Left main coronary artery occlusion by external compression with a large pulmonary artery in Eisenmenger syndrome

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Introduction

The compression of the left main coronary artery (LMCA) with a dilated pulmonary artery (PA) is an important and life-threatening issue, possibly related to sudden death in patients with advanced pulmonary arterial hypertension (PAH). Here we present an exceptional and extreme case with an LMCA occlusion, who fortunately survived due to well-developed coronary collateral retrograde flow.

Case Report

A 39-year-old woman suffering from exertional dyspnea and frequent anginal episodes was referred to our center. PAH, secondary to unoperated patent ductus arteriosus, had been diagnosed 13 years before, and the patient was on a triple-combination therapy (bosentan, tadalafil, and inhaled iloprost). The clinical presentation involving cyanosis and clubbing jugular vein distension, loud S2P2, and parasternal lift was concordant with Eisenmenger's syndrome. Electrocardiography indicated the right axis deviation, right ventricular strain pattern and anterolateral ischemia was suspected (Fig. 1a). Echocardiography revealed a dilated PA and right chambers with a leftward septal shift (Videos 1 and 2), and the estimated PA systolic pressure was 110 mm Hg. The LMCA compression was suspected. Computed tomography showed a complete LMCA occlusion due to external compression by the PA aneurysm (Fig. 1b and 1c). Cardiac catheterization revealed an advanced PAH (mean PA pressure, 88 mm Hg; pulmonary vascular resistance, 22.2 woods unit; cardiac index, 1.6 L/min/m^2). Aortagraphy and selective coronary angiography confirmed the LMCA occlusion (Fig. 1d and 1e, Videos 3 and 4). Coronary angiography also proved that the circumflex coronary artery originated from the right sinus of Valsalva, and the patient survived because of an efficient retrograde flow from the circumflex and right coronary arteries (Fig. 1f and 1g; Videos 5 and 6). Coronary by-pass surgery and PA aneurysm repair was offered. However, the patient refused surgery, and it was decided to switch the inhaled iloprost to parenteral prostanoid therapy.

Discussion

Sudden cardiac death is a common mode of mortality in patients with PAH, and it may be related with mechanical complications (1). The severe LMCA compression rate was 8.2% in a recent study and required intervention in most of the patients (2). The LMCA compression risk is related to the PA diameter. When the PA diameter exceeds 40 mm, it is defined as a PA aneurysm (3). PA aneurysms are mostly located in the main PA and can cause symptoms due to compression of adjacent tissues. Idiopathic, iatrogenic, infectious or connective tissue diseases, and vasculitis can cause a PA aneurysm, but the majority of the cases are related with congenital heart disease and PAH (3). As the LMCA compression can exist without anginal symptoms, physicians should be aware of the sudden death risk, and computed tomography should be performed to define the LMCA compression in patients at risk; particularly if the PA diameter is over 40 mm, ensuring the existence of a PA aneurysm (1, 2). Recent evidence indicates the coronary stenting as a viable option for resolution of LMCA compression (2). But in this case, a complete obstruction of LMCA was not amenable to stenting, so a surgical intervention was the only option. In addition to the mechanical compression of LMCA, considering the potentially mortal pulmonary rupture and dissection risk, our decision was also concordant with current recommendations for the management of PA aneurysms (3).

Conclusion

A marked dilatation of PA in patients with PAH may cause symptoms due to the compression of adjacent tissues, and particularly the LMCA compression may expose them to the risk of sudden

