Radiology Case Reports 19 (2024) 671–674

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

# Pulmonary arterial hypertension associated with portal hypertension: Noninvasive comprehensive assessment using computed tomography ☆

*Hidetaka Hayashi, MD[a], Seitaro Oda, MD, PhD[a,*], Masafumi Kidoh, MD, PhD[a], Miki Anai, MD[b], Kyoko Hirakawa, MD, PhD[b], Seiji Takashio, MD, PhD[b], Eiichiro Yamamoto, MD, PhD[b], Hiroto Takamure, MD[a], Naoya Moriguchi, MD[a], Kenichi Tsujita, MD, PhD[b], Toshinori Hirai, MD, PhD[a]*

[a] *Department of Diagnostic Radiology, Faculty of Life Sciences, Kumamoto University, 1-1-1 Honjyo, Chuo-ku, Kumamoto, 860-8556, Japan*

[b] *Department of Cardiovascular Medicine, Graduate School of Medical Sciences, Kumamoto University, 1-1-1 Honjyo, Chuo-ku, Kumamoto, 860-8556, Japan*

ARTICLE INFO

*Article history:*
Received 15 August 2023
Revised 2 October 2023
Accepted 5 November 2023
Available online 25 November 2023

*Keywords:*
Pulmonary hypertension
Portal hypertension
Dual energy computed tomography
Pulmonary perfusion
Extracellular volume

ABSTRACT

Pulmonary arterial hypertension associated with portal hypertension, known as portopulmonary hypertension (PoPH) is one of the important and serious pulmonary complications in patients with portal hypertension. Although there are a large number of patients with portal hypertension due to mainly liver cirrhosis, the number of cases diagnosed with PoPH are far fewer because the causes of dyspnea in patients with cirrhosis are diverse and the disease entity of PoPH is poorly recognized by clinicians. We report here the case with PoPH suggested and assessed comprehensively by dual energy computed tomography (CT) including high-resolution pulmonary CT angiography, pulmonary perfusion imaging, myocardial late iodine enhancement imaging, and myocardial extracellular volume analysis. This refined CT imaging protocol can be used in conjunction with standard chest evaluation and offers a practical and useful approach for the noninvasive "one-stop shop" evaluation of PoPH.



☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: seisei0430@nifty.com (S. Oda).

https://doi.org/10.1016/j.radcr.2023.11.013

## Introduction

Pulmonary arterial hypertension associated with portal hypertension, known as portopulmonary hypertension (PoPH) is classified as a type of pulmonary artery hypertension (PAH) (subset of group 1 pulmonary hypertension [PH]). PoPH is one of the important and serious pulmonary complications in patients with portal hypertension and occurs in 2%-10% of these patients [1,2]. Although there are a large number of patients with portal hypertension due to mainly liver cirrhosis, the number of cases diagnosed with PoPH is far fewer because the causes of dyspnea in patients with cirrhosis are diverse and the disease entity of PoPH is poorly recognized by clinicians. Therefore, accurate assessment and suggestion of PoPH with noninvasive imaging are crucial. We report here the case with PoPH suggested and assessed comprehensively by dual energy computed tomography (CT) including high-resolution pulmonary CT angiography, pulmonary perfusion imaging, myocardial late iodine enhancement (LIE) imaging, and myocardial extracellular volume analysis (ECV).

## Case report

A 72-year-old man presented with shortness of breath and symptoms of World Health Organization functional class II. His medical history included alcoholic cirrhosis and portal hypertension (Figs. 1A and B). He had a low blood pressure of 93/74 mm Hg, a heart rate of 65 beats/min, and an oxygen saturation of 92% with room air. Chest radiography showed enlargement of the central pulmonary arteries, with an increase in the hilar to thoracic ratio of 0.56. He had elevated levels of plasma B-type natriuretic peptide (270.1 pg/mL). Echocardiography showed findings, such as right ventricular enlargement, right ventricular wall thickening, ventricular septal flattening, and increased tricuspid regurgitation pressure gradient of 72.0 mm Hg, which suggested PH. A ventilation-perfusion (V/Q) scan to differentiate chronic thromboembolic PH showed no abnormal findings (Fig. 2A). He underwent CT imaging using a dual-layer spectral detector CT system (IQon Spectral CT; Philips Healthcare, Best, The Netherlands). A scan protocol for comprehensive assessment consisting of 2 contrast acquisitions (Supplementary Material) was used. This protocol included high-resolution pulmonary CT angiography, pulmonary perfusion imaging, myocardial late enhancement imaging, and extracellular volume analysis. CT showed dilation of the central pulmonary arteries, but no pulmonary embolism or lung lesions were observed (Fig. 2B). Pulmonary perfusion imaging revealed bilateral diffuse, inhomogeneous, patchy, and nonsegmental hypoperfusion (Figs. 2C and D). Delayed-phase cardiac CT revealed LIE at the right ventricular insertion point (RVIP), which is indicative of focal myocardial damage associated with right ventricular overload, and myocardial ECV fraction elevation in the affected segments (41% [reference range: 23%-28%]) (Figs. 2E and2F). Cardiac magnetic resonance imaging (MRI) was performed using a 3.0-T MRI scanner (Ingenia CX; Philips Healthcare) to confirm the severity of right ventricular dysfunction. Right ventricular dilatation, hypertrophy, dysfunction (right ventricle: end-diastolic volume, 162 mL; end-systolic volume, 134 mL; ejection fraction, 17%), and abnormal septal bowing with a D-shaped left ventricle were noted in cine images. In velocity-encoded phase contrast images, low velocity (mean velocity, 7.26 cm/s) in the main pulmonary artery and a normal pulmonary-to-systemic blood flow ratio (Qp/Qs, 1.0) were calculated. Focal late gadolinium enhancement was noted at the RVIP (Fig. 2G). The ECV calculated from T1 mapping before and after gadolinium-based contrast administration for the affected anterior RVIP was markedly elevated (46% [reference range: 23%-28%]) (Fig. 2H). These cardiac MRI findings suggested severe PH and were similar to the CT results. Right heart catheterization (RHC) was performed and resulted in increased mean pulmonary artery pressure (mPAP) of 45 mm Hg, pulmonary vascular resistance (PVR) of 10.7 Wood units (WU), hepatic venous pressure gradient of 15 mm Hg, decreased cardiac output (CO) of 3.54 L/min and cardiac index (CI) of

