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Left Main Ostial Compression in a Patient with Pulmonary Hypertension: Dynamic Findings by IVUS

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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Conflict of interest: None declared

Patient: Female, 39
Final Diagnosis: Idiopathic pulmonary arterial hypertension
Symptoms: Chest pain
Medication: —
Clinical Procedure: Percutaneous coronary intervention
Specialty: Cardiology and Pulmonology

Objective: Rare co-existence of disease or pathology

Background: Pulmonary artery dilatation is a common feature among patients with severe pulmonary hypertension. Left main coronary artery extrinsic compression by an enlarged pulmonary artery is a rare complication and a potential cause for chest pain and sudden cardiac death in patients with pulmonary hypertension. This situation is very rare and few reports have described it. Currently, the appropriate management of these patients remains unknown.

Case Report: In the present report we describe the case of a 39-year-old woman who presented with a 2-year history of cardiac symptoms related to exercise. The patient underwent a 64-slice multidetector computed tomography (MDCT) coronary angiography, which showed left main coronary artery (LMCA) compression by a markedly enlarged pulmonary artery trunk (44 mm), without intraluminal stenosis or coronary artery calcium, as determined by the Agatston score. This compression was considered to be the cause of the cardiac symptoms. To confirm and plan the treatment, the patient underwent cardiac catheterization that confirmed the diagnosis of pulmonary hypertension and LMCA critical obstruction. Taking into account the paucity of information regarding the best management in these cases, the treatment decision was shared among a "heart team" that chose percutaneous coronary intervention with stent placement. An intra-vascular ultrasound was performed during the procedure, which showed a dynamic compression of the left main coronary artery. The intervention was successfully executed without any adverse events.

Conclusions: This case illustrates dynamic compression of the LMCA by IVUS, visually demonstrating the mechanism of the intermittent symptoms of myocardial ischemia in this kind of patient. It also shows that percutaneous stenting technique may be an appropriate treatment for this unusual situation.

MeSH Keywords: Hypertension, Pulmonary • Stents • Ultrasonography, Interventional

Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/895668>



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Background

Pulmonary arterial hypertension (PAH) is a syndrome resulting from restricted flow through the pulmonary arterial circulation, resulting in increased pulmonary vascular resistance and ultimately in right heart failure [1]. Pulmonary artery (PA) dilatation is a common feature among these subjects, present in 76.6% of the cases in a cohort of severe pulmonary hypertension patients [2]. The PA dilatation is also considered an independent risk factor for death not explained by right ventricular failure or comorbidities in patients with PAH [3]. One of the causes of death related to pulmonary artery dilatation is left main coronary artery extrinsic compression, which is a rare complication but a potential cause for chest pain and sudden cardiac death in patients with pulmonary hypertension [3]. The diagnosis of this mechanism of myocardial ischemia may be challenging among patients without known history of PAH. Upon identifying this rare complication, the traditional treatment option used to be open surgery, but reports with limited data in the last decade have shown encouraging results of percutaneous treatment.

Case Report

We describe the case of a 39-year-old woman without comorbidities or use of medication who presented with a 2-year history of chest pain, dyspnea, and palpitations after moderate exertion. Additionally, the chest pain was worsened by left-side decubitus and leaning forward. No other symptoms, such as fever, paroxysmal nocturnal dyspnea, or orthopnea, were present. Physical examination revealed a prominent pulmonary component of S₂, without jugular venous distention or cardiac murmurs, and with clear lungs. A 12-lead electrocardiogram (Figure 1) showed normal sinus rhythm and a T-wave inversion in precordial leads. Chest radiograph (Figure 1) revealed right prominent pulmonary arteries. The transthoracic echocardiography (Figure 1) showed a dilated pulmonary trunk, with preserved function of the right and left ventricles

and normal-sized bilateral chambers. The patient underwent a 64-slice multidetector computed tomography (MDCT) coronary angiography, which showed left main coronary artery (LMCA) compression by a markedly enlarged pulmonary artery trunk (44 mm), without intraluminal stenosis or coronary artery calcium, as determined by the Agatston score (Figure 2). A right cardiac catheterization was performed, and demonstrated a mean right atrial pressure of 10 mmHg and pulmonary artery pressure of 85/40 mmHg (mean of 55 mmHg). Coronary angiography demonstrated 95% diameter LMCA ostial stenosis (Figure 2, Video 1).

