted in our patient to provoke the spontaneous rupture of the anterior wall of his aortic arch, with left recurrent laryngeal nerve compression. Another interesting aspect has been the presence of a chronic, markedly thickened pericardium with a moderate hemorrhagic effusion, acting as a constrictive-effusive pericarditis. It is, to the best of our knowledge, the first reported association of these pathologies.

While the atherosclerotic lesions are most likely responsible for spontaneous atraumatic aortic rupture, the exact mechanisms of rupture have only recently been identified. Matrix metalloproteinases (MMP) have been implicated in the pathogenesis of degenerative aortic diseases. The concentration of MMP-9, a gelatinase belonging to the MMP family, in the arterial wall correlates positively with aneurysm size [3] and its expression by macrophages induces acute atherosclerotic plaque disruption [4]. Furthermore, its concentration, as well as that of MMP-8 is increased at the site of abdominal aortic aneurysm rupture [5]. Finally, deficiency in the tissue inhibitor of metalloproteinases type 1 (TIMP-1) results in spontaneous pseudoaneurysm formation in an experimental model [6]. This family of enzymes seems to act as the final effector in degenerative arterial lesions.

Aortic arch surgery has been drastically modified recently by the advent of interventional techniques. Coil embolization has been described in pseudoaneurysms with a suitable anatomy [7], however, most aortic arch aneurysms benefit nowadays from a combined surgical-endovascular repair procedure. Rerouting of the aortic arch branches is done surgically in a first step, which allows for a subsequent covered-stent graft placement in the aortic arch [8]. This treatment option significantly reduces the morbidity associated with hypothermic arrest, however, it was not available in our case due to economic constraints.

While remaining rare, spontaneous aortic rupture is diagnosed with increasing frequency and should be taken into account in the differential diagnosis of acute thoracic syndromes. Treatment may be surgical, endovascular or by a combined two-step procedure, according to the type of the offending lesion, its localization, availability and team-experience.
References