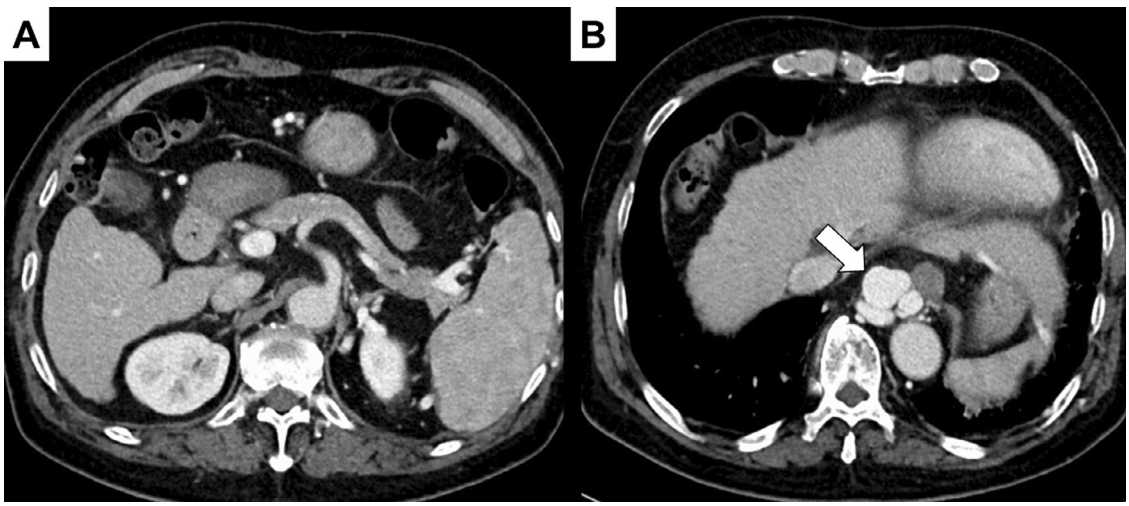


**Fig. 1 – Abdominal contrast-enhanced CT. Irregular liver margins and splenomegaly suggestive of cirrhosis and portal hypertension (A), as well as development of collateral circulation (B) (arrows).**

