

Case Report

Pulmonary capillary hemangiomatosis: An unusual cause of primary pulmonary hypertension in a child with characteristic computed tomography imaging features

D Manjubashini, K Nagarajan, B Rajesh Kumar¹

Departments of Radiodiagnosis and ¹Pulmonary Medicine, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India

ABSTRACT

Pulmonary capillary hemangiomatosis (PCH) is a rare cause of primary pulmonary hypertension (PPH) diagnosed in children and young adults with a nonspecific clinical presentation of dyspnea, cough, chest pain, and fatigue. It is characterized by extensive proliferation of pulmonary capillaries within alveolar septa. The imaging features include diffuse centrilobular ground-glass opacities with features of pulmonary hypertension. We present a case of PCH in an 11-year-old boy who was diagnosed with PPH in echocardiography and referred for diagnostic imaging.

KEY WORDS: Multidetector computed tomography, primary pulmonary hypertension, pulmonary capillary hemangiomatosis

Address for correspondence: Dr. K Nagarajan, Department of Radiodiagnosis, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry - 600 506, India. E-mail: lknagarajan1@gmail.com

INTRODUCTION

Pulmonary capillary hemangiomatosis (PCH) is a rare disorder characterized by extensive alveolar capillary proliferation within the alveolar septa leading to the infiltration of thin-walled capillaries into the peribronchial and perivascular interstitium of the lung parenchyma and even the pleura.^[1-6] PCH is rare; and to the best of our knowledge, there are about 42 reported cases in literature.^[2-5] The disease has no sexual predilection,^[7] and it is diagnosed in children or young adults usually with nonspecific complaints such as dyspnea and cough. It presents with features similar to idiopathic pulmonary hypertension or pulmonary veno-occlusive disease (PVOD)-like features.^[5] The distinction of PCH or PVOD from idiopathic pulmonary arterial hypertension (IPAH) is important because pulmonary vasodilators may lead to deleterious complications in patients with PCH and PVOD, resulting

in hemoptysis and hemothorax. The cross-sectional computed tomography (CT) imaging features include diffuse centrilobular ground-glass opacities with features of pulmonary hypertension. The disease has poor prognosis, and lung transplantation is the only definitive treatment.^[3] Routine screening using multidetector computed tomography imaging of young-onset primary pulmonary hypertension (PPH) and the familiarity of this entity might increase the chance of identifying the diagnosis of PCH.

CASE REPORT

An 11-year-old boy presented to our hospital with 2 months' history of progressive exertional dyspnea. He

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Manjubashini D, Nagarajan K, Rajesh Kumar B. Pulmonary capillary hemangiomatosis: An unusual cause of primary pulmonary hypertension in a child with characteristic computed tomography imaging features. Lung India 2019;36:157-9.

Access this article online	
Quick Response Code: 	Website: www.lungindia.com
	DOI: 10.4103/lungindia.lungindia_122_18

was apparently healthy before 2 months. There was no history of fever, cough, wheezing, or hemoptysis. Physical examination was unremarkable. Chest auscultation revealed mild tachycardia. There were no signs of cardiac failure, no cardiac murmurs, and no adventitial sounds. Chest X-ray was taken which showed diffuse, fine reticulonodular opacities uniformly distributed in both the lung fields, and dilated main pulmonary artery [Figure 1]. There were no Kerley B lines or pleural effusion. The electrocardiography showed the right axis deviation and right ventricular hypertrophy with ST-T changes. He underwent echocardiography which showed the right ventricular hypertrophy and dilated main pulmonary trunk. The left ventricular systolic function, mitral valve, and pulmonary veins were normal. The estimated right ventricular systolic pressure was 90–100 mmHg. The right atrium was dilated. There was no other medical history of note and, in particular, no history of any chronic liver disease or hereditary hemorrhagic telangiectasia.

He was then referred for imaging evaluation of pulmonary vasculature. CT pulmonary angiogram was taken with arterial and venous phases, along with high-resolution CT of thorax which showed innumerable, randomly distributed tiny, and centrilobular ground-glass nodules in bilateral lung parenchyma [Figure 2]. These nodules

were measuring 6–8 mm in size. No lobar predilection was observed. The main pulmonary trunk was dilated with a luminal diameter of 3.5 cm [Figure 3]. The right and left branches of the pulmonary artery were also enlarged with peripheral pruning of the branches. Concentric hypertrophy of the right ventricle with bulging of the interventricular septum toward the left ventricle was observed along with enlarged right atrium features suggestive of pulmonary artery hypertension. There was a small 10-mm lymph node in the right paratracheal region which was considered insignificant [Figure 4]. There was no interstitial pulmonary fibrosis evident on imaging.