After discussion of this patient's case in a "Heart Team" meeting, we decided to treat the lesion with percutaneous angioplasty. Before this procedure, an intra-vascular ultrasound (IVUS) was performed, demonstrating dynamic compression of the LMCA with a minimal luminal area of 5.7 mm² and no atherosclerotic disease (Figure 2, Video 2). The patient underwent LMCA stenting with a 5.0×13 mm (18 atm) bare-metal stent (Figure 2, Video 3) and a subsequent IVUS revealed adequate expansion of the stent (Video 4). MDCT after angioplasty demonstrated an adequately placed stent, without any compression of the LMCA (Figure 2). Workup for the cause of pulmonary hypertension was consistent with primary pulmonary hypertension and the patient was started on sildenafil. The patient had complete resolution of the symptoms after PCI and still asymptomatic one year after the PCI.

Discussion

Chest pain is a common complaint among patients with pulmonary hypertension and is frequently attributed to distension of the pulmonary artery or right ventricular ischemia [4,5]. The symptoms could be intermittent because arterial, ventricular, and pulmonary abnormalities exacerbate during exercise among patients with PAH [4–6]. Because these patients are often young women, investigation of pain etiology usually does not encompass a thorough workup for coronary heart disease, especially among



Figure 1. Twelve-lead electrocardiogram showed normal sinus rhythm and a T-wave inversion in precordial leads (A). Chest radiograph revealed a prominent pulmonary artery (B). Transthoracic echocardiography in the parasternal short axis view showing a dilated pulmonary artery (48-mm) (C).

