Radiology Case Reports 19 (2024) 319–325

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Case Report

Large main pulmonary artery aneurysm: Case report and brief review of the literature ☆,☆☆

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ARTICLE INFO

Article history:
Received 22 June 2023
Revised 11 October 2023
Accepted 14 October 2023

Keywords:
Pulmonary artery
Aneurysm
Computed Tomography
Echocardiography

ABSTRACT

Pulmonary artery aneurysms are a rare but often fatal clinical entity with an estimated incidence of 1 in 14,000 individuals in postmortem studies. They can be congenital or acquired. No specific guidelines regarding their optimal management, medical or surgical, currently exist and treatment is planned on a case-by-case basis since data regarding their clinical course and prognosis are limited. We present the case of a 77-year-old male patient who presented at the Emergency Department of our hospital with a complaint of exertional dyspnea and dull substernal pain over 1 week. Upon investigation, a main pulmonary artery true aneurysm measuring 61 mm on Computed Tomography was detected. The patient's history was remarkable for heavy smoking, arterial hypertension, dyslipidemia, known ascending aortic aneurysm, moderate COPD, and past tuberculosis. He was admitted to the Cardiology unit and treated as a case of decompensated heart failure with preserved ejection fraction. His symptoms improved with intravenous diuretics. A past chest MRI report, 7 years before his current event, described the main PA aneurysm measuring 51-52 mm. Regarding the main PA aneurysm, the heart team decided to follow a conservative approach with regular

☆ Acknowledgments: The authors would like to acknowledge Mr Athanasios Vakalopoulos and Mr Lazaros Grigoriadis, radiologic technologists at the General Hospital of Florina "Eleni Th. Dimitriou" for their professionalism and meticulous collaboration in performing high-quality CT scans at our hospital. They also express their appreciation to Mr. Lazaros Grigoriadis for his invaluable assistance regarding the extraction of 3D volume-rendered images for the current case report.
☆☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.
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https://doi.org/10.1016/j.radcr.2023.10.038

follow-up visits based on the patient's comorbidities, functional status, and slow growth rate of the PA aneurysm. Management of pulmonary artery aneurysms requires a heart-team approach in the context of the patient's underlying conditions and symptoms. More data are required in order to guide a treatment plan with an acceptable risk – benefit profile for each patient.

Introduction

Pulmonary artery (PA) aneurysms are considered a rare clinical entity with an estimated incidence of 1 in 14,000 individuals based on postmortem examination [1]. In Computed Tomography (CT), a true PA aneurysm (involving all of the arterial layers) is defined as a focal dilatation of the main PA above 29 mm or of the interlobar PA above 17 mm whereas other specialists set the upper limit of normal to 40 mm for the main PA [2]. It can be congenital (due to heart defects or connective tissue disorders) or acquired (infections, vasculitis, pulmonary arterial hypertension, chronic pulmonary embolism, neoplasm, iatrogenic and idiopathic) [3]. We report the case of a 77-year-old male patient with a large main PA aneurysm diagnosed by transthoracic echocardiography and CT angiography.

Case report

We present the case of a 77-year-old male patient who visited our Emergency Department of our secondary-care hospital with a complaint of progressive exertional dyspnea and dull substernal pain over 1 week. The patient's history was remarkable for smoking (120 pack-years), arterial hypertension, dyslipidemia, known ascending aortic aneurysm, moderate Chronic Obstructive Pulmonary Disease (COPD) staged as GOLD 2-3 (postbronchodilator FEV1/FVC = 0.59 and FEV1% = 59.3, measured 1 year before his current visit) supported by intermittent oxygen therapy and CPAP at home and past history of tuberculosis. The patient was under treatment with amlodipine, cilazapril, torasemide, cardvedilol, simvastatin, inhaled tiotropium plus formoterol, and inhaled beclometasone. Upon clinical examination, his BP was 146/75 mm Hg, his pulse was regular at 87 beats/min and the patient was afebrile. Oxygen saturation on room air was 86% with the patient reporting a baseline measurement of 91% at home. No stigmata of dermatologic disease or features of Marfan's syndrome were found. A 2/6 early diastolic murmur was heard at the left upper sternal border. Decreased lung sounds were revealed and wheezing. An ECG showed sinus rhythm with an isolated premature ventricular contraction. Chest X-ray (CXR) revealed an increased cardiothoracic index, a widened middle mediastinum, a prominent right atrial contour, and a prominent main PA (Fig. 1).

