


Coronary artery fistula as major source of right lung circulation in a patient with isolated right pulmonary artery agenesis

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A 36-year-old patient was sent for diagnostic work-up of dry cough, chest tightness, and 2 years of increasing dyspnoea. Previously, the patient was physically well and only known for an asymptomatic heart murmur since childhood. A grade III/VI systolic murmur was heard at the left sternal border. Electrocardiogram revealed right ventricular hypertrophy and occasional atrial premature beats. Chest radiograph revealed prominent left pulmonary trunk with increased vascular markings (Panel A), and echocardiography showed enlargement of right atrium and ventricle, moderate tricuspid regurgitation, and pulmonary hypertension (estimated pulmonary pressure 85 mmHg). Cardiac magnetic resonance imaging suspected right pulmonary artery agenesis. We then performed cardiac catheterization for pulmonary hypertension workup. Pulmonary artery pressure was elevated (84/19/46) and angiography showed a total absence of right pulmonary artery (RPA) with left pulmonary artery (LPA) engorgement (Panel B). Incidentally, we found a tortuous coronary fistula raising from the right coronary artery (RCA) supplying the right lung field (Panels C and D). To reveal a possible coronary steal, we suggested further stress study by Treadmill exercise test but the patient refused and was treated with oral nifedipine for control of pulmonary hypertension thereafter. Her estimated pulmonary pressure was the same during half-year follow-up, and her symptoms were stationary.

In patients with pulmonary atresia with ventricular septal defect, unilateral pulmonary artery hypoplasia with collateral blood flow, mostly from systemic and rarely from coronary supplying the corresponding lung territory, is common. The anomaly was thought to result from early recanalization of coronary artery to bronchopulmonary anastomoses due to regional reduction in pulmonary blood flow. Here, we report a rare case of isolated RPA agenesis with right lung blood supply mainly from RCA possibly through the same embryologic mechanism.

Supplementary material is available at European Heart Journal online.

Panel A. Chest radiograph showing engorged left pulmonary trunk with increased infiltrates.

Panel B. Main pulmonary artery angiography showed absence of right pulmonary artery with engorged left pulmonary artery.

Panels C and D. Right coronary angiography showed tortuous coronary artery fistula (arrows) supplied right lung field (right anterior oblique and left anterior oblique view).