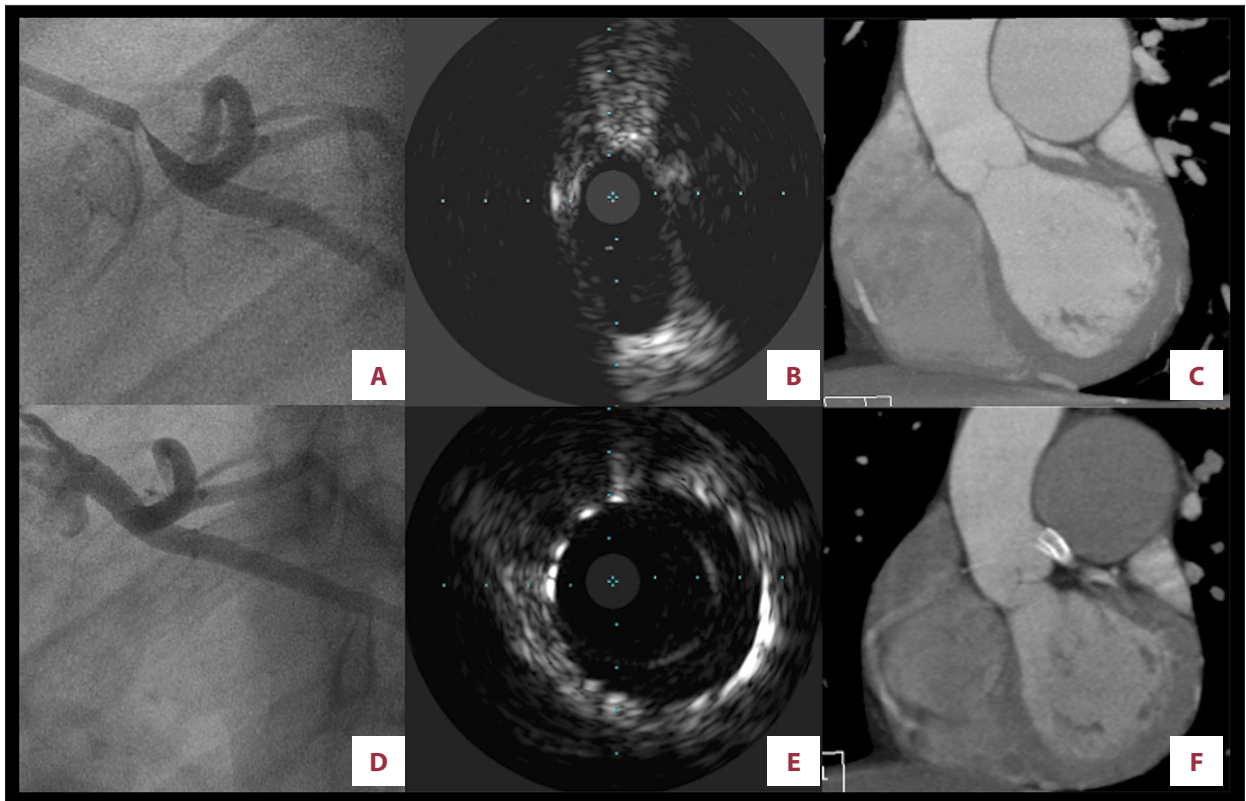
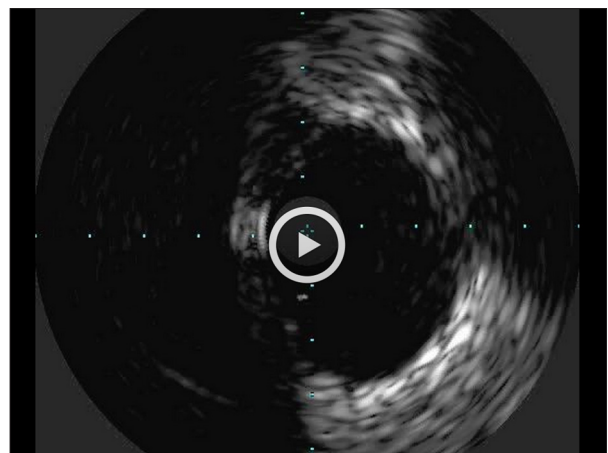


Figure 2. Coronary angiography demonstrating severe LMCA ostial stenosis (A). Intravascular ultrasound (IVUS) image of the LMCA showing ostial compression by the pulmonary trunk, with minimal luminal area of 5.8 mm² (B). Multidetector computed tomography (MDCT) coronary angiography showed left main coronary artery (LMCA) compression by a markedly enlarged pulmonary artery trunk (44mm) (C). The patient underwent LMCA angioplasty with a 5.0×13 mm bare-metal stent, with excellent angiographic result (D). IVUS after angioplasty showing a well-apposed stent, with minimal luminal area of 17.5 mm² (E). MDCT after angioplasty demonstrating an adequately placed stent, without LMCA compression by the pulmonary artery trunk (F).



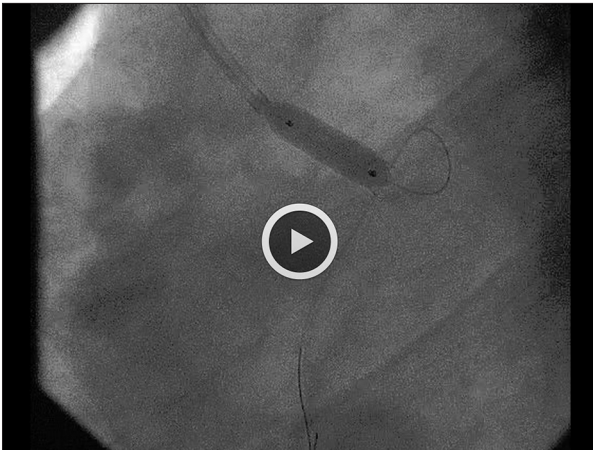
Video 1. LMCA ostial stenosis demonstrated by coronary angiography.