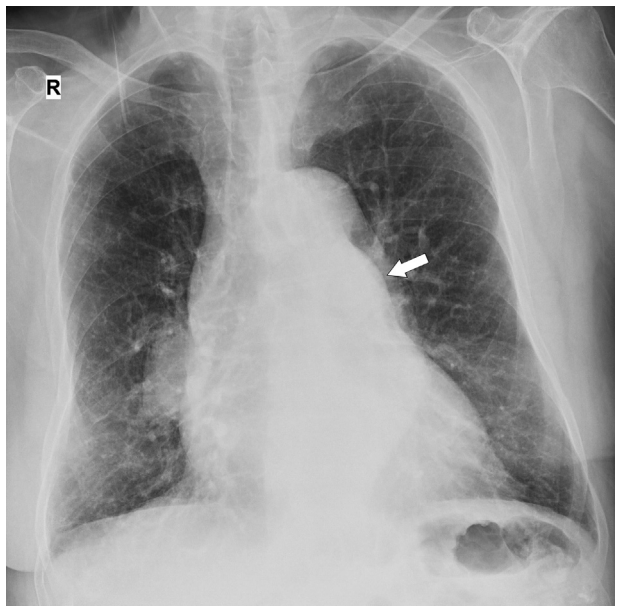

Fig. 1 – Chest X-Ray of the patient after his examination at the Emergency Department revealing a prominent main PA (white arrow).

Cardiac enzymes were negative for the presence of myocardial ischemia, d-dimers were within normal range, and complete blood count, basic metabolic panel, and basic liver panel were unremarkable. Transthoracic echocardiography (TTE) revealed mild concentric left ventricular hypertrophy, a borderline normal left ventricular ejection fraction (= 50%), a moderately dilated left atrium, dilatation of the aortic root at the level of the sinuses of Valsalva (max. diameter = 46 mm) and proximal ascending aorta (max. diameter = 44 mm), a tricuspid aortic valve with mild regurgitation, a mildly dilated right ventricle with normal contractility, mild tricuspid regurgitation and an estimated right ventricular systolic pressure (RVSP) of 48 mm Hg indicative of moderate pulmonary hypertension. Upon TTE, a parasternal short-axis view at the level of the great vessels revealed a markedly dilated main PA measuring 6.0 cm (Fig. 2). Mild to moderate regurgitation of the pulmonary valve was also present. Color flow mapping along the main PA was not suspicious for a patent ductus arteriosus.

A CT aortic angiography was performed to initially rule-out an acute aortic syndrome and visualize the pulmonary vessels and lung parenchyma. It confirmed the diagnosis of a large main PA aneurysm, without signs of rupture, measuring 61.4 mm (axial plane), 55.3 mm (sagittal plane), and 66.2mm (coronal plane) (Figs. 3 and 4). The right PA was also markedly dilated, measuring 43 mm. The known ascending aortic aneurysm was measured at 49 mm without signs of rupture. The lung parenchyma exhibited a "honeycomb" appearance with features typical of emphysema. The patient was