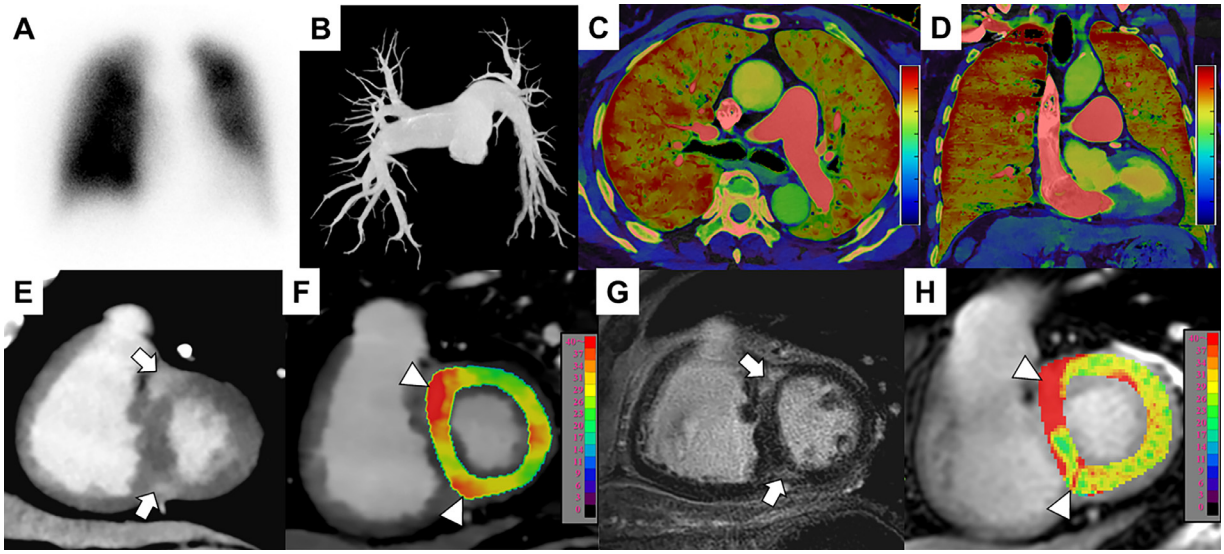


**Fig. 2 – Noninvasive diagnostic imaging of pulmonary arterial hypertension associated with portal hypertension. (A) Normal ventilation-perfusion scan. (B) Pulmonary CT angiography shows no pulmonary embolism. (C and D) Pulmonary perfusion imaging reveals bilateral diffuse, inhomogeneous, patchy, and nonsegmental hypoperfusion. (E and F) Delayed-phase CT reveals LIE at the RVIP (arrows) and ECV elevation (arrow heads). (G and H) On MRI, focal late gadolinium enhancement is noted at the RVIP (arrows), and the ECV is markedly elevated (arrow heads).**

1.98 L/min/m$^2$, and normal pulmonary arterial wedge pressure (PAWP) of 7 mm Hg. According to these examination findings and a history of cirrhosis and portal hypertension, and ruling out other causative conditions such as collagen vascular diseases, he was eventually diagnosed with PoPH. He started PAH-targeted treatment with macitentan (endothelin receptor antagonist). His symptoms improved, and a check-up RHC showed improvement with a mPAP of 32 mm Hg, a PVR of 6.0 WU., CO of 4.67 L/min, and CI of 2.86 L/min/m$^2$.

## Discussion

The diagnosis of PoPH requires demonstration of portal hypertension and precapillary PH with exclusion of other PH groups. Precapillary PH is defined as a resting mPAP of ≥20 mm Hg with a PAWP of ≤15 mm Hg and a PVR of ≥3 WU measured by RHC. The definitive diagnosis of portal hypertension is based on mainly noninvasive imaging findings (presence of splenomegaly, gastroesophageal varices, and a portosystemic shunt).

The CT scan protocol reported here, including high-resolution pulmonary CT angiography, pulmonary perfusion imaging, myocardial LIE imaging, and ECV analysis, enables comprehensive evaluation of patients with PH [3]. This refined CT imaging protocol can be used in conjunction with standard chest evaluation and offers a practical and useful approach for the noninvasive "one-stop shop" evaluation of PH.

This case had diffuse inhomogeneous pulmonary hypoperfusion despite a normal V/Q scan. An inhomogeneous patchy pulmonary hypoperfusion pattern in PAH has been reported on pulmonary perfusion scintigraphy and CT, and has been found to be associated with disease severity and to be a poor prognostic factor [4,5]. These studies focused on idiopathic PAH, and CT pulmonary perfusion and V/Q scan findings in PoPH have not been previously reported. CT pulmonary perfusion may be more sensitive than a V/Q scan for detecting pulmonary hypoperfusion in PoPH.

## Conclusion

This comprehensive CT imaging protocol can support appropriate and effective diagnostic strategies for PoPH. Diffuse inhomogeneous pulmonary hypoperfusion detected by CT pulmonary perfusion imaging may be the diagnostic key for PoPH.

## Patient consent

Written informed consent was obtained from the patient's parents for anonymized patient information to be published in this article.

## Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2023.11.013.

## REFERENCES


[1] Chen HS, Xing SR, Xu WG, Yang F, Qi XL, Wang LM, et al. Portopulmonary hypertension in cirrhotic patients: prevalence, clinical features and risk factors. Exp Ther Med 2013;5(3):819–24.

[2] Hadengue A, Benhayoun MK, Lebrec D, Benhamou JP. Pulmonary hypertension complicating portal hypertension: prevalence and relation to splanchnic hemodynamics. Gastroenterology 1991;100(2):520–8.

[3] Oda S, Kidoh M, Nagayama Y, Nakaura T, Hirakawa K, Yamamoto E, et al. Non-invasive imaging in pulmonary hypertension- comprehensive assessment using dual-layer spectral computed tomography. Circ J 2021;85(3):316.

[4] Wang M, Ma R, Wu D, Xiong C, He J, Wang L, et al. Value of lung perfusion scintigraphy in patients with idiopathic pulmonary arterial hypertension: a patchy pattern to consider. Pulm Circ 2019;9(1):2045894018816968.

[5] Lefebvre B, Kyheng M, Giordano J, Lamblin N, de Groote P, Fertin M, et al. Dual-energy CT lung perfusion characteristics in pulmonary arterial hypertension (PAH) and pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis (PVOD/PCH): preliminary experience in 63 patients. Eur Radiol 2022;32(7):4574–86.