Case Report

Left Main Coronary Artery Compression in Pulmonary Arterial Hypertension: Percutaneous Treatment to Improve Symptoms

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ABSTRACT

A 51-year-old woman with pulmonary arterial hypertension presented with progressive chest pain and dyspnea. Computed tomography imaging showed significant enlargement of her main pulmonary artery (PA) and was suggestive of left main coronary artery (LMCA) compression by the PA. The patient underwent percutaneous coronary intervention, which confirmed the diagnosis and a stent was deployed to the LMCA. Three months after the procedure the patient has near resolution of her symptoms. LMCA compression by an enlarged PA is an important cause of chest pain in patients with pulmonary arterial hypertension and can be managed safely and effectively with percutaneous coronary intervention and stenting.

RÉSUMÉ

Une femme de 51 ans atteinte d'hypertension artérielle pulmonaire présentait des symptômes progressifs de dyspnée et de douleur thoracique. Les images de la tomographie par ordinateur ont révélé un élargissement de l'artère pulmonaire (AP) principale et indiquaient une compression de l'artère coronaire gauche principale (ACGP) par l'AP. L'intervention coronarienne percutanée subie par la patiente a confirmé le diagnostic, et une endoprothèse coronaire a été déployée à l'intérieur de l'ACGP. Les symptômes de la patiente avaient pratiquement disparu trois mois après l'intervention. La compression de l'ACGP par une AP élargie est une cause importante de douleur thoracique chez les patients atteints d'hypertension artérielle pulmonaire qui peut être traitée de façon sûre et efficace par une intervention coronarienne percutanée et une endoprothèse coronaire.

This case highlights an under-recognized cause of chest pain in a patient with pulmonary arterial hypertension (PAH). There is a high incidence of typical and atypical angina in these patients, and it is often attributed to the consequences of elevated right-sided pressures. In this case report we describe a scenario in which the patient's chest pain was secondary to external compression of the left main coronary artery (LMCA) by an enlarged pulmonary artery (PA), causing coronary ischemia.

Case

A 51-year-old woman with a 17-year history of idiopathic PAH presented to the emergency department with 12 months

of progressive exertional chest pain accompanied by worsening exertional dyspnea. Her PAH was managed with macitentan 10 mg daily and riociguat 1.5 mg 3 times daily along with furosemide and digoxin. Her functional status was World Health Organization Class II. Her most recent right and left heart catheterization was done 15 months earlier because of palpitations and presyncope. Her coronary circulation at that time showed no atherosclerosis but there was a 50% narrowing of the LMCA. Her mean PA pressure at that time was 67 mmHg. Apart from idiopathic PAH, she had a history of *in situ* pulmonary thrombosis, paroxysmal atrial tachycardia, ulcerative colitis, and alopecia.

Upon presentation to hospital, she had a blood pressure of 100/55 mm Hg and otherwise normal vitals. On cardiac examination she had a loud P₂ with a right ventricular (RV) heave and a holosystolic murmur consistent with tricuspid regurgitation. Her jugular venous pressure was not elevated and there was no pedal edema. Breath sounds were normal. Her electrocardiogram showed sinus rhythm with diffuse nonspecific ST-T wave abnormalities, as well as changes consistent with RV hypertrophy and strain. The troponin level was newly elevated at 348 ng/L (0–30 ng/L), whereas her creatine kinase and D-dimer were normal. A chest radiograph

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Ethics Statement: Research reported has adhered to the relevant ethical guidelines. Patient informed consent was obtained prior to publication of this case report.

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See page 692 for disclosure information.

Novel Teaching Points

- Compression of the LMCA by an enlarged PA can be a cause of chest pain in patients with advanced PAH.
- Computed tomography coronary angiogram is the best first step for evaluating extrinsic LMCA compression.
- PCI with stent deployment is a safe and effective treatment.

showed clear lung fields but significant enlargement of the central pulmonary arteries.

A computed tomography (CT) scan of the chest was ordered to rule out an aortic dissection and pulmonary embolism. There was no evidence of either, however the scan showed severe PA enlargement, with the main PA measuring 5.3 cm in diameter. In contrast, the ascending aorta measured 2.3 cm (Fig. 1A). The main PA was 4.8 cm on a cardiac magnetic resonance imaging study 2 years earlier. Careful review of the nongated CT chest study showed compression of the LMCA by the enlarged PA (Fig. 1B) and the take-off angle of the LMCA from the aortic root was 52°.

A repeat coronary angiogram was arranged to assess if there had been progression of the LMCA stenosis (Fig. 2A). The angiogram demonstrated a > 90% stenosis of the ostial LMCA (Fig. 2B), which made it difficult to seat the catheter. The rest of the coronary segments were normal. Percutaneous coronary intervention (PCI) was performed using the radial approach with unfractionated heparin and dual antiplatelet pretreatment, using a JL3.5 guide catheter (Cordis, Miami, FL). A second-generation drug-eluting stent (3.5 × 9 mm Resolute; Medtronic, Minneapolis, MN) was deployed at maximal pressure of 18 atm at the site of stenosis in the LMCA, with complete resolution of the obstruction to blood flow (Fig. 2C). A 9-mm stent length was required to avoid the distal left main bifurcation. The patient was prescribed clopidogrel along with her previous medications, which included rivaroxaban.

Three months after the procedure, the patient has near resolution of her chest pain and her exercise tolerance has improved 1 grade. She is able to perform all activities of daily living.

Discussion

Chest pain is a common symptom in patients with PAH.^{1,2} Classically it is thought to occur secondary to elevated right-sided pressures leading to RV strain, impairment of coronary perfusion, and RV ischemia.² In this report we present a less common and under-recognized cause of anginal symptoms in a patient with advanced PAH and extrinsic compression of the LMCA by an enlarged PA. Compression of the LMCA by an enlarged PA is more likely with PA diameter > 4.0 cm or a PA to ascending aorta diameter ratio of > 1.5.¹ It should be considered in PAH patients with chest pain, because it can lead to complications including myocardial infarction,³ cardiogenic shock,³ and arrhythmias that might contribute to the incidence of sudden death in PAH patients.⁴ Interestingly, despite a relatively modest increase in diameter of the PA from 4.8 cm to 5.3 cm, there was a significant increase in the degree of external compression on the LMCA. To our knowledge, measurements of LMCA stenosis and PA diameter at multiple time points has not previously been described.

Anginal-like chest pain and worsening dyspnea on exertion are the most common presenting symptoms.¹ Among a large cohort of patients with PAH, those with angina (16%) underwent CT coronary angiogram (CTCA) studies.¹ Forty percent of those with angina had LMCA stenosis ≥ 50%. Those whose CTCA were suggestive of LMCA compression went on to have coronary angiography. The CTCA findings that were most predictive of significant stenosis were clear compression by the PA, but also a take-off angle of the left coronary artery from the aorta of less than 60°, as was the case with our patient.

PCI with placement of a stent to the LMCA can produce good results. Galie et al. described 45 patients with extrinsic LMCA compression who received PCI stenting.¹ Most were treated with dual antiplatelet therapy for at least 1 month, and > 95% of patients had improvement or resolution of symptoms. Nearly 90% of patients had stent patency at 9 months. This cohort was followed-up over a mean of 4.5 years, and during that time survival was similar compared with a matched population with PAH and angina who did not have extrinsic compression of the LMCA.⁵ A second-generation drug-eluting stent was used most often, on the basis of its superior radial strength and low restenosis rate. Because of the absence of atherosclerosis, Galie et al. concluded that neither

Figure 1. (A) Computed tomography image showing the enlarged pulmonary artery (PA) with a diameter of 5.3 cm compared with the ascending aorta (A) diameter of 2.3 cm. (B) The left main coronary artery (LMCA) exiting from the aortic root (A) and compression by a large adjacent PA can be seen. Take-off angle of the LMCA is visualized at 52°.

Figure 2. (A) Coronary angiogram done 1 year before the current presentation. There is 50% stenosis of the ostial left main coronary artery (LMCA) with otherwise normal anatomy. (B) At presentation, there is > 90% stenosis at the ostial LMCA. (C) After deployment of a drug-eluting stent to the LMCA, there is normal blood flow.

aggressive postdilation, or intravascular ultrasound are mandated. Because stent positioning can be more challenging without atherosclerosis, they also suggested a Judkins left guide catheter and suggested avoiding very short stents.⁵ Surgical approaches such as PA aneurysm repair have been described, but the perioperative risk in patients with PAH is high.⁶ Because of the high likelihood of success with PCI, it is a reasonable first therapeutic option for extrinsic compression of the LMCA by an enlarged PA.

Conclusion

In conclusion, extrinsic compression of the LMCA by an enlarged PA in patients with PAH is an important cause of angina. It should be suspected in a PAH patient with typical or atypical anginal symptoms. CTCA is a useful first test in evaluating for this condition. Coronary angiography is needed to confirm the LMCA stenosis and permit PCI with stent deployment, which can lead to long-term improvement in symptoms.

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Disclosures

The authors have no conflicts of interest to disclose.

References

- Galie N, Saia F, Palazzini M, et al. Left main coronary artery compression in patients with pulmonary arterial hypertension and angina. *J Am Coll Cardiol* 2017;69:2808-17.
- Thenappan T, Ormiston ML, Ryan JJ, Archer SL. Pulmonary arterial hypertension: pathogenesis and clinical management. *BMJ* 2018;360:1-27.
- Vaseghi M, Lee JS, Currier JW. Acute myocardial infarction secondary to left main coronary artery compression by pulmonary artery aneurysm in pulmonary arterial hypertension. *J Invasive Cardiol* 2007;19:E375-7.
- Dalonzo GE, Barst RJ, Ayres SM, et al. Survival in patients with primary pulmonary-hypertension - results from a national prospective registry. *Ann Intern Med* 1991;115:343-9.
- Saia F, Dall'Ara G, Marzocchi A, et al. Left main coronary artery extrinsic compression in patients with pulmonary arterial hypertension. *JACC Cardiovasc Interv* 2019;12:319-21.
- Van Tittelboom N, Devos D, Thierry B, De Pauw M, Vandecasteele E. Recurrent chest pain and dyspnoea in a patient with pulmonary arterial hypertension. *Acta Clinica Belgica* 2019;74:292-5.