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Pulmonary artery dissection in a patient with undiagnosed pulmonary hypertension – A case report and review of literature

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ABSTRACT

Pulmonary artery dissection is rare and highly lethal and most reports in the literature are from autopsies. We describe a patient with undiagnosed primary pulmonary hypertension suffering from pulmonary artery dissection that subsequently underwent surgical repair and in addition review the current literature on this topic.

Introduction

Pulmonary artery dissection is extremely rare and has been reported in less than 100 patients, most of them diagnosed post-mortem, indicating that these patients succumb to cardiogenic shock or sudden death.[1] Of the known cases, 24 were diagnosed alive. Successful repair of the pulmonary artery has been described in only seven cases. We report the outcome after surgical resection of the central pulmonary artery (PA) and reconstruction with grafts in a patient suffering from aneurysm and dissection of the pulmonary artery.

Case report

A 55-year old man with a previous history of malignant melanoma and one episode of metastasis to the lung, allegedly in remission for 20 years, was admitted due to progressive exertional dyspnea. He was diagnosed with a pulmonary artery aneurysm and dissection by CT-scan (Fig. 1a–c) and referred for surgical intervention. The CT-scan did not reveal any pericardial effusion. Preoperative ultrasound demonstrated a hypertrophic right ventricle with a peak systolic pulmonary artery pressure of 80 mm Hg. After thorough evaluation and discussion with the patient it was decided to repair the dissected pulmonary artery.

The patient was premedicated orally with oxazepam (5 mg). Before induction of anesthesia, an arterial cannula was placed in the left radial artery and a triple lumen (Arrow International, Inc., 2400 Bernville Road, Reading, PA 19605, USA) catheter was inserted into the left subclavian vein. A norepinephrine infusion was started to maintain mean arterial pressure (MAP) >70 mm Hg in order to sustain coronary blood flow and maintain interventricular dependence. By augmenting aortic root pressure in the setting of increased right ventricular (RV) afterload, RV ischemia was prevented.[2]

After 5 min of preoxygenation with F_IO_2 100% and 20 ppm NO, anesthesia was induced with ketamine (1.2 mg/kg), propofol (1 mg/kg) and fentanyl (5 μg/kg). Tracheal intubation was facilitated with rocuronium (0.8 mg/kg). Anesthesia was maintained with sevoflurane in oxygen and 20 ppm inspired NO with a F_IO_2 necessary to keep P_aO_2 >15 kPa. Ventilation was volume-controlled to maintain P_aCO_2 between 5.0 and 5.5 kPa. A rapid infusion (AVA HF, Edwards Lifesciences, LLC, Irvine, CA 92614-5686 USA) catheter was inserted through the right internal jugular vein and a continuous cardiac output thermo dilution flow directed pulmonary artery (Edwards Lifesciences) catheter (PAC) was positioned in the superior caval

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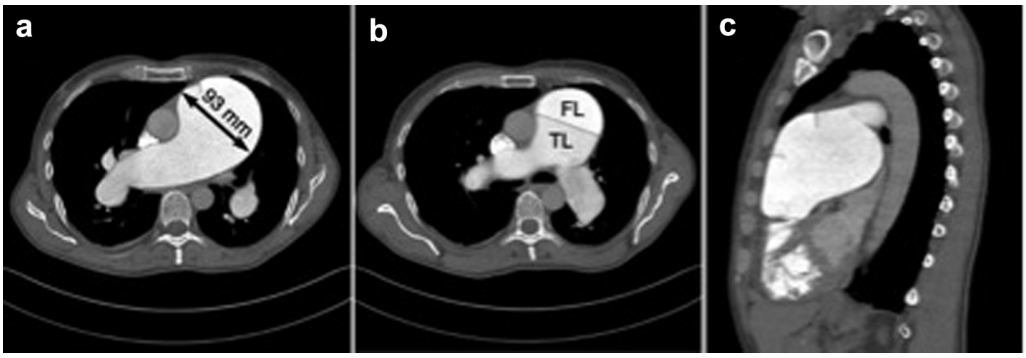

Fig. 1. (a) Preoperative CT-scan demonstrating an aneurysm of the pulmonary artery (PA). In the ventral medial portion of the aneurysm is a small flap of the dissection membrane seen. (b) Preoperative CT-scan demonstrating a dissection of the pulmonary artery (PA). False (FL) and true lumens (TL) are seen. (c) Sagittal view showing the pulmonary valve and the proximal and distal dissection membranes in the ventral wall of the aneurysm of the pulmonary artery.

vein. A 6Tc TEE probe (Vivid E9, GE Healthcare) was used together with a Vivid E9 cardiovascular ultrasound system (Vivid E9 GE Healthcare, 9900 Innovation Drive, Wauwatosa, WI 53226, USA) to monitor cardiac function. Near infrared spectroscopy (INVOS, Somanetics Corporation, Troy, Michigan 48084, USA) with sensors placed on forehead was used during the entire procedure.