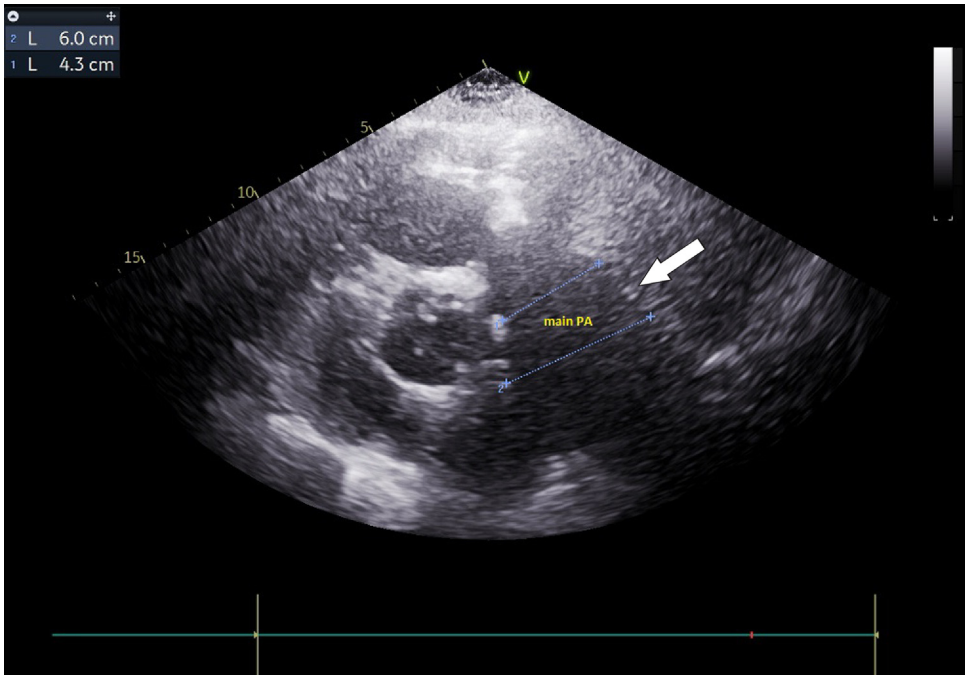

Fig. 2 – Transthoracic Echocardiography. Parasternal short axis view at the level of the great vessels, focused to the main PA revealing its dilatation measuring 6.0 cm (white arrow).

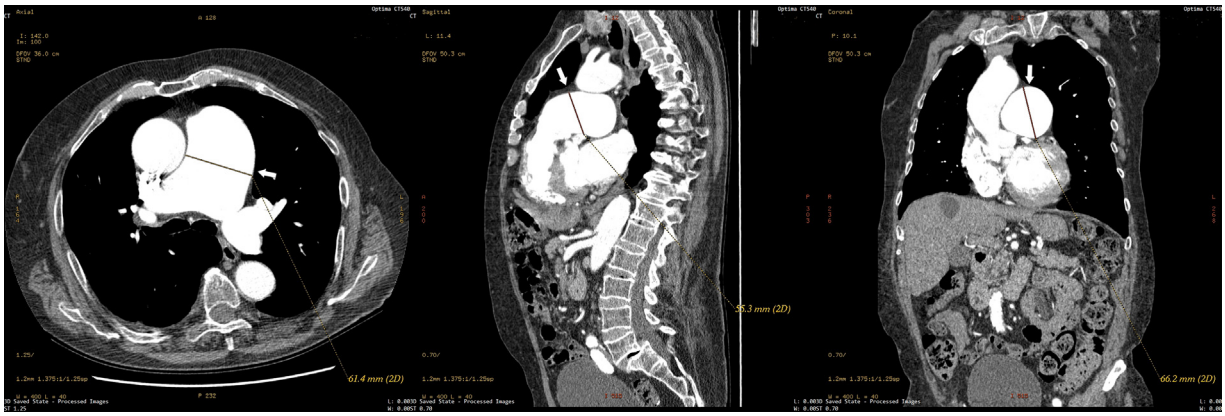

Fig. 3 – CT aortic angiography. Main PA aneurysm (white arrows) measuring 61.4 mm at the axial plane (first tile from the left), 55.3 mm at the sagittal plane (second tile from the left), and 66.2 mm at the coronal plane (last tile from the left).

admitted to the Department of Cardiology with a diagnosis of decompensated heart failure with preserved ejection fraction (HFpEF), responded well to intravenous diuretics and his symptoms alleviated in 2 days.

