Primary pulmonary hypertension is often associated with angina-like chest pain of uncertain etiology. Left main coronary artery compression by the pulmonary artery is a treatable cause of angina and should be considered in these patients. We describe a patient presenting with primary pulmonary hypertension, clinical angina and extrinsic compression of the left main coronary artery by the pulmonary artery, who was treated with direct stenting.

**Key words:** Primary pulmonary hypertension. Left main coronary artery. Stent.

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**INTRODUCTION**

Angina-like chest pain is a common symptom in patients with primary pulmonary hypertension (PPH). Because this disease affects young people generally without cardiovascular risk factors, these pains have not been associated with the presence of coronary arteriosclerosis. It has been proposed that the etiology of these symptoms may be due to the painful dilation of the pulmonary artery or to right ventricular ischemia. However, another potential cause of angina in these patients has been reported: the extrinsic compression of the left main coronary artery (LMCA) by the pulmonary artery.

**CASE REPORT**

A 31-year-old woman with no relevant medical history nor modifiable cardiovascular risk factors, whose primary clinical manifestation was a 3-year history of progressive dyspnea triggered by even moderate exertion. She also had a syncopal episode and symptoms consistent with angina on moderate exertion. On examination, the most notable finding was the presence of an accentuated pulmonic component of the second heart sound.

The electrocardiogram revealed sinus rhythm with the QRS axis at 120°, right bundle branch block, and signs of right ventricular hypertrophy. Chest x-ray showed cardiomegaly involving the right cavities, with a notable enlargement of the pulmonary arch. Standard laboratory workup—including pulmonary function testing and screening for systemic diseases and those related to human immunodeficiency virus (HIV)—was normal. The echocardiogram—although quality images could not be obtained because the patient was a poor candidate for echocardiography—revealed a nondilated left ventricle.
with preserved systolic function, a significant dilation of the right cavities, and paradoxical septal motion. The valvular apparatus was normal and the maximum pulmonary artery systolic pressure as measured by Doppler was 88 mm Hg. No pulmonary embolism was evident on computed tomographic angiography, although severe dilation of the pulmonary artery and its principal branches was found. The diagnosis was suspected primary pulmonary hypertension. A hemodynamic and angiographic study was indicated to complete the examination and definitively rule out the presence of shunts. The mean right atrial pressure was 18 mm Hg; pulmonary artery pressure was 82/34 mm Hg (mean, 49 mm Hg); pulmonary capillary wedge pressure, 9 mm Hg, with a cardiac index of 2.0 L/min/m². Total pulmonary resistance was 1139 dyne.s.cm⁻⁵ (of which 921 was vascular). The presence of left-to-right shunting was ruled out by angiography. However, aortic angiography revealed a left coronary artery with a narrowed ostium that appeared stenotic. As a result, left coronary angiography was carried out, revealing 80% stenosis of the left coronary artery ostium in the form of a “pencil point” that progressively recovered its diameter (Figure 1A). The rest of the coronary tree was normal with no evidence of arteriosclerosis.

It was suspected that this stenosis was related to extrinsic compression by the dilated pulmonary artery. Magnetic resonance imaging (MRI) was performed and the images—although not clear—suggested extrinsic compression caused by the lower right part of the pulmonary artery (whose diameter was 4 cm) on the left coronary artery. Continuous subcutaneous epoprostenol infusion therapy was initiated via a portable infusion pump with reservoir. The patient’s symptoms partially improved and subjective improvement of the dyspnea was noted (this was not measured objectively, however). No further episodes of chest pain occurred during this time and, after the patient was stabilized 4 weeks later, a coated 3.5 mm×8 mm Taxus stent was directly implanted in the LMCA at 17 atm, with excellent angiographic results (Figure 1B) and no complications. Six months later, the patient was still undergoing continuous epoprostenol infusion therapy and, from a clinical standpoint, presented complete remission of his chest pains and an apparent subjective improvement of his dyspnea.