Due to the risk of rupture of the aneurysm, the right femoral artery and vein were cannulated after heparinization and extracorporeal bypass circulation (ECC) was initiated. Then, standard sternotomy and pericardiotomy was safely performed. A view of the operative field is shown in Fig. 2a. During and after ECC, a continuous infusion of propofol was administered as well as incremental doses of fentanyl. The superior vena cava (SVC) was dissected free. Without clamping of the aorta, the main pulmonary trunk was incised down towards the left PA and the dissecting membrane was visualized (Fig. 2b). Another incision of the right PA was performed between the aorta and SVC, and extended into the right hilum passing the right upper lobe artery. A 30 mm Dacron graft was sutured with a 5-0 Prolene to the right PA and subsequently tunneled toward the left side in the remaining native pulmonary artery and sutured to the left PA with 5-0 Prolene, as described by Senbaklavaci et al.[3] A 3 cm incision was performed in the middle of the interconnected graft and another 30 mm graft was sutured with 5-0 Prolene in a T-shaped fashion. Finally the proximal end of this graft was sutured to the sino-tubular junction of the pulmonary valve (Fig. 2c). The PAC was now floated into the pulmonary artery. Coming off bypass resulted in excessive diffuse bleeding due to pulmonary hypertension with systolic PA pressures around 100 mm Hg. Bypass was restarted and the pulmonary aneurysmal sac was closed around the graft. Before another attempt of weaning from bypass, the following treatment was instituted: inhalation of 20 ppm NO and 10.000 ng/mL, 10 mL/h prostaglandin I_2 (PGI_2, epoprostenol, Flolan®) in 100% oxygen was started (due to the synergistic vasodilator effect on pulmonary vascular resistance),[1,2,4–8] as soon as the ventilator was turned back on. The external pacemaker was set at DDD 90/min. A bolus dose of milrinone 50 μg/kg was given and a milrinone infusion of 0.375 μg/kg/min was started. Norepinephrine was infused targeting a MAP of 80 mm Hg. Pulmonary pressure remained high and almost equal to systemic pressure. CVP was targeted at 15–18 mm Hg. This second attempt was successful and followed by thorough hemostasis. The heparin effect was reversed with protamine sulfate until normal ACT values were achieved. Plasma, cryoprecipitate and platelets were given according to the ThromboElastoGram (TEG5000, Haemonetics Corporation, 400 Wood Road, Braintree, MA 02184, USA). Hemoglobin was 17 g/dL at the start of the operation and was kept above 10 g/dL by infusion of erythrocyte concentrates. At the end of surgery, the NO could gradually be weaned. The wound was packed with gauze and without closing the sternum the patient was taken to the intensive care unit (ICU) with on-going infusion of milrinone, norepinephrine and propofol as well as aerosolized PGI_2.

Almost equalized pressures in the pulmonary and systemic circulation characterized the first couple of the days in ICU, despite the use of aerosolized PGI_2. Two days later the patient was taken back for reoperation. Moderate amounts of blood clots were rinsed from the wound. The patient was edematous exhibiting hemorrhagic diathesis. Again, the wound was packed with gauze and the patient was taken to the ICU.

At the fourth postoperative day PGI_2 was discontinued and the sternum was closed. During closure the patient developed atrial fibrillation, which did not respond to electro conversions. During the evening the patient became increasingly unstable and required high doses of inotropic support (using Milrinone, Norepinephrine and Epinephrine). Despite treatment with Amiodarone, Metoprolol, Adenosine and numerous electro conversions, the atrial arrhythmias persisted. Veno-arterial extracorporeal membrane oxygenator (ECMO) was considered but refrained because of the poor prognosis. The sternum was re-opened in the ICU, but with no improvement in cardiac output. The patient died later that evening in cardiogenic shock.

Discussion

Dissection of the pulmonary artery is a rare condition with less than 100 reported cases in the literature. The vast majority of these are found in autopsies. The most common cause is congenital heart disease (most commonly persistent ductus arteriosus) but other causes include primary or secondary pulmonary hypertension, vascular inflammatory disease, aorto-pulmonary fistulas, connective tissue disease and catheter induced vessel wall injury. We have identified twenty-four cases where the patients were alive at the time of diagnosis (Table 1) in the literature. No clear consensus regarding how these patients are best managed exists. In fourteen cases the clinical outcome was reported as stable (follow-up 4 days–6 years), in four cases the patients died shortly after diagnosis, one patient survived 12 months and in five cases the outcome was not reported. The reports of surviving patients demonstrate the feasibility of both conservative medical management (seven cases) and surgical repair (seven cases). In one case of surgical repair, bilateral lung transplantation was performed after 10 months,[3] and in two cases of medical treatment heart and lung transplantation was performed after 49[30] and 50[18] days, respectively. Five of the patients had pericardial effusion at the time of diagnosis. Two of these underwent successful surgical repair, the third patient died before the operation was started.