During his uneventful stay the patient provided a past report of a chest MRI and a TTE exam, both performed seven years before his current event as well as older TTE reports. Despite the fact that the main PA aneurysm was not mentioned in any of the old TTE reports, chest MRI described a main PA aneurysm measuring 51- 52 mm (Fig. 5), a dilated right PA measuring 32 mm, an ascending aortic aneurysm measuring 47-48 mm, a Ghon lesion at the right lower lobe and ipsilateral calcified lymph nodes (Ranke complex attributed to past tuberculosis infection). The patient mentioned that no surgical or PA aneurysm-specific consultation was provided back then.

During his stay at our unit, tertiary cardiac surgery consultation and subsequently the heart team recommended a conservative approach with regular follow-up visits, considering the slow growth rate of the main PA aneurysm (1 cm over 7 years), the patient's comorbidities and functional capacity. The patient was discharged on optimal HFpEF therapy and was referred as an outpatient to a specialized center for a CT Coronary Angiography which revealed non-obstructive coronary artery disease (CAD-RADS 2), confirmed the abovementioned findings regarding the main PA aneurysm and excluded

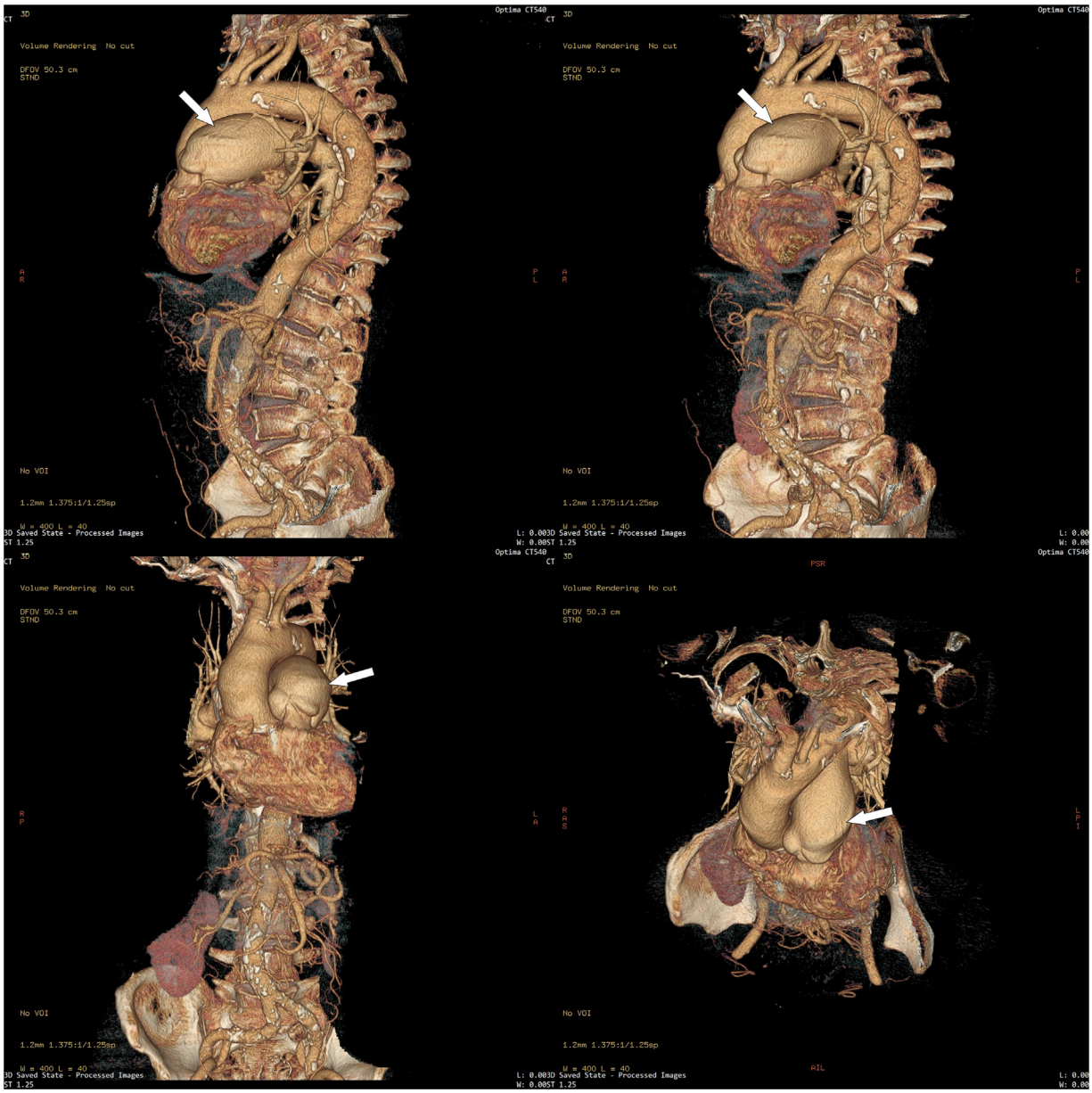

Fig. 4 – 3D Volume rendered images obtained from the CT aortic angiography. The large main PA aneurysm is marked with white arrows.

the presence of left to right shunting. The patient was also referred to a tertiary Pulmonary Arterial Hypertension center for further evaluation.

Discussion

PA aneurysms may be silent and asymptomatic or present with devastating symptoms like massive hemoptysis [3]. Considering the rarity of such finding and the lack of clear treatment guidelines, the main PA aneurysm of our patient remained unnoticed until his chest MRI scan 7 years before his current hospitalization and no specific guidance had been provided to him ever since. Surprisingly, the aneurysm followed a benign course growing only 10 mm in 7 years without any signs of rupture. In case of high diagnostic suspicion (eg, compatible symptoms, significant lung disease and probable pulmonary hypertension, suspicious CXR with prominent main PA, history of past significant infectious disease or connective tissue disorders etc.) the cardiologist performing TTE should insistently optimize the Parasternal Short Axis view (PSAX) at the level of the great vessels and focus to the main PA in order to adequately visualize its lateral border and obtain an approximate measurement of its diameter. The PSAX view in patients with a poor acoustic window (e.g. barrel chest, emphysema, obesity etc.) is challenging to perfect, requires patience and technical skill, therefore PA pathology can be missed. A plausible pathophysiological scenario regarding our patient's PA aneurysm could involve his

