Percutaneous Coronary Intervention for Left Main Compression Syndrome due to Severe Idiopathic Pulmonary Arterial Hypertension: One Year Follow-up Using Intravascular Imaging

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Abstract

A 60-year-old woman presented with ST-elevation myocardial infarction due to extrinsic compression of the left main coronary artery (LMCA) caused by a dilated pulmonary artery (PA) with idiopathic pulmonary hypertension and was successfully treated with intravascular ultrasound- and optical coherence tomography-guided stenting. Continuous subcutaneous epoprostenol infusion therapy was initiated immediately after the procedure and increased aggressively. Imaging modalities were extremely useful in making the diagnosis and providing follow-up of LMCA compression syndrome in this case. Over the one-year observation period, a sufficient hemodynamic improvement was obtained, without exacerbation of the PA dilatation, resulting in the absence of compression of the LMCA.

Key words: coronary artery stenting, left main coronary artery compression, pulmonary hypertension, intravascular imaging

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Introduction

Left main coronary artery (LMCA) disease may occur in association with the development of a dilated pulmonary artery (PA) in patients with severe pulmonary hypertension (PH) (1). Extrinsic compression of the LMCA by the dilated PA results in myocardial ischemia, infarction or sudden cardiac death (1). This report describes the case of a patient with severe idiopathic pulmonary arterial hypertension (IPAH) who presented with ST-elevation acute myocardial infarction (STEMI) due to extrinsic compression of the LMCA by a dilated PA. She subsequently underwent emergency coronary angiography (CAG) and percutaneous coronary intervention (PCI) of the LMCA, and continuous epoprostenol infusion was started immediately after the procedure. Her symptoms and hemodynamics dramatically improved, and intravascular imaging confirmed stent patency up to one year later.

Case Report

A 60-year-old woman was hospitalized for sustained chest pain and loss of consciousness in June 2011. At 42 years of age in 1993, she had undergone DDD pacemaker implantation for sick sinus syndrome. At 57 years of age in 2009, she received a detailed examination for progressive dyspnea triggered by moderate exertion. At that time, right heart catheterization (RHC) showed a systolic/diastolic/mean PA pressure of 73/26/46 mmHg, pulmonary arterial wedge pressure of 7 mmHg and pulmonary vascular resistance (PVR) of 725 dynes·sec·cm⁻⁵. She was therefore diagnosed with IPAH based on the diagnostic algorithm and treated with bosentan. She remained in stable condition with a WHO functional class II status, although transthoracic echocardiography revealed a dilated and hypertrophied right ventricle and the estimated right ventricle systolic pressure remained over 90 mmHg at her monthly clinic visits.
In the beginning of 2011, the patient’s symptoms suddenly became exacerbated. She was thus diagnosed with progressive PH symptoms and newly prescribed sildenafil for PH therapy intensification; however, this treatment was not effective. The most notable findings of a physical examination performed on admission were the presence of an accentuated pulmonic component of the second heart sound and a third heart sound at Erb’s point. Her blood pressure was decreased to 80 over 30, and her heart rate was increased at 130/min. An electrocardiogram revealed a sinus rhythm with the QRS axis at 120°, right bundle branch block and ST elevation in leads aVR and V1-2. A chest X-ray showed notable enlargement of the pulmonary arch, and the laboratory data revealed elevation of the white blood cell count (10,320/μL) and CK (1,293 IU/L) and CK-MB (113 IU/L) levels. A computed tomography (CT) scan obtained for one year. RHC performed after the one-year follow-up visit. The PA diameter in the present case was 7.6 cm, impinging upon the origin of the LMCA (Fig. 1A). In the emergency room, the patient appeared severely distressed, with ongoing chest pain and vomiting. She was therefore diagnosed with STEMI presenting with a Killip class IV status and underwent emergency CAG. The CAG procedure showed 90% stenosis in the ostium of the LMCA with a thrombolysis in myocardial infarction trial (TIMI) grade 3 flow (Fig. 2A); however, no other sites of coronary artery stenosis were noted. Intravascular ultrasound (IVUS) and optical coherence tomography (OCT) disclosed that the LMCA ostium was deformed, with a slit-like narrowing appearance, suggestive of compression (Fig. 3A, D), with no atheroma throughout the LMCA. She immediately underwent PCI and a 3.5*22-mm bare metal stent was implanted in the LMCA (Fig. 2B) with intra aortic balloon pumping (IABP) support. Subsequently, IVUS and OCT revealed adequate stent expansion in a circle without malapposition, resulting in the complete resolution of the LMCA stenosis (Fig. 3B, E). After PCI, RHC showed a systolic/diastolic mean PA pressure of 97/62/76 mmHg (systemic arterial pressure of 85 mmHg) and PVR of 1,710 dynes·sec·cm⁻5 (Table). In addition, the patient’s chest pain resolved promptly after the PCI procedure. Continuous epoprostenol infusion therapy was subsequently initiated on day 1 of hospital admission. The dose of epoprostenol was slowly increased until the patient became hemodynamically stable, and she was successfully weaned off intra-aortic balloon pumping and catecholamine support five days after admission. At discharge, 37 days after admission, RHC showed a systolic/diastolic mean PA pressure of 65/30/42 mmHg and PVR of 494 dynes·sec·cm⁻5 under the infusion of 21.8 ng/kg/min of epoprostenol (Table). The dose of epoprostenol was therefore increased up to 42.4 ng/kg/min at the outpatient care clinic, and the patient went about her usual daily life, without any symptoms, in a WHO functional class I status for one year. RHC performed after the one-year follow-up showed a systolic/diastolic mean PA pressure of 42/20/31 mmHg and PVR of 433 dynes·sec·cm⁻5 (Table), CAG, IVUS, OCT and CT revealed a thoroughly expanded and apposed stent without compression (Fig. 1B, 2C, 3C, 3F). In addition, the main PA diameter was not increased at the one-year follow-up visit.

