



## Left main coronary artery compression in a patient with portopulmonary hypertension

Iara Teixeira de Araújo<sup>1,a</sup>, Pammela Jacomeli Lembi<sup>1,b</sup>,  
Eduardo Belisario Falchetto<sup>2,c</sup>, Ricardo de Amorim Corrêa<sup>3,4,d</sup>

### TO THE EDITOR,

We report the case of a 54-year-old male patient with a diagnosis of portopulmonary hypertension (POPH) secondary to alcoholic liver cirrhosis (Child-Pugh class C and Model for End-Stage Liver Disease = 16). The patient was referred to the Outpatient Clinic for Disorders of the Pulmonary Circulation of the Federal University of Minas Gerais *Hospital das Clínicas*, located in the city of Belo Horizonte, Brazil, for assessment to inform decision-making about liver transplantation. At the initial evaluation, he complained of retrosternal pain that was brought on by moderate physical exertion and resolved with rest, without syncope or presyncope. Physical examination revealed a regular cardiac rhythm and fixed splitting of the second heart sound over the pulmonary area, and pulmonary auscultation was unremarkable. An echocardiogram revealed mild enlargement of the right atrium and right ventricle, with an estimated pulmonary artery systolic pressure of 40 mmHg. His spirometry results were as follows: FEV<sub>1</sub> = 3.80 L (101% of predicted); FVC = 4.95 L (104% of predicted); FEV<sub>1</sub>/FVC ratio = 0.77 (97% of predicted); and a negative bronchodilator test. Lung radionuclide imaging findings suggested a low probability of pulmonary thromboembolism. A computed tomography scan of the chest showed normal attenuation of the lung parenchyma, with no pleural effusion, masses, or lymph node enlargement. Right heart catheterization revealed a mean pulmonary artery pressure of 48 mmHg, pulmonary artery resistance of 6.22 Woods units, a diastolic gradient of 25 mmHg, and a cardiac index of 3.17 L/min/m<sup>2</sup>. On the basis of these findings, the patient received a confirmed diagnosis of POPH, with limitation corresponding to functional class II, and stable angina, class III, due to extrinsic compression of the left main coronary artery (LMCA).

The patient was then hospitalized to undergo further diagnostic assessment and therapeutic planning. Coronary computed tomography angiography (Figures 1A and 1B) showed severe ostial LMCA stenosis that was probably due to extrinsic compression by the main pulmonary artery (MPA). Coronary cineangiography (Figure 1C) revealed severe stenosis in the proximal third (80%) of the LMCA.

The patient underwent percutaneous angioplasty with placement of a drug-eluting stent into the LMCA (Figure 1D), followed by antiplatelet therapy with acetylsalicylic

acid and clopidogrel. He was started on sildenafil 20 mg three times daily to treat pulmonary arterial hypertension (PAH). The patient experienced significant improvement in symptoms and functional capacity.

Angina symptoms are usually associated with coronary atherosclerosis, although they may be due to other conditions such as inflammatory diseases and extrinsic compression of coronary arteries.<sup>(1)</sup> Of those conditions, LMCA compression by a dilated MPA has been reported as a potentially reversible cause of angina and ventricular dysfunction in patients with PAH. This type of compression is primarily related to congenital heart disease or idiopathic PAH; however, other forms of PAH, such as POPH, can trigger it. Unlike atherosclerotic coronary disease, which is more prevalent in elderly patients, extrinsic compression of the LMCA seems to affect younger patients.<sup>(2)</sup>

Extrinsic compression of the LMCA can cause chest pain, left ventricular dysfunction, ischemia, and sudden death. Its implications in terms of function and prognosis remain poorly characterized, but malignant arrhythmias and ventricular dysfunction can contribute to increasing the incidence of sudden death in patients with PAH.<sup>(3)</sup> LMCA compression seems to be related to MPA diameter (greater than 49 mm) and to the ratio of MPA diameter to aorta diameter, as measured by echocardiography.<sup>(4)</sup>

The current literature on PAH has emphasized the need to generate and investigate a second diagnostic hypothesis for angina or angina-like symptoms in patients with PAH. In the case reported here, the patient underwent coronary computed tomography angiography and coronary cineangiography to elucidate the cause of his angina symptoms. Currently, coronary cineangiography is the gold standard test for making this diagnosis. When viewed with contrast, the LMCA appears narrowed at its origin and has a dilated tubular appearance distal to the obstruction (Figure 1).<sup>(2)</sup> Noninvasive techniques, such as coronary computed tomography angiography, cardiac magnetic resonance imaging, and transesophageal echocardiography, can demonstrate the origin and proximal course of the coronary arteries.

LMCA compression in patients with PAH is a treatable cause of angina and left ventricular ischemia. However, despite the fact that numerous treatment options have been described for LMCA compression, there is still controversy regarding optimal therapy for this condition.