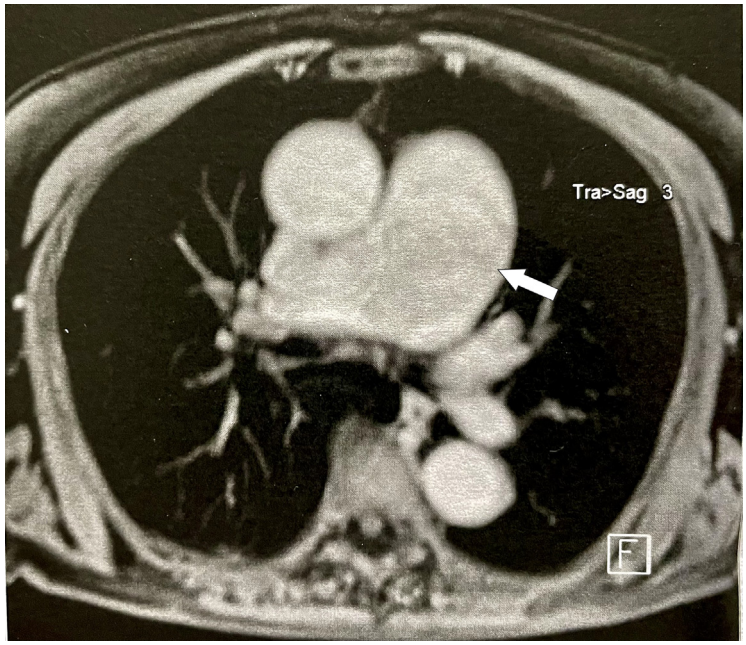

Fig. 5 – Frame obtained from the past chest MRI exam of the patient, seven years before his current event, revealing the main PA aneurysm (marked with a white arrow) measuring 51-52 mm.

Table 1 – Select case reports and case series describing main PA aneurysms, concomitant diseases for each patient, and subsequent management of the aneurysm.

Authors/Year	Patient's age in years/gender	Maximal main PA Aneurysm diameter in mm (diagnostic imaging modality)	Concomitant disease/findings	Management
Nair et al./2001 [5]	63/M	65 (CT)	Idiopathic	Main PA plication
Senbaklavaci et al./2001 [6]	34/F	80 (TTE)	Primary pulmonary hypertension - main PA dissection	T-shaped prosthetic graft replacement of the main PA (inclusion technique)
Agarwal et al./2002 [7]	20/F	60 (TTE + MRI + ANGIO)	Severe PV regurgitation	Aneurysmal sac excision and PV reconstruction with pericardial patches
Veldtman et al./2003 [8]	Case 1: 59/F	90 (TTE)	PV regurgitation, tiny PDA	Surgical - Hancock valved conduit
	Case 2: 37/M	80 (TTE + TOE + MRI)	Dextrocardia – PV stenosis and regurgitation	Carpentier-Edwards bioprosthesis and aneurysmorrhaphy of the main PA
	Case 3: 64/F	70 (TTE)	Congenital PV stenosis – COPD - PAD	Carpentier-Edwards porcine bioprosthesis. Plication of main PA
	Case 4: 45/M	100 (TTE)	Severe PV regurgitation	Carpentier-Edwards porcine bioprosthesis. Aneurysmorrhaphy of the main and proximal PA
Imazio et al./2004 [9]	64/M	60 (TOE + CT)	Idiopathic - mild PV regurgitation	Aneurysmorrhaphy
Deb et al./2005 [10]	62/M	65 (CT)	Idiopathic - PV regurgitation	Main PA resection and reconstruction with pulmonary SynerGraft homograft
Shih et al./2007 [11]	24/M	52 (CT)	Idiopathic - Systolic murmur at PV area	Aneurysmectomy and polytetrafluoroethylene (PTFE) graft replacement

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Table 1 (*continued*)

Authors/Year	Patient's age in years/gender	Maximal main PA Aneurysm diameter in mm (diagnostic imaging modality)	Concomitant disease/findings	Management
Vistarini et al./2007 [12]	51/M	52 (CT)	PV stenosis + RV dilatation	Surgical excision of pulmonary trunk and its left branch reconstruction with Dacron graft
Kotwica et al./2008 [13]	56/F	58 (TTE + CT)	Idiopathic	Regular follow-up
Muthialu et al./ 2010 [14]	25/F	60 (TTE + CT)	Severe PV regurgitation	Aneurysmectomy and reconstruction to indexed size
Shankarappa et al./2010 [15]	19/F	48 (CT)	Pulmonary hypertension – RV dysfunction	Medical therapy (sildenafil)
Seguchi et al./2011 [16]	45/M	70 (CT + TTE)	Idiopathic - PV regurgitation	Aneurysmectomy of the pulmonary main trunk with synthetic graft and valvuloplasty of pulmonary annulus
Orman et al./2013 [17]	16/F	38 (CT)	Idiopathic	Elective aneurysmectomy
Theodoropoulos et al./2013 [18]	Case 1: 41/F	65 (CT)	Idiopathic	Surgical main PA reconstruction
	Case 2: 76/F	44 (CT + MRA)	PFO	Aneurysmectomy and PFO closure
	Case 3: 61/F	40 (CT)	"Bovine" aortic arch – coronary artery calcifications	Serial follow-up
Hou et al./2016 [19]	Case 4: 28/F	50 (CT)	Idiopathic	Aneurysmectomy
	Case Series of 21 patients - 16 patients with full clinical data	35 – 80	11 cases secondary to Behcet's disease – 2 cases VSD – 2 cases post stenotic – 2 cases idiopathic – 4 cases PDA, ASD, tuberculosis, vasculitis respectively	5 cases surgical 8 cases medication 2 cases EVAR + medication 1 case transcatheter PDA closure
Sa-kong et al./2017 [20]	67/F	65 (CT)	Idiopathic - mild PV and TV regurgitation	Medical therapy (beraprost)

