

Pulmonary Capillary Hemangiomatosis without Pulmonary Hypertension: An Early Stage of Disease?

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A 14-year-old male visited the First Affiliated Hospital of Guangzhou Medical University with a complaint of a 2-year history of progressive exertional dyspnea and fingertips cyanosis. Physical examination revealed remarkable desaturation measured by pulse oximetry (80% at rest) and marked cyanosis of lips. The high-resolution computed tomography scanning revealed no significant abnormality in the lung field [Figure 1a and 1b]. Computed tomography of pulmonary angiography showed no filling defect in trunk or branches of pulmonary arteries, no dilated central pulmonary arteries, and no cardiomegaly [Figure 1c and 1d]. Cardiac catheterization showed a mean right atrial pressure of 6 mmHg, pulmonary artery pressure with mean of 12 mmHg, pulmonary capillary wedge pressure of 9 mmHg, cardiac output of 6.2 L/min, and cardiac index of 4.10 L·min⁻¹·m⁻². Pulmonary vascular resistance was slightly elevated at 4.52 wood units. Lung biopsy revealed that lower lobe alveolar spaces were filled with red blood cells or hemosiderin cells, and alveolar septum was wider than normal. Hematoxylin and eosin staining shows that capillary proliferations within alveolar and bronchiolar walls were visible throughout most areas [Figure 2a-2c]. Immunostaining of lung section was strongly positive for CD34 [Figure 2d], CD31 [Figure 2e], and F8 [Figure 2f]. The clinical manifestations, pathological findings, and immunohistochemistry supported definitive diagnosis of pulmonary capillary hemangiomatosis (PCH) with normal pulmonary artery blood pressure and normal radiological appearance. Because of financial difficulties, the patient abandoned heart–lung transplantation and left hospital without therapeutic intervention. Follow-up to nearly a years, the patient was stable with no prominent hypoxia on 2 L/min supplemental nasal cannula oxygen at home. Written informed consent was obtained from the parents of patient for publication of this case report and any accompanying images.

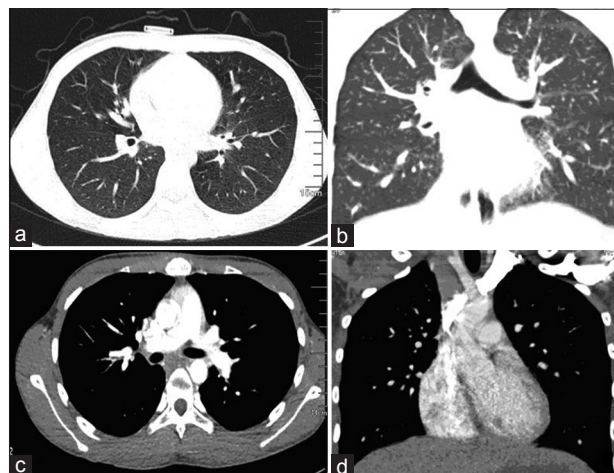


Figure 1: High-resolution computed tomography scanning revealed no significant abnormality (a and b) and computed tomography pulmonary angiography showed that no filling defect in trunk or branches of pulmonary arteries (c and d).

All patients with PCH usually have elevated pulmonary arterial pressures measured by right heart catheterization with normal or low pulmonary capillary wedge pressure and this have been demonstrated.^[1] The infiltration of pulmonary vessel and intrapulmonary shunting caused by uncontrolled proliferation of the pulmonary capillaries might be responsible for the pulmonary hypertension and

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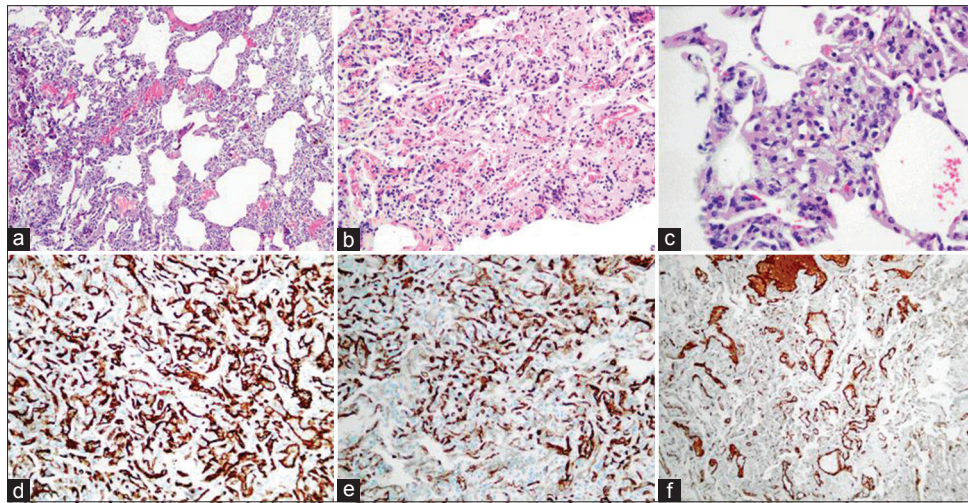


Figure 2: Microscopic examination of the lung section biopsy specimen showing numerous interlacing capillary channels filled with red blood cells within the lung interstitium (H and E, a: $\times 200$; b: $\times 200$; c: $\times 400$). Immunohistochemically, lung section showed proliferation of capillaries within the alveolar walls immunostaining with CD34, CD31, F8 (d: $\times 200$; e: $\times 200$; f: $\times 200$).

hypoxia.^[2] In our case, chest radiological findings did not show any specific changes of the disease and pulmonary artery pressure measured by right heart catheterization was normal, this fact urged us to proceed to the open lung biopsy. To our knowledge, although not all cases performed right heart catheterization, PCH without pulmonary hypertension has not been reported previously. One case reported that a 29-year-old man with PCH had been stable for 3.5 years since the diagnosis without symptoms of pulmonary hypertension but lack of cardiac catheterization data.^[3] While our case has been followed up for a year during which there has been no clinical deterioration, this might suggest that PCH is stable for a period of time as the patient did not have pulmonary hypertension, and this case might possibly be at an early phase of PCH. Therefore, the chest radiological findings matched the right heart catheterization findings. However, the prognosis of PCH is poor and the progressive nature is only 3 years from the time of initial clinical manifestations.^[4] Some cases demonstrated that clinical outcome might be quite variable even within a family,^[5] so the case should be followed up closely. Before performing lung biopsy, PCH should be differentiated from pulmonary veno-occlusive disease (PVOD), which is the potential cause of profound hypoxemia, and the notable clinical manifestation is also exertional dyspnea. PCH and PVOD are clinically indistinguishable especially when without obvious pulmonary hypertension and radiologic changes. Lung or heart–lung transplantation is the only treatment for this disease. More extensive understanding of PCH would resolve the therapeutic difficulties in the future.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parents have given consent for images and other clinical information to be reported in the journal. The patient's parents understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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