Type A aortic dissection complicated with fistulization into the right atrium and right-to-left shunt

Sebastian Pagni, Christopher Mascio, Jaimin Trivedi and Jiapeng Huang

INTRODUCTION

Aorto-right atrial fistulization (ARAF) is a rare occurrence after proximal aortic dissection [1, 2]. The presentation is dramatic, often with cardiogenic shock. Transoesophageal echocardiography (TEE) is the most suitable diagnostic tool in the unstable patient. Emergent surgical intervention is required before progression to end-organ failure.

A 69-year-old white female presented with acute severe, persistent mid-sternal chest pain, shortness of breath, tachycardia (heart rate of 110 bpm) and cardiogenic shock. She had a mid-right sternal border systolic murmur and faint femoral pulses. The troponin level was 10 µg/dl. The electrocardiogram (EKG) showed an inferior and posterior T-wave inversion and Q-waves in the inferior leads. She had a myocardial infarction in 2005 (followed by coronary artery bypass surgery), baseline creatinine 1.8 mg/dl, atrial fibrillation, hypertension and breast cancer (left radical mastectomy, 1990). A bedside TEE confirmed the diagnosis of type A aortic dissection (AAD) complicated by a periarticular false aneurysm. It also showed severe right atrium and right ventricle enlargement and a fistula between the false aneurysm and right atrium (Figs 1 and 2). Right-to-left shunting was seen through a patent foramen ovale.

The patient was explored emergently. After heparinization, cardiopulmonary bypass (CPB) was established cannulating the right axillary artery and right common femoral vein. A repeat sternotomy and mediastinal dissection were performed. Left ventricular venting and coronary sinus catheters were used. The graft to the left anterior descending artery was identified and flow interrupted. The aorta was clamped below the innominate artery, and myocardial protection was established by antegrade and retrograde coronary perfusion. During systemic cooling, we dissected the proximal aortic area (Fig. 2). There was a large false aneurysm (5 × 7 cm) communicating with the ruptured aortic false lumen and a fistula to the right atrium. There was a fresh clot in the false lumen. The dissected layers of the sinus wall and root were reattached using BioGlue (Cryolife, Inc., Kennesaw, GA, USA), and the commissures resuspended using 4.0 monofilament sutures. Hypothermic circulatory arrest was instituted at a core temperature of 15°C with electroencephalogram (EEG) silence, and the antegrade cerebral perfusion monitored by cerebral oxymetry and trans-cranial dopplers.

The ascending aorta was resected including the tear, and the proximal arch cuff was prepared by using BioGlue as neo-media and thea velved # 28 Dacron graft (Gelwave, Terumo, Inc.) was sutured. The right atrium was explored and the large patent foramen ovale was closed primarily before rewarming and systemic CPB resumed. A vein graft was anastomosed to the posterior descending coronary artery due to closure of the vein graft. The Dacron tube graft was anastomosed to the proximal aortic cuff to complete the repair, and the proximal vein anastomosis completed before aortic unclamping. The hypothermic circulatory arrest, CPB and myocardial arrest times were 25, 231 and 122 min, respectively.

The postoperative course was complicated by acute renal insufficiency and respiratory distress due to laryngeal oedema, requiring mechanical ventilation for 7 days. She was discharged home on postoperative day 25. She remained alive 23 months after surgery without complications related to the dissection repair surgery.

COMMENT

Rupture of the aorta into the right atrium is a rare, but often catastrophic, complication of AAD [2]. Without early recognition,
it can be fatal like most AAD, but with the added risk of acute congestive heart failure, shock and rapid progression to end-organ failure [3]. ARAF has been described as a post-operative complication after the repair of AAD or in patients with AAD and previous cardiac surgery. We speculate that the adhesions from previous cardiac surgery contain the aortic rupture, which eventually develops into an false aneurysm and then ruptures into a right heart chamber. Heart failure with circulatory collapse is often the presentation of ARAF [4].

