Cardiology in the Young

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A rare variant of scimitar syndrome characterised by right pulmonary vein drainage to portal vein with eventeration of diaphragm

Thejaswi Puthiyedath, Pramod Sagar and Kothandam Sivakumar

Department of Pediatric Cardiology, Institute of Cardio Vascular Diseases, Madras Medical Mission, Chennai, India

Abstract

While infradiaphragmatic total anomalous pulmonary venous drainage to portal vein is well described, hemianomalous drainage of right pulmonary veins to portal vein in Scimitar syndrome has not yet been reported.

Images in Congenital Cardiac Disease

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Corresponding author:
K. Sivakumar;
Email: drkumarsiva@hotmail.com

Chest X-ray in a 3-year-old boy was diagnostic of eventeration of right diaphragm and cardiac dextroposition during a respiratory infection. Echocardiography showed right pulmonary artery hypoplasia and did not reveal the right pulmonary veins. The right-heart chambers were not enlarged and pulmonary artery pressures were normal. CT confirmed diaphragmatic eventeration with mild right lung hypoplasia. The small tortuous right pulmonary veins entered the diaphragm and liver and drained into left branch of the portal vein (Figs. 1, 2). A cardiac catheterisation was done to assess anatomy and haemodynamics. Eventeration of diaphragm and left lobe of liver caused a tortuous right-angled drainage of inferior caval vein (Video 1, 2). Repeated oximetry runs did not show any left-to-right shunt. Selective angiogram showed hypoplastic right pulmonary artery and tortuous small pulmonary veins coursing caudally and draining into the left branch of portal vein in the eventerated left lobe of liver through two separate venous channels (Video 3, 4, Figs. 3, 4). Even though the right pulmonary vein drainage as well as the ultimate blood flow through the left branch of portal vein were anatomically complex, the redistribution of the pulmonary blood flows to the left lung demonstrated on pulmonary angiogram did not result in significant shunt. The pulmonary artery and wedge pressures were normal.

Hemi anomalous pulmonary venous connection to portal vein is not reported earlier. This image highlights the fact that despite the anomalous drainage of right pulmonary veins to a systemic vein, the resistance in the pulmonary venous pathway is determined by the length, size,

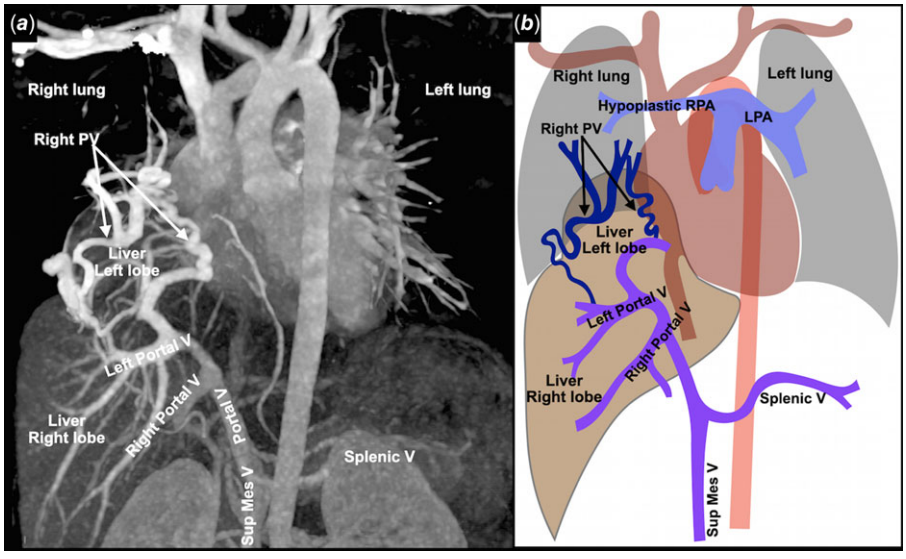

Figure 1. CT (***a***) and accompanying cartoon (***b***) show eventeration of right diaphragm with herniation of left lobe of liver. The right pulmonary veins drain through two tortuous channels to the left branch of portal vein. The right portal vein, splenic and superior mesenteric vein are also illustrated.