Video 2. IVUS demonstrating compression of the LMCA with no atherosclerotic disease.

patients without previous diagnosis of PAH. However, left main coronary artery (LMCA) extrinsic compression by an enlarged pulmonary artery is a potential cause for chest pain and sudden

cardiac death in such patients. The extrinsic compression of the left main coronary artery (LMCA) secondary to pulmonary artery trunk (PA) dilatation is a relatively new syndrome associated



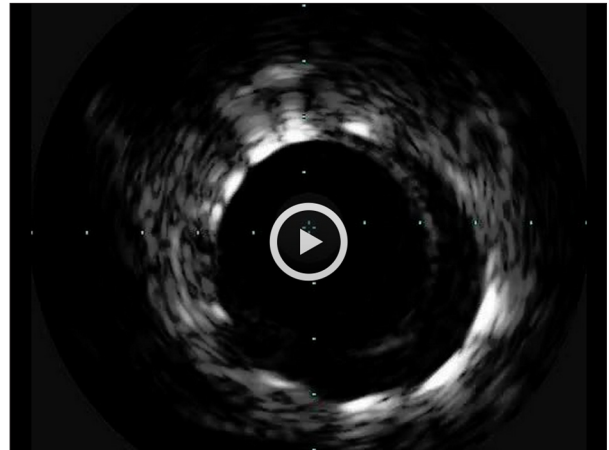
Video 3. Left main coronary artery stenting.

with pulmonary hypertension in cyanogen congenital cardiopathies [5,7–10] and also with a few cases secondary to idiopathic pulmonary arterial hypertension reported in the literature [11–16]. The pulmonary artery diameter is the most important aspect of the LMCA stenosis in these cases, but other aspects like mean pulmonary artery pressure or angina symptoms were not significantly associated with coronary compression [16]. Previous analysis suggested that these cases of ischemic syndrome are also related with an LMCA ostium located on the right side of the left sinus of Valsalva, and that a take-off angle $<45^\circ$ was associated with sudden death [5,17]. These observations show that the positional relationship between the LMCA and pulmonary artery is also an important aspect of this syndrome.

It is important to confirm myocardial ischemia caused by stenosis due to LMCA compression before indicating a treatment [18]. There are patients in whom the hemodynamics can be stabilized only by treating the cause of pulmonary hypertension. This effect was reported in cardiac defects [19,20], but in primary hypertension the reported results are conflicting [4,21,22]. A study showed improvement in angina symptoms after the administration of sildenafil and prostacyclin [4], while others showed that coronary revascularization was necessary due to the lack of improvement in angina symptoms after treatment with drugs such as bosentan [4,21]. Regarding the best option for revascularization, some authors believe in the need for surgical coronary revascularization in cases with significant LMCA obstruction [19]. However, pulmonary hypertension has been shown to increase the rate of post-operative

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Video 4. Post-PCI IVUS demonstrating adequate expansion of the stent.

mortality in patients with LMCA stenosis caused by pulmonary artery compression [22–24], especially as a consequence of acute right ventricular failure. Given the high surgical mortality in patients with PH, LMCA stenting has been used for revascularization and several authors have reported successful results in this kind of patient [24,25]. All of the reported cases involved compression of the ostium or proximal LMCA and required single-stent placement. In the current report, the IVUS findings showed clearly the dynamic changes of the lumen pre-PCI. Also in the reported case, the post-PCI analysis of the LMCA by IVUS showed that the stent effectively treated the compression.

Conclusions

This case illustrates by IVUS the dynamic compression of the LMCA and demonstrates successful treatment using a percutaneous stenting technique. Physicians should have a high index of suspicion for this complication because pulmonary artery dilatation is common among patients with PAH but the LMCA compression is rare and may be dynamic with intermittent clinical manifestations.

Statement

The authors report no financial relationships or conflicts of interest regarding the content herein.

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