The computed tomography angiography is the gold standard for diagnosis; however, such as shown in our patient, the haemodynamic instability precluded its use. Bedside and/or intraoperative TEE is invaluable in rapidly diagnosing AAD with ARAF and in determining the tear site, aortic valve function and presence of an false aneurysm.

Kuipers et al. [3] first reported a case of ARAF post-dissection at autopsy in 1963. In 1966, Temple et al. [2] reported the first successful repair of ARAF in a patient with chronic proximal aortic dissection with fistulization and acute right heart failure. Few reports have followed, with mixed results, all sharing the common finding of a difficult initial diagnosis, right heart failure and often previous cardiac surgery. Obvious reasons for the sub-optimal outcomes are the need for emergent repair of two major problems in the setting of a reoperation requiring hypothermic circulatory arrest and complex aortic reconstruction. Few cases of ARAF with acute dissection have been reported [1–5] with high perioperative mortality (30–50%). Only 2 cases of ARAF in the setting of a previous CABG have been described and, including ours, all have survived, indicating better outcomes than ARAF with prior aortic surgery.

We speculate that the right ventricle dysfunction was related to the acute ischaemia in the right coronary territory due to acute closure of the saphenous vein graft to the right coronary artery. The finding of a right-to-left shunt was very distinct, as shown in Fig. 2.

To our knowledge, this report is the first to describe the finding of a severe right-to-left shunt in the setting of an ARAF and right heart failure. Although the significance of the shunt is unclear, it might have helped the patient by venting into the left atrium through patent foramen ovale, thus avoiding rapid circulatory collapse.

In conclusion, ARAF is a very rare and fatal complication of AAD. The diagnosis should be suspected when an atypical murmur and right heart failure are present in the setting of an acute aortic syndrome with shock. TEE is an extremely useful diagnostic tool at the bedside, and prompt surgical intervention is life-saving.

Conflict of interest: none declared.
REFERENCES


eComment. Aorto-right atrial fistula in type A aortic dissection

Author: Jamil Hajj-Chahine

Department of Cardio-Thoracic Surgery, University Hospital of Poitiers, Poitiers, France
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I read with great interest the paper by Pagni et al. concerning the management of a 59-year-old female patient with acute type A aortic dissection complicated by a fistula to the right atrium [1]. This article has two interesting features that could be of use to all of us when faced with a similar case.

The optimal surgical approach to patients who present with acute type A aortic dissection in the setting of previous cardiac surgery is not always apparent. Adhesions from previous cardiac surgery have been proposed to prevent free rupture of the ascending aorta, this lowered risk of rupture and tamponade has led some to suggest delaying the operation with the intention of obtaining cardiac catheterization, especially in the setting of prior coronary artery bypass grafting surgery. Although extensive adhesions might protect against free rupture, there is another risk — fistulization or contained rupture of the aorta into neighbouring cardiac chambers and pulmonary artery. In this case scenario, surgical intervention is emergently required to prevent dismal prognosis secondary to preoperative cardiogenic shock.

In a recently published study, Kledell et al. [2] showed that patients with acute type A aortic dissection with a history of cardiac surgery are at increased risk for aortic rupture when compared to aortic dissection without previous cardiac surgery (29% vs 3.2%, P=0.012). However, the rupture is contained in the majority of cases (67%) without tamponade or cardiogenic shock. Only two patients presented with aorto-atrial fistula and cardiogenic shock. These patients died despite aortic repair. Estera et al. [3] operated on two patients with aorto-pulmonary fistulas in the setting of aortic dissection in redo cases. Unfortunately, these two patients also died after the operation.

Reports from the literature are sparse and sporadic [4]: rupture and haemodynamic instability are rare because of postcardiotomy scarring and protective periarticular fibrosis [5]. Aortic fistula to the right heart cavities is a dreaded complication that poisons any increased risk of mortality and dismal postoperative outcomes. As outlined in this case report, successful management requires timely recognition along with prompt surgical intervention.

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References