ANGIO, angiography; ASD, atrial septal defect; CT, computed tomography; EVAR, endovascular aneurysm repair; F, female; M, male; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging; PAD, peripheral artery disease; PDA, patent ductus arteriosus; PFO, patent foramen ovale; PV, pulmonary valve; RV, right ventricle; TOE, transesophageal echocardiography; TTE, transthoracic echocardiography; TV, tricuspid valve; VSD, ventricular septal defect.

COPD-related pulmonary hypertension (Group 3 pulmonary hypertension). Past tuberculosis infection is known to cause distal PA aneurysms (Rasmussen's aneurysms) a mechanism which does not fit our patient's case [1]. Our patient's main PA aneurysm cannot be classified as idiopathic based on the criteria proposed by Greene and colleagues who defined an idiopathic PA aneurysm as one which satisfies the following criteria: (1) simple dilatation of the pulmonary trunk with or without involvement of the rest arterial circulation; (2) absence of abnormal intracardiac or extracardiac shunts; (3) absence of chronic cardiac or pulmonary disease, clinical and at autopsy; and (4) absence of arterial disease such as syphilis, or more than simple atheromatosis or arteriolar sclerosis [4]. After a review of the available literature, a selection of main PA aneurysm case reports and case series is presented in Table 1. The majority of cases have been managed surgically with patients' ages ranging from 16 to 76 years old and an aneurysm size from 35 to 100 mm. Regarding the surgical approach, the heart team has to take the patient's comorbidities into consideration as well as the realistic possibility of a successful PA aneurysm operation. The surgical strategy of choice remains controversial. Extracorporeal circulation is mandatory for PA aneurysm operations with the introduction of venous and arterial cannulas. More specifically, the introduction of an arterial cannula into the ascending aorta with concomitant aortic cross-clamping is required. In addition, a venous cannula in the right atrium should be inserted and cold cardioplegia for diastolic arrest should be introduced to the myocardium. The femoral artery is also a good choice for the insertion of an arterial cannula as well as the femoral vein for a venous cannula. The Dacron graft is a satisfactory substitute for the replacement of the main PA as well as for the right and left PAs [18,21]. Theodoropoulos and colleagues describe a PA aneurysm replacement through the introduction of the patient to extracorporeal circulation under moderate hypothermia. However, in the 4th case, the PA aneurysm was replaced through mild hypothermia (32°C) [18]. There is no consensus on a specific strategy regarding the target temperature of the patients who will undergo PA aneurysm replacement. Beside PA aneurysms, there are reports of co-existing congenital heart disease such as ventricular septal defects, atrial septal defects and patent ductus arteriosus hence a concomitant operation might be needed [22]. Postsurgical complications include PA dissection and rupture, airway compression and PA thrombosis [23]. The current case report contributes to the existing body of relevant knowledge which includes a small number of published cases and there are currently no specific guidelines to advise the heart team about a PA aneurysm inter-

vention with an appropriate timing and optimal risk-benefit profile.

Patient consent

The patient provided written informed consent regarding this publication.

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