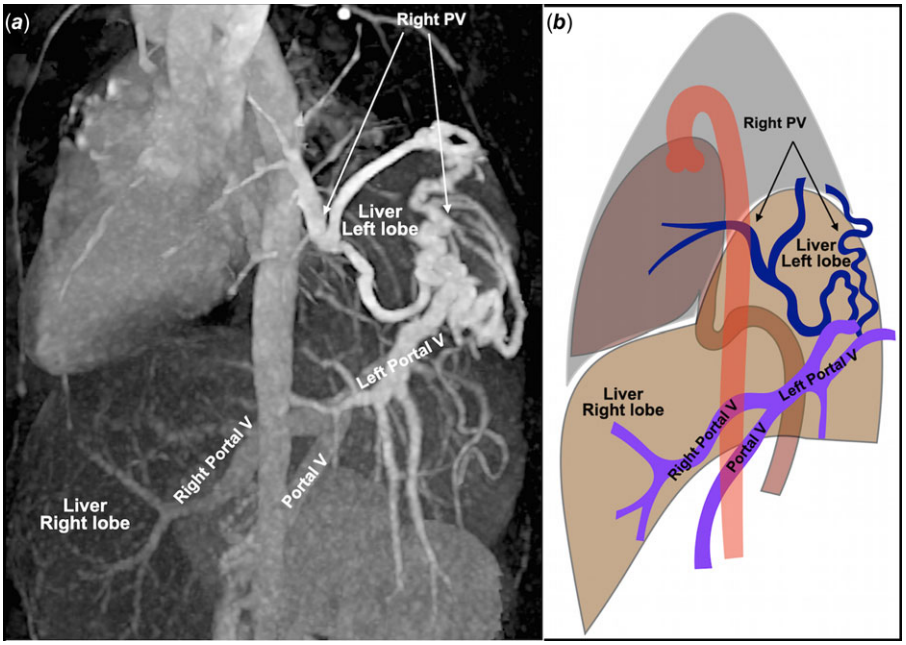

Figure 2. In a sagittal projection, as seen from lateral side, CT (***a***) and cartoon (***b***) show the anomalous thin and tortuous right pulmonary veins entering the eventerated right diaphragm and the left lobe of the liver.

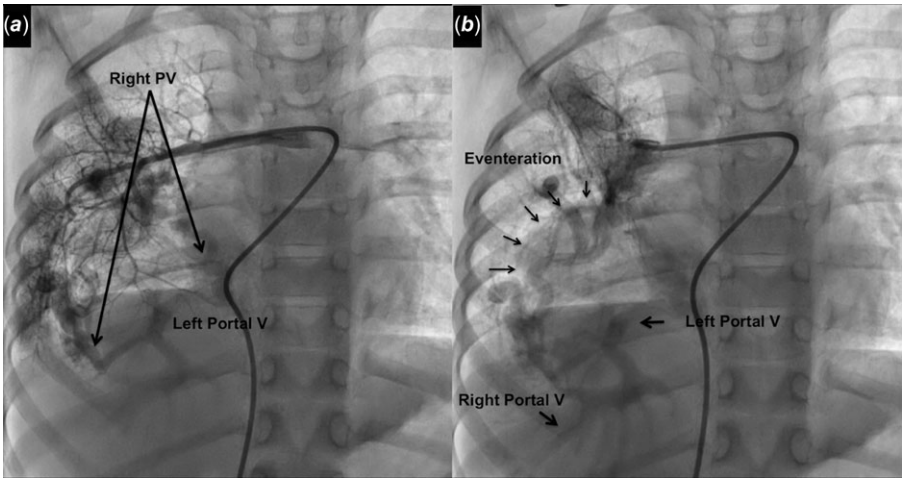

Figure 3. Venous phase of right pulmonary arteriogram (***a***) demonstrates two tortuous right pulmonary veins draining to left branch of portal vein that fills well on a later frame (***b***). Short arrows point to the eventeration of the diaphragm with herniation of left lobe of liver.

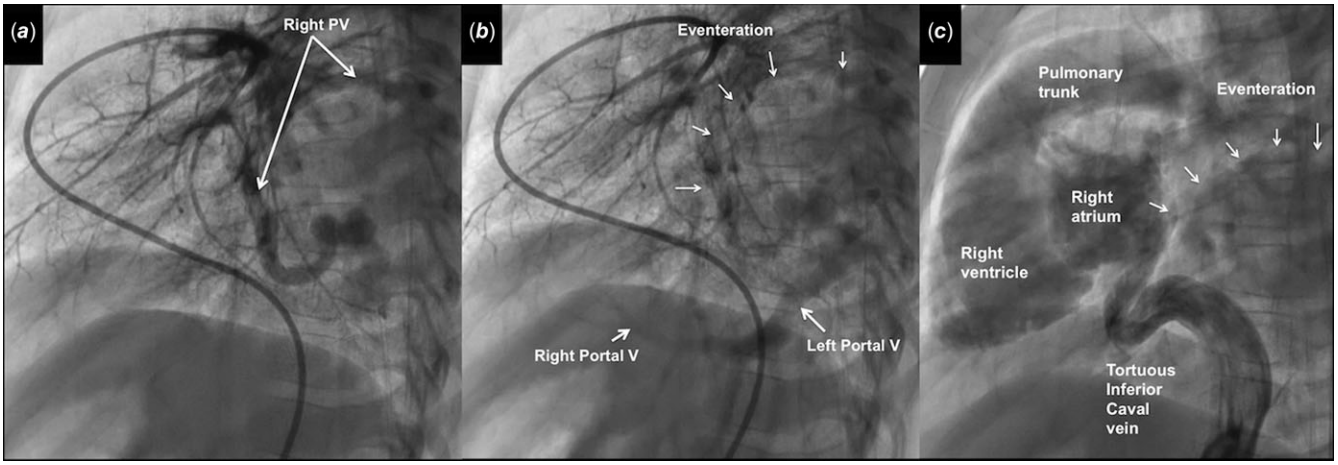

Figure 4. Venous phase of right pulmonary arteriogram in lateral view (***a***) demonstrates the right pulmonary veins, penetrating the eventerated diaphragm (short arrows) and draining into the left branch of portal vein (***b***). The herniated left lobe of liver led to an abnormal right-angled tortