Rapid progression of pulmonary artery dilatation in pulmonary hypertension

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Abstract

We report the case of a 47-year-old woman who was admitted to the Cardiac Department for worsening dyspnea. The last chest computed tomography (CT) showed a rapid increase in pulmonary artery dimension (65 mm in 2019, 76 mm in 2021). The symptoms reported by the patient were due to important extrinsic compression of the left main coronary artery (LMCA). In this case, it is very difficult to choose the best therapeutic strategy. In the end, we decided to treat the left main coronary for prevention. After 3 months no new clinical symptoms have developed.

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Introduction

We described a case-report of a 47-year-old woman with a diagnosis from 2018 of primary pulmonary arterial hypertension treated with macitentan, sildenafil, selexipag and concomitant pulmonary artery dilation. After 2 years form diagnosis, a chest CT showed a rapidly increasing of pulmonary artery dimension (65 mm in 2019, 76 mm in 2021). Due to left main coronary artery (LMCA) extrinsic compression we decided to perform percutaneous coronary intervention (PCI) with unprotect drug-eluting stent (DES) on LMCA for primary prevention with good final result.

Case Report

A 47-year-old woman with a diagnosis of primary pulmonary arterial hypertension from 2018, diagnosed by right ventricular catheterization (systolic pulmonary artery pressure, PAPs 107 mmHg, mean pulmonary artery pressure PAPm 32 mmHg, diastolic pulmonary artery pressure, PAPd 50 mmHg) associated with a performance of 380 m at 6-minute walking test (6MWT), where she had dyspnea after 4 min. She had regular annually follow-up with cardiologic visit (included transthoracic echocardiogram, TTE), chest computed tomography (CT) and 6MWT. She was treated at the beginning with macitentan and sildenafil, after one year (2019) we decided to introduce selexipag for worsening at 6MWT (320 m with dyspnea at 3 min). In January 2021 she presented to the cardiology office for worsening dyspnea. The electrocardiogram showed sinus rhythm, without alterations. The transtoracic echocardiogram (TTE) showed right ventricular dilation (basal right ventricular diameter 60 mm) and hypokinetic (tricuspid annular plane systolic excursion, TAPSE 16 mm), high pulmonary hypertension (PAPs 90 mmHg) with minimal tricuspid regurgitation and medium regurgitation of the pulmonary valve. Left ventricular function was normal (ejection fraction, 60%) with paradoxical septum movement. The pulmonary artery (PA) was markedly enlarged (diameter 76 mm). Moreover, a CT imaging demonstrated a rapid and marked increasing of pulmonary artery dimension (65 mm in 2019, 76 mm in 2021) and its main branches (50 mm vs 47 mm at right, 42 mm vs 36 mm at left) (Figure 1). A right ventricular catheterization was performed, too: PAPs 120 mmHg, pulmonary artery wedge pressure (PAWP) 100 mmHg. A coronary angiography was then indicated, and revealed 80% of left main coronary artery (LMCA) stenosis in absence of atheromatous plaques. The LMCA was then assessed by

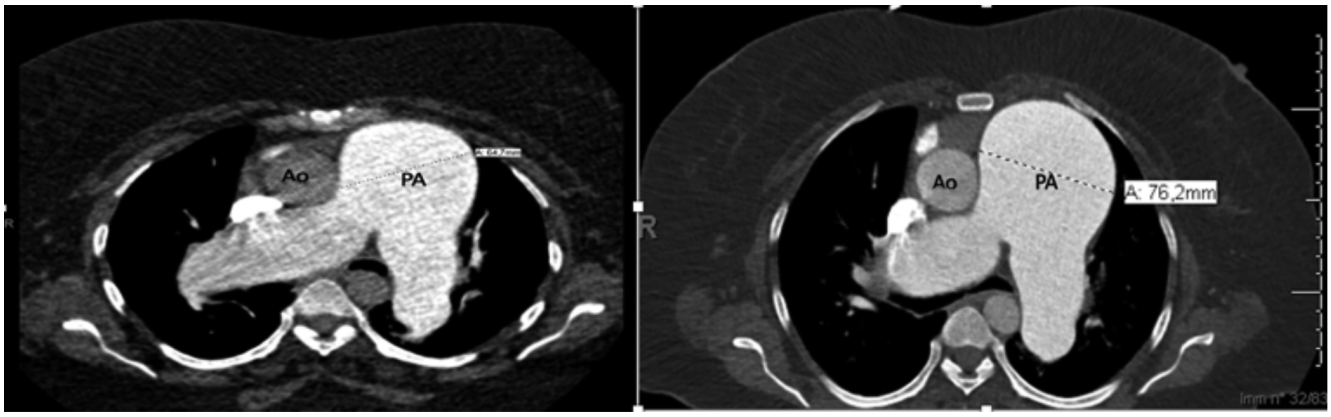

Figure 1. Pulmonary artery at 2019 CT. The CT evidenced a PA dimension of 64.7 mm (left) and a rapid PA enlargement of 76.2 mm (right). PA, pulmonary artery; Ao, aorta.

intravascular ultrasound (IVUS), which confirmed the presence of significant luminal narrowing. Considering the important extrinsic compression of LMCA in patient without angina or loss of consciousness, we decided to perform a percutaneous coronary intervention (PCI) with unprotect drug-eluting stent (DES) on LMCA with good final result. No complications occurred during and after the procedure. She was discharged after 48 h in clinical good conditions. On continued outpatient follow-up at Pulmonary Artery Hypertension Ambulatory, after 3 months no new clinical symptoms have developed.

An informed consent was signed by patient.

Discussion

Our patient presents a rapid enlarging of PA with compression of LMCA without symptoms except dyspnea. Treatment with PCI + DES on LMCA was therefore aimed not only at preventing symptoms, but also sudden death, especially in those patients who have rapid pulmonary artery dilation like ours (11 mm in 16 months).

This was the first time we decided to treat a LMCA compression by PA through PCI+ DES despite the absence of angina or loss of consciousness. Our patient, in fact, presents a rapid enlarging of PA with compression of LMCA without symptoms.

According to literature, pulmonary artery hypertension (PAH) is responsible of LMCA compression from 5% to 19% among patients, especially in those with PAH [1,2]. The mechanism was an extrinsic by dilatation of PA. LMCA compression by an enlarged PA is more often associated with congenital heart diseases, particularly atrial septal defect, ventricular septal defect, patent ductus arteriosus, or tetralogy of Fallot. The significant myocardial ischemia depends both on the degree of LMCA compression and its angle with the left sinus of Valsalva (particularly if less than 30°), and the ratio of the main PA to aorta of 2 or higher is also considered to be a risk factor for LMCA compression [3-5]. Therefore, Galiè *et al*. [6] demonstrated that a PA diameter of at least 40 mm represented the best predictor of LMCA stenosis of 50% or greater.

There is a paucity of protocols for LMCA compression in the setting of PAH. Aorto-coronary bypass and unprotected LMCA stent implantation are the only currently strategies. Given the high surgical mortality in patients with PAH, LMCA stenting has been favored as the revascularization strategy of choice, and several authors have reported successful results in this kind of patients [7].

In our case, we agreed with the literature to treat LMCA and thus prevent adverse events.

Therefore, an effective screening algorithm for the diagnosis of rapid PA dilatation and LMCA compression was needed, independently of symptoms [8,9].

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