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IMAGE

A Case of Pulmonary Hypertension Due to Fistulas Between Multiple Systemic Arteries and the Right Pulmonary Artery in an Adult Discovered for Occulted Dyspnoea

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Background Pulmonary hypertension (PH) can be caused by a fistula between the systemic and pulmonary arteries. Here, we report a case of PH due to multiple fistulas between systemic arteries and the right pulmonary artery where the ventilation/perfusion scan showed no perfusion in the right lung.

Methods A 32-year-old male patient was hospitalised for community-acquired pneumonia. After treatment with antibiotics, the pneumonia was alleviated but dyspnoea persisted. Pulmonary hypertension was diagnosed using right heart catheterisation, which detected the mean pulmonary artery pressure as 37 mmHg. The anomalies were confirmed by contrast-enhanced CT scan (CT pulmonary angiography), systemic arterial angiography and pulmonary angiography.

Results Following embolisation of the largest fistula, the haemodynamics and oxygen dynamics did not improve, and even worsened to some extent. After supportive therapy including diuretics and oxygen, the patient's dyspnoea, WHO function class and right heart function by transthoracic echocardiography all improved during follow-up.

Conclusions Pulmonary hypertension can be present even when the right lung perfusion is lost. Closure of fistulas by embolisation, when those fistulas act as the proliferating vessels, may be harmful.

Keywords Pulmonary hypertension • Fistula • Dyspnoea • Embolisation • Diuretics

Abbreviations: PH, pulmonary hypertension; RHC, right heart cathererisation; mPAP, mean pulmonary artery pressure; sPAP, systolic pulmonary artery pressure; CTPA, CT pulmonary angiography; PA, pulmonary angiography; PO_2, partial pressure of oxygen; TAPSE, tricuspid annular plane systolic excursion

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Background

Pulmonary hypertension (PH) is a common complication of patients with fistulas which increase pulmonary artery blood flow. Here, we present a report of an adult case of PH due to multiple fistulas between systemic arteries and the right pulmonary artery for occulted dyspnoea whose ventilation/perfusion (V/Q) scan detected no perfusion of the right lung.

Case Report

A 32-year-old male was admitted to our hospital due to community-acquired pneumonia. After treatment, most symptoms disappeared except dyspnoea, which persisted. The physical examination showed a 2/6 systolic murmur in the tricuspid area. Pulmonary valve closure (P_2) sound > aortic valve closure (A_2) sound was heard. Arterial blood gas analysis showed that partial pressure of oxygen (PO_2) was 66 mmHg. Transthoracic echocardiography detected tricuspid regurgitation velocity 3.88 m/s, and the estimated systolic pulmonary artery pressure (sPAP) was 70 mmHg. WHO Functional Class was evaluated as II.

Echocardiography, right heart contrast echocardiography, V/Q scan, lung function, CT pulmonary angiography (CTPA), and laboratory tests such as autoimmune antibodies were screened for the aetiology of PH. Transthoracic echocardiography showed a normal sized left ventricle and a relatively normal function of left ventricle [left ventricle ejection fraction (LVEF) = 68%], no regional wall motion abnormality, right ventricular (RV) was enlarged with a right ventricular end diastolic dimension of 52 mm, and medium RV systolic dysfunction [tricuspid annular plane systolic excursion (TAPSE) of 18.5 mm and Tei index = 0.93]. The right heart contrast echocardiography denied bypass within the heart. Ventilation/perfusion scan showed that ventilation was normal in the bilateral lung but no perfusion was detected in the right lung (Figure 1). Lung function demonstrated that FEV_1/FVC was 72.82% and FEV_1 of 84.8% after bronchodilator reversibility testing. Significantly, multiple fistulas between systemic arteries including several intercostal arteries, internal mammary artery and the right pulmonary artery were shown on CTPA (Figure 2). Laboratory results did not reveal signs of other diseases such as autoimmune disease and metabolic disease.

Pulmonary angiography (PA) and right heart catheterisation (RHC) was performed via the right jugular vein using a Swan-Ganz catheter. Systemic arterial angiography was performed via right femoral artery. Systemic arterial angiography and PA confirmed multiple fistulas between systemic arteries including several intercostal arteries, phrenic artery, internal mammary artery and the right pulmonary artery (Figure 3). Right heart catheterisation detected the mean pulmonary artery pressure (mPAP) as 37 mmHg and the PO_2 of systemic artery (femoral artery) as 71 mmHg while the PO_2 of right and left pulmonary artery as 67 and 39 mmHg respectively. The largest fistula, that is the fistula between the internal mammary artery and the right pulmonary artery, was embolised. After embolisation, the haemodynamics were re-evaluated. Right heart catheterisation detected the mPAP as 40 mmHg and the PO_2 of systemic artery as 68 mmHg while the PO_2 of right pulmonary artery as 72 mmHg (Table 1).

Given the supportive therapy including diuretics and oxygen, the patient was followed up regularly. One month later, the PO_2 of systemic artery (radial artery) was 82 mmHg without oxygen and the estimated sPAP was 68 mmHg and TAPSE and Tei index all improved by echocardiography. Three months and six months later, the estimated sPAP decreased gradually to 54 mmHg and TAPSE and Tei index all improved (Table 2). WHO Functional Class improved from II to I.

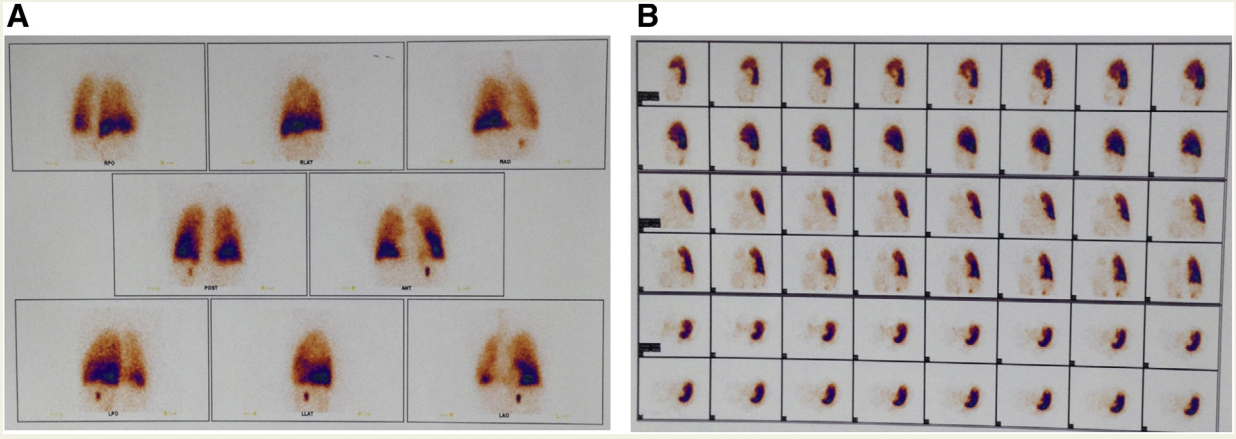

Figure 1 Ventilation and Perfusion scan (V/Q scan) showed no perfusion in the right lung.
A, Ventilation scan detects no defect in bilateral lung. B, Perfusion scan detects no perfusion in the right lung.

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