**Discussion**

LMCA compression syndrome occurs at a frequency of 5% to 19% among patients with PH (2, 3). Extrinsic LMCA compression by an enlarged PA should be suspected in patients with PH who present with angina. This syndrome is primarily seen in patients with idiopathic PH or PH due to uncorrected (or incompletely corrected) CHD (1). Mesquita et al. reported that compression of the LMCA occurs at a PA diameter of >4.0 cm or PA-to-aortic diameter ratio of >1.2 (3). The PA diameter in the present case was 7.6 cm, with a PA-to-aortic diameter ratio of 2.4.
The treatment of symptomatic extrinsic compression of the LMCA remains controversial. Coronary artery bypass graft surgery and unprotected LMCA stent implantation are currently the only available revascularization strategies (4, 5). Due to the high rate of surgical mortality among patients with PH, LMCA stenting is usually favored for revascularization strategy (6). Recently, several papers have described the use of LMCA stenting in patients with LMCA compression induced by a dilated PA, with excellent early post-procedural and short-term outcomes (1, 7, 8). However, there are few reports regarding the application of intravascular imaging for follow-up and the detailed medical condition of PH after the procedure. To the best of our knowledge, this report provides the first proof indicating the feasi-

Figure 2. Coronary angiograms obtained (A) before (B), after and (C) one year after treatment. (A) A long, smooth area of 90% stenosis in the LMCA ostium without further evidence of coronary artery disease. (B) Percutaneous coronary intervention of the unprotected LMCA with direct stenting resulted in the complete restoration of the patency of the LMCA. (C) At the one-year follow-up visit, there was no restenosis in the LMCA. *LMCA indicates the left main coronary artery.

Figure 3. Intravascular ultrasound and optical coherence tomography images (each slice obtained at the same time table on IVUS and OCT are of the same segments). IVUS and OCT showed a deformity of the ostial LMCA, with slit-like narrowing, suggesting the presence of extrinsic compression before stenting (A, D). Optimal stent expansion in a circle without compression after LMCA stenting (B, E). A thoroughly expanded and apposed stent without compression one year later (C, F). IVUS: intravascular ultrasonography, OCT: optical coherence tomography, LMCA: left main coronary artery.
bility and therapeutic effectiveness of LMCA stenting, with detailed intravascular imaging follow-up findings and the use of long-term continuous epoprostenol infusion for IPAH. Since compression of the LMCA was caused by a dilated PA in association with PH in this case, it was necessary to reduce the PA pressure in order to decrease the likelihood of extrinsic compression on the LMCA, and thus prevent adverse events. Although there are some reports of the use of PAH-target drugs [e.g., bosentan (9), sildenafil (7, 10) and epoprostenol (11, 12)] after revascularization, treatment with these agents did not obtain a sufficient improvement in hemodynamics. In the current case, continuous epoprostenol infusion therapy was started immediately after PCI and the dose of epoprostenol was increased aggressively, as shown in the Table. Consequently, a sufficient hemodynamic improvement was obtained without exacerbation of the PA dilatation for up to one year, with no repeat compression of the LMCA. Our experience suggests that controlling PH is an important factor for preventing restenosis after LMCA stenting in patients with IPAH.

We experienced a case of STEMI resulting from extrinsic compression of the LMCA by a dilated PA associated with IPAH, without recurrence of adverse cardiac events for more than one year. Intravascular imaging modalities, such as IVUS and OCT, are extremely useful for making the diagnosis of LMCA compression syndrome and providing follow-up to monitor the therapeutic outcome. Intracoronary stent implantation should be considered in the acute phase to preserve the coronary flow under extrinsic compression. Moreover, achieving aggressive reduction of the PA pressure appears to be an important factor for preventing adverse events in the chronic phase in patients with PH who undergo LMCA stenting for LMCA compression syndrome.

The authors state that they have no Conflict of Interest (COI).

References