1. Faculdade de Medicina, Universidade Federal de Minas Gerais – UFMG – Belo Horizonte (MG) Brasil.

2. Hospital Felício Rocho, Serviço de Hemodinâmica, Belo Horizonte (MG) Brasil.

3. Programa de Pós-Graduação de Ciências Aplicadas à Saúde do Adulto, Departamento de Clínica Médica, Faculdade de Medicina, Universidade Federal de Minas Gerais – UFMG – Belo Horizonte (MG) Brasil.

4. Ambulatório de Doenças da Circulação Pulmonar, Serviço de Pneumologia e Cirurgia Torácica, Hospital das Clínicas, Universidade Federal de Minas Gerais – UFMG – Belo Horizonte (MG) Brasil.

a. <http://orcid.org/0000-0001-5246-637X>; b. <http://orcid.org/0000-0001-7336-2051>;

c. <http://orcid.org/0000-0003-1717-2223>; d. <http://orcid.org/0000-0003-1779-0443>



**Figure 1.** Coronal (in A) and axial (in B) images from coronary computed tomography angiography showing extrinsic compression of the left main coronary artery (LMCA) by the main pulmonary artery (MPA). In C, coronary cineangiography showing severe stenosis in the proximal third of the LMCA (arrow). In D, coronary cineangiography showing the LMCA after stenting (arrow). Ao: aorta.

Treatment options include percutaneous coronary intervention (PCI), coronary artery bypass grafting, and heart-lung transplantation.<sup>(3)</sup> Although coronary artery bypass grafting remains the treatment of choice for LMCA revascularization, PCI may be considered in patients with anatomic conditions associated with a low risk of PCI procedural complications or in patients with clinical conditions that predict an increased risk of adverse surgical outcomes. In 2017, Galiè et al.<sup>(5)</sup> presented data on the prevalence of extrinsic compression of the LMCA by a dilated MPA in patients with PAH and angina, as well as the results of PCI. Of 765 patients with PAH, 121 (16%) presented with symptoms that were consistent with angina pectoris. Of those 121 patients, 94 (78%) had abnormal computed tomography angiography findings. Significant LMCA stenosis was detected by coronary angiography in 48 (40%) of the 121 symptomatic patients. A total of 45

patients underwent PCI with stenting, with symptom relief occurring in 91% of the patients, and 5 (11%) developed restenosis.<sup>(5)</sup> In the case reported here, the patient underwent percutaneous angioplasty with drug-eluting stenting in the LMCA, followed by antiplatelet therapy with acetylsalicylic acid and clopidogrel. For treating PAH, he was started on sildenafil at a dose of 20 mg three times daily. The patient experienced significant improvement in symptoms and functional capacity.

In conclusion, retrosternal pain caused by extrinsic compression of the LMCA by a dilated MPA should be considered in the differential diagnosis of angina or left ventricular dysfunction in patients with PAH. Percutaneous stent implantation can yield good angiographic and clinical outcomes. Further studies are needed in order to determine the impact of PCI on the prognosis of such patients.

## REFERENCES

- Rubín JM, Arias JC, Lambert JL. Unusual case of coronary stenosis caused by an external compression. *Int J Cardiol.* 1997;62(2):167-9. [https://doi.org/10.1016/S0167-5273\(97\)00199-X](https://doi.org/10.1016/S0167-5273(97)00199-X)
- Kajita LJ, Martínez EE, Ambrose JA, Lemos PA, Esteves A, Nogueira da Gama M, et al. Extrinsic compression of the left main coronary artery by a dilated pulmonary artery: clinical, angiographic, and hemodynamic determinants. *Catheter Cardiovasc Interv.* 2001;52(1):49-54. [https://doi.org/10.1002/1522-726X\(200101\)52:1<49::AID-CCD1012>3.0.CO;2-0](https://doi.org/10.1002/1522-726X(200101)52:1<49::AID-CCD1012>3.0.CO;2-0)
- Lindsey JB, Brilakis ES, Banerjee S. Acute coronary syndrome due to extrinsic compression of the left main coronary artery in a patient with severe pulmonary hypertension: successful treatment with percutaneous coronary intervention. *Cardiovasc Revasc Med.* 2008;9(1):47-51. <https://doi.org/10.1016/j.carrev.2007.07.003>
- Mesquita SM, Castro CR, Ikari NM, Oliveira SA, Lopes AA. Likelihood of left main coronary artery compression based on pulmonary trunk diameter in patients with pulmonary hypertension. *Am J Med.* 2004;116(6):369-74. <https://doi.org/10.1016/j.amjmed.2003.11.015>
- Galiè N, Saia F, Palazzini M, Manes A, Russo V, Bacchi Reggiani ML, et al. Left Main Coronary Artery Compression in Patients With Pulmonary Arterial Hypertension and Angina. *J Am Coll Cardiol.* 2017;69(23):2808-2817. <https://doi.org/10.1016/j.jacc.2017.03.597>