The patient had no other symptoms and remained clinically stable. The patient was followed up for few months as an outpatient. Pulmonary biopsy confirmation was suggested; however, the patient refused biopsy. As the patient was clinically stable and the imaging findings were strongly suggesting PCH, histological confirmation was



Figure 1: Chest radiograph posteroanterior view showing diffuse, fine reticulonodular opacities throughout bilateral lung fields, dilated right and left pulmonary arteries, and main pulmonary trunk

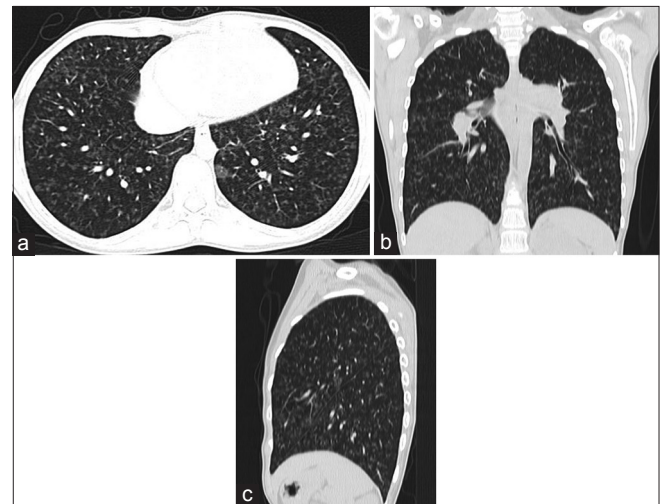


Figure 2: High-resolution computed tomography lung axial section (a) showing many, randomly scattered small, and centrilobular ground-glass nodules in bilateral lung parenchyma without lobar predilection. Coronal (b) and sagittal (c) reformations showing the same innumerable, tiny, and centrilobular ground-glass nodules

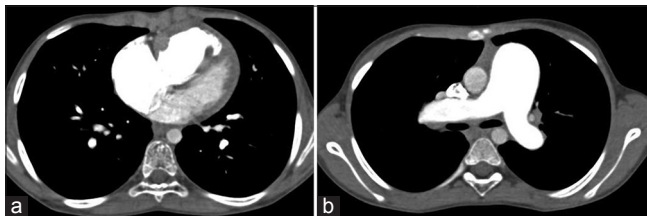


Figure 3: Computed tomography angiogram axial section at ventricular level (a) showing hypertrophy of the right ventricle with mild bulge of the interventricular septum toward the left ventricle. The right atrium also enlarged. Section (b) at higher level showing dilated main pulmonary trunk and right and left branches of the pulmonary arteries



Figure 4: Contrast computed tomography axial section showing a small solitary right lower paratracheal lymph node measuring about 10 mm in short axis

deferred. He was started on interferon- α and doxycycline as an angiogenesis inhibitor and is being followed-up on out-patient basis.

DISCUSSION

PCH, first described by Wagenvoort *et al.*, in 1978, is a rare cause of PPH.^[4] It is characterized by uncontrolled proliferation of small capillaries infiltrating the perivascular and bronchial interstitium. These small proliferating microvessels are prone to bleeding. When this occurs, it results in accumulation of hemosiderin-laden macrophages in alveolar spaces.^[6] The disease occurs sporadically, with few cases reported to have occurred in families.^[8] PCH is occasionally associated with other entities such as Takayasu arteritis,^[9] Kartagener's syndrome, systemic lupus erythematosus, and hypertrophic cardiomyopathy. Common clinical features of PCH are progressive dyspnea and features of pulmonary hypertension. Hemoptysis, pleural effusion, and right heart failure can rarely occur.^[3] Clinically, PCH is hard to distinguish from PVOD.^[1,5,8]

Characteristic CT findings in PCH include diffuse tiny and poorly circumscribed centrilobular nodular opacities. Scattered centrilobular opacities have wide differential diagnosis including atypical adenomatous hyperplasia, bronchioloalveolar carcinoma, pulmonary lymphoproliferative disorder, and organizing pneumonia with fibrosis.^[10] In the presence of pulmonary hypertension corroborated by echocardiography in a young patient, the differential diagnosis can be narrowed down to one of the pulmonary vascular etiologies – primary pulmonary arterial hypertension (PAH), PCH, and PVOD. Centrilobular nodules may be present in primary PAH as cholesterol granulomas.^[11-13] However, the findings of interlobular septal thickening, engorged lymphatics or lymphadenopathy, pulmonary edema, or alveolar hemorrhage-like features are not present in primary PAH.^[14] Raised pulmonary arterial pressures and normal/low pulmonary capillary wedge pressures are seen in both PCH and PVOD^[15] and may not help to differentiate among the two. Interstitial septal lines are present in both PCH and PVOD; however, they are more common in PVOD than PCH.^[10] Mediastinal lymphadenopathy may be seen in PCH rather than PVOD. As the imaging is characteristic in the background of clinical features and PAH, in most cases, there may not be a need for histopathological confirmation. It may be resorted to in patients with interstitial fibrosis in whom differentiation from PVOD becomes relatively difficult.

In contrast to IPAH, epoprostenol therapy is contraindicated in PCH which may show deleterious effects such as pulmonary edema and intrapulmonary bleeding.^[3,5] Symptomatic treatment with diuretics, oxygen, and warfarin has been used widely, and interferon α -2a has been proposed as specific therapy. Prognosis is poor with median survival of 3 years.^[6] Lung transplantation is the only definitive treatment.^[3]

In summary, PCH is a very rare cause of PPH diagnosed in children and young adults with nonspecific respiratory symptoms. Although imaging findings may mimic PVOD, the presence of centrilobular opacities alone as in our case makes the probability of PCH more likely and plan the management to avoid vasodilators which may worsen the outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients 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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Anderson TJ, Larsen ET, Wyse DG, Lester WM. Primary pulmonary hypertension for 30 years. *Am J Cardiol* 1991;68:284-5.
2. de Carvalho AC, Hovnanian AL, Fernandes CJ, Lapa M, Jardim C, Souza R, *et al.* Tadalafil as treatment for idiopathic pulmonary arterial hypertension. *Arq Bras Cardiol* 2006;87:e195-7.
3. O'Keefe MC, Post MD. Pulmonary capillary hemangiomas: A rare cause of pulmonary hypertension. *Arch Pathol Lab Med* 2015;139:274-7.
4. Wagenvoort CA, Beetstra A, Spijker J. Capillary haemangiomas of the lungs. *Histopathology* 1978;2:401-6.
5. Aston K, Riddell GJ, Sheppard MN, Wells AU, Riley MS. Pulmonary capillary haemangiomas – An unusual cause of hypoxia. *Respir Med Case Rep* 2012;7:12-4.
6. Almagro P, Julià J, Sanjaume M, González G, Casalots J, Heredia JL, *et al.* Pulmonary capillary hemangiomas associated with primary pulmonary hypertension: Report of 2 new cases and review of 35 cases from the literature. *Medicine (Baltimore)* 2002;81:417-24.
7. Eltorkey MA, Headley AS, Winer-Muram H, Garrett HE Jr, Griffin JP. Pulmonary capillary hemangiomas: A clinicopathologic review. *Ann Thorac Surg* 1994;57:772-6.
8. Langleben D, Heneghan JM, Batten AP, Wang NS, Fitch N, Schlesinger RD, *et al.* Familial pulmonary capillary hemangiomas resulting in primary pulmonary hypertension. *Ann Intern Med* 1988;109:106-9.
9. Savage BJ, Gupta RK, Angle J, Okusa MD. Takayasu arteritis presenting as a pulmonary-renal syndrome. *Am J Med Sci* 2003;325:275-81.
10. Kim HY, Shim YM, Lee KS, Han J, Yi CA, Kim YK, *et al.* Persistent pulmonary nodular ground-glass opacity at thin-section CT: Histopathologic comparisons. *Radiology* 2007;245:267-75.
11. Horton MR, Tuder RM. Primary pulmonary arterial hypertension presenting as diffuse micronodules on CT. *Crit Rev Comput Tomogr* 2004;45:335-41.
12. Nolan RL, McAdams HP, Sporn TA, Roggli VL, Tapson VF, Goodman PC, *et al.* Pulmonary cholesterol granulomas in patients with pulmonary artery hypertension: Chest radiographic and CT findings. *AJR Am J Roentgenol* 1999;172:1317-9.
13. Resten A, Maitre S, Humbert M, Sitbon O, Capron F, Simoneau G, *et al.* Pulmonary arterial hypertension: Thin-section CT predictors of epoprostenol therapy failure. *Radiology* 2002;222:782-8.
14. Chaisson NF, Dodson MW, Elliott CG. Pulmonary capillary hemangiomas and pulmonary veno-occlusive disease. *Clin Chest Med* 2016;37:523-34.
15. Frazier AA, Franks TJ, Mohammed TL, Ozbudak IH, Galvin JR. Pulmonary veno-occlusive disease and pulmonary capillary hemangiomas. *Radiographics* 2007;27:867-82.