**DISCUSSION**

Angina is a documented symptom of PPH affecting approximately 40% of patients.¹ Its etiology is not entirely clear. It is believed that one of its mechanisms may be related to subendocardial ischemia of the right ventricular wall caused by intramyocardial compression of the arterioles or a decrease in the coronary perfusion gradient due to an increase in right atrial pressure.² Alternatively, it has been suggested that the cause of these pains is the acute dilation of the pulmonary artery during transitory increases in pulmonary artery pressure.³ No study demonstrating a relation between angina in these patients and the presence of myocardial ischemia has been published. Moreover, coronary angiography is not systematically applied to the management of PPH because patients are generally young and without risk factors, with a low prevalence of coronary arteriosclerosis; in our
coronary artery and Eisenmenger syndrome had been associated with the anomalous origin of the left pulmonary artery might explain the symptoms of pulmonary hypertension. In 1997, the first case of extrinsic compression of the LMCA in a PPH patient (its association with the anomalous origin of the left coronary artery and Eisenmenger syndrome had been described previously) was reported.6-12 After an exhaustive review of the literature, we found only 6 reported cases in which extrinsic compression of the LMCA associated with PPH had been described.6,13-16 All of the patients, as in many others with pulmonary hypertension, had symptoms of chest pain consistent with angina. The diagnosis of coronary artery compression was made in all cases by the hemodynamic-coronary angiography study that these patients undergo to quantify and evaluate the etiology of pulmonary hypertension. In none of these patients was a coronary angiography indicated based on the existence of prior myocardial ischemia studies. The extrinsic compression of the LMCA—which can occur in patients with PPH and those with congenital heart disease presenting with pulmonary hypertension—produces an unusual narrowing of the coronary ostium with progressive distal recovery of the diameter while the rest of the coronary circulation remains normal.13,16 Few cases have been reported and even then LMCA compression was—to an extent—often an incidental finding; however, some authors believe that the incidence of this functional obstruction in PPH patients with angina may be underestimated due to the postmortem collapse experienced by the blood vessels. In 1 case, a PPH patient with clear angiographic signs of LMCA stenosis underwent a heart and lung transplant and no macroscopic signs of coronary disease were found anywhere.6 Likewise, because extrinsic compression of the coronary artery is treatable, some authors believe that a coronary angiography should be performed on all PPH patients presenting with exertional angina or left ventricular dysfunction.13 It is important to keep in mind that PPH is a complex, difficult to manage disease with a poor prognosis; the reported incidence of angina, left ventricular dysfunction, and sudden death is 41%, 20%, and 26%, respectively.1,4,17 Any of these manifestations could have a physiopathologic basis in the impairment of the LMCA. MRI is an emerging technique that has proven useful in evaluating this type of patient, although in some cases, such as ours, it is not diagnostic due to its limited sensitivity.14,18

The therapeutic approach to this problem is based on a very limited number of case histories. Surgical mortality in PPH patients who undergo coronary revascularization surgery is very high, in large part due to postoperative right ventricular failure.39 Moreover, unprotected LMCA stenting has been associated with significant morbidity, although it can be safely performed in high-risk patients at experienced hospitals.20-22 In 2001, Rich et al13 reported the first—and only—2 cases of patients with PPH and extrinsic compression of the LMCA treated by percutaneous insertion of an intracoronary stent. As in our case, the angiographic result was very good, with no intraoperative complications. It should be noted that 1 of the patients described by these authors had severe left ventricular systolic dysfunction (ejection fraction <0.25), which normalized after the procedure. Therefore, myocardial ischemia may be an etiological mechanism of left ventricular systolic dysfunction when extrinsic compression of the LMCA is present. Prior to the intervention, both patients were administered continuous intravenous epoprostenol infusion therapy for 30 days to achieve clinical stabilization.

Based on the case histories described above, we also opted for stenting (postponing the intervention for 30 days after initiation of intravenous epoprostenol treatment) when presented with a patient with chest pain without other signs of ischemia and a documented 80% stenosis of the left coronary ostium. The patient’s signs and symptoms improved, although it is notable that no apparent change in her mean pulmonary artery pressure was found in the 2 hemodynamic studies performed. Moreover, an MRI revealed no perceptible change in the diameter of the pulmonary artery; nor was any change evident in the angiographic morphology of the coronary stenosis caused by the compression. This suggests to us that the results of long-term treatment with epoprostenol alone are unlikely to match those of surgical intervention. On the other hand, because the patient continued to receive chronic continuous infusion of epoprostenol—an agent with a proven beneficial effect on symptoms and prognosis—it is difficult to precisely identify the role played by LMCA stenting in the long-term sustained improvement experienced by this patient.23

However, we believed the surgical insertion of a stent in the LMCA was the best option for this “symptomatic” patient, basing our decision on the small body of literature available as well as our center’s extensive experience in coronary surgery. It should be noted that the optimal treatment for PPH patients with extrinsic LMCA compression has not been clearly established; it is not known whether the functional and prognostic significance in these cases is

Gómez Varela S, et al. Stenting in Primary Pulmonary Hypertension With Compression of the Left Main Coronary Artery

Rev Esp Cardiol 2004;57(7):695-8 697
similar to that for patients with occlusive arteriosclerosis involving the LMCA.

However, as occurred with our patient, the angina may be alleviated by stent revascularization, but no data is available on the long-term benefits of this technique. The long-term impact of treatment with epoprostenol or endothelin inhibitors in this type of coronary impairment is also unknown.

CONCLUSIONS

Extrinsic compression of the LMCA is a treatable cause of angina in patients with PPH. Although coronary angiography is not part of the diagnostic protocol, some authors recommend it be added to the hemodynamic study of PPH patients with symptoms of exertional angina or left ventricular systolic dysfunction. Stenting of the LMCA can be carried out with good angiographic results at experienced centers, although too few case histories are available to ascertain its impact on the prognosis of these patients.

REFERENCES