Heritable Pulmonary Arterial Hypertension With Elevated Pulmonary Wedge Pressure

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A 50-year-old man, who is an active smoker without a previous medical history, was admitted for recent dyspnea (New York Heart Association functional class III) and an episode of exertional syncope. His familial history included the sudden death of his mother at the age of 50. His blood pressure was 123/90 mm Hg; heart rate, 80 beats/min; peripheral oxygen saturation, 95%; and clinical examination found a prominent pulmonary component of S2, jugular vein distension, and normal lung sounds. His ECG showed complete right bundle-branch block. Chest radiography showed central pulmonary artery, right atrium, and ventricle enlargements without major abnormalities of pulmonary parenchyma (Figure 1). Brain natriuretic peptide was 758 pg/mL (normal, <100 pg/mL) and troponin T was raised to 0.22 ng/mL (normal, ≤0.14 ng/mL).

Transsthoracic echocardiography showed severe dilatation of the right chambers, paradoxical wall motion septum, and pulmonary hypertension with a tricuspid regurgitant jet velocity of 4.5 m/s and an estimated systolic pulmonary arterial pressure of 96 mm Hg (Figure 2). Left ventricle, aortic and mitral valves, and left ventricle filling pressures were normal. High-resolution computed tomography of the chest showed neither acute nor chronic thromboembolic disease, nor severe parenchymal lung disease; in particular, radiological signs of pulmonary veno-occlusive disease were absent. The different causes of precapillary pulmonary hypertension were excluded by exhaustive investigations, including pulmonary function tests, ventilation/perfusion lung scan, antinuclear antibodies, HIV and hepatitis serologies, and abdominal ultrasound.

Right heart catheterization was then performed to confirm the diagnosis of precapillary pulmonary hypertension; systolic and diastolic pulmonary arterial pressure were, respectively, 100 mm Hg and 48 mm Hg, with a mean pulmonary arterial pressure of 67 mm Hg. The assessment of pulmonary wedge pressure (PWP) was difficult technically because of the severity of pulmonary hypertension and was achievable only with the tip of the balloon catheter located in the right lower lobe (West zone 3), but not in the right upper lobe or in the left pulmonary arteries (Figure 3). A complete wedging of the balloon catheter was then obtained with evidence of distinct A and V waves, allowing correct measurements of PWP at the end of expiration. Surprisingly, we measured increased PWP at 37 mm Hg, suggesting postcapillary pulmonary hypertension. Because of this discrepancy between the value of PWP and the clinical and echocardiographic records, we decided to perform concomitantly a left ventricular catheterization to measure left ventricular end diastolic pressure, which was normal at 7 mm Hg (Figures 4 and 5), confirming precapillary pulmonary hypertension.

Cardiac computed tomography was then performed and showed major dilatation of the right chambers, with an enlarged right atrium causing compression of the right inferior pulmonary vein (Figures 6 and 7). This extrinsic compression explained the falsely elevated PWP in this patient with authentic pulmonary arterial hypertension. Interestingly, genetic testing was done in this patient, who...
had a family history of pulmonary arterial hypertension, and a BMPR2 gene mutation was identified. As demonstrated previously,\(^1\,2\) heritable pulmonary arterial hypertension is associated with a poor outcome, and the patient died suddenly 5 days later before any specific treatment could be started.

To our knowledge, we report here the first case illustrating the possible discrepancy between PWP and left ventricular end diastolic pressure resulting from pulmonary vein compression by important dilatation of the right atrium in a patient with severe, heritable pulmonary arterial hypertension. Because true diagnosis is crucial for the treatment strategy, this case report shows the potential complementary role of left ventricular catheterization,\(^3\) performed concomitantly with right heart catheterization, in the assessment of pulmonary hypertension, especially in difficult cases.

Figure 2. In the parasternal long-axis view (A), transthoracic echocardiography shows paradoxical wall motion septum and a severe dilatation of the right ventricle (RV) and right atrium (RA) resulting from pulmonary hypertension, causing compression of the left ventricle (LV) and left atrium (LA). Systolic pulmonary artery pressure was assessed using peak velocity of tricuspid flow regurgitation with continuous-wave Doppler (B). Right atrial pressure was estimated at 15 mm Hg because of a noncompressible dilated inferior vena cava. Thus, systolic pulmonary arterial pressure was calculated at 96 mm Hg.

Disclosures

None.

References

Figure 3. Chest radiograph during right heart catheterization. The assessment of pulmonary wedge pressure with a complete wedging of the balloon catheter was achievable only in the right lower lobe. Pulmonary wedge pressure was increased at 37 mm Hg.

Figure 4. Concomitant left ventricle pressure and pulmonary artery pressure curves. Right heart catheterization showed pulmonary hypertension, with pulmonary arterial pressure estimated at 100/48 mm Hg. Left ventricle catheterization found a normal left ventricular end diastolic pressure (LVEDP) at 7 mm Hg.

Figure 5. Concomitant left ventricular catheterization and pulmonary wedge pressure assessment confirmed precapillary pulmonary hypertension by showing normal left ventricular end diastolic pressure (LVEDP) at 7 mm Hg, whereas pulmonary wedge pressure was increased at 37 mm Hg. LV indicates left ventricle; PWP, pulmonary wedge pressure.

Figure 6. Cardiac computed tomography showed huge dilation of right chambers and extrinsic compression of the right inferior pulmonary vein by the right atrium, explaining the falsely elevated pulmonary wedge pressure. LA indicates left atrium; LV, left ventricle; RA, right atrium; and RV, right ventricle.
Figure 7. Cardiac computed tomography with 3-dimensional volume rendering: superior views (A, B), inferior view (C), and frontal view after fusion volume rendering with a 2-dimensional reformat (D). The ostium of the right inferior pulmonary vein (RIPV) was compressed (white arrow), with a notch aspect, by the huge dilatation of the right atrium. Of note, the right superior pulmonary vein (RSPV) was also compressed by the right atrium. LA indicates left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.
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