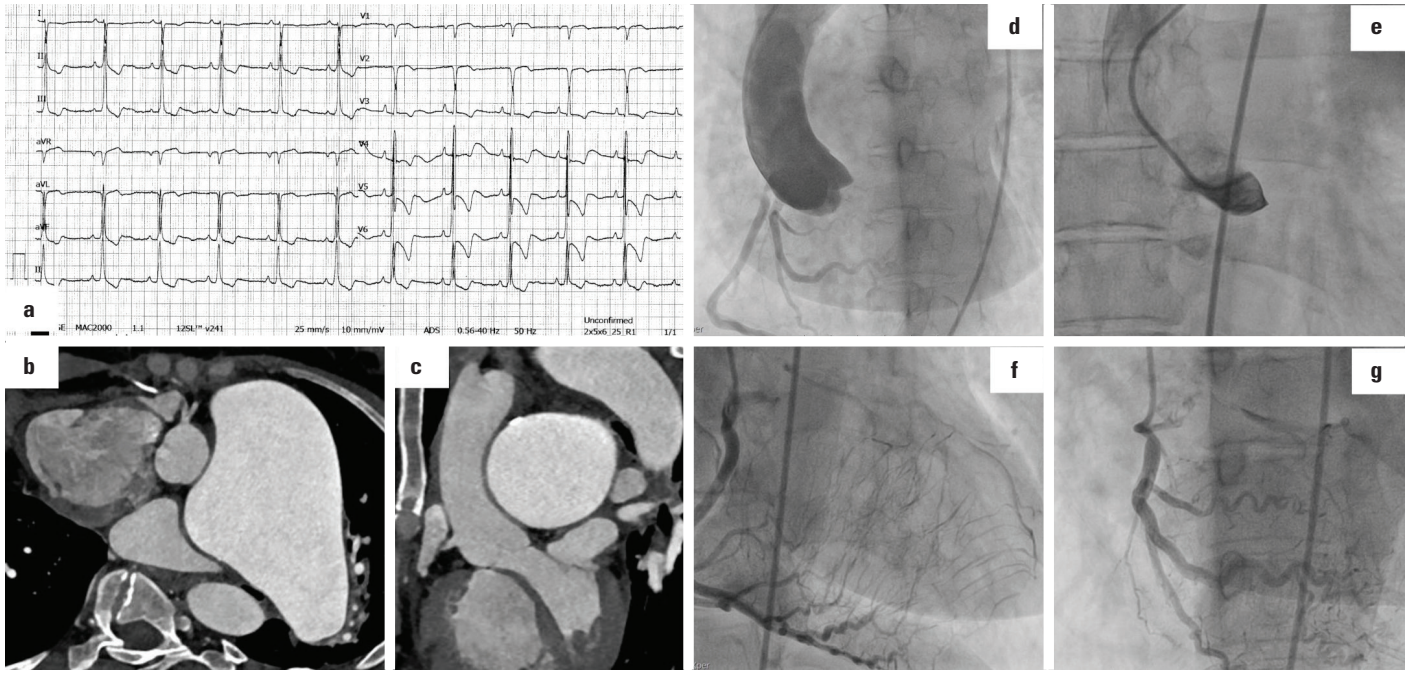

Figure 1. Beyond an apparent right ventricular strain pattern in inferior leads, electrocardiography (a) reveals a symmetrical deep T-wave inversion in lateral derivations arousing the suspicion of ischemia. Also, a mild ST elevation in aVR is notable, indicating the LMCA disease. An axial computed tomography (CT) image (b) depicts a huge (65 mm) pulmonary artery aneurysm compressing the LMCA, while the right and anomalous circumflex coronary arteries originate from the right sinus of Valsalva. The sagittal CT image (c) confirms an obliteration of LMCA by the pulmonary artery aneurysm. Aortagraphy (d) shows the right coronary artery and the right take-off circumflex coronary artery, but LMCA is not opacified during aortagraphy. Selective engagement attempts fail due to compression, but a small tip of the origin of the LMCA can be visible (e). Selective angiography of the right (f) and anomalous circumflex (g) coronary arteries depicts an efficient retrograde filling of the left anterior descending coronary artery. The retrograde flow reaches almost to the LMCA trunk

cardiac death. Physicians should consider this issue and as the LMCA compression can exist without anginal symptoms, computed tomography should be utilized liberally to define the LMCA compression in patients at risk, especially if the PA diameter is >40 mm.

Informed consent: An informed consent was obtained from the patient.

Video 1. Transthoracic echocardiography in the parasternal short axis view depicts massive dilatation (65 mm) of main pulmonary artery

Video 2. Transthoracic echocardiography in parasternal short axis view shows a dilated right ventricle and D-shaped septum indicating severe pulmonary hypertension

Video 3. Aortagraphy shows a complete occlusion of LMCA due to external compression by the PA aneurysm. The circumflex coronary artery arising from the right sinus of Valsalva is noted. Late opacification of the left anterior descending coronary artery is evident via retrograde collateral flow

Video 4. Selective engagement of the left main coronary artery could not be achieved because of the external compression by pulmonary aneurysm. The origin of the compressed left main coronary artery is observed as a tiny tip in the left sinus of Valsalva

Video 5. Selective right coronary angiography indicates prominent retrograde collateral flow to the left anterior descending coronary artery

Video 6. The circumflex coronary artery was abnormally originating from the right sinus of Valsalva and selective angiography reveals collateral flow to the left coronary artery almost reaching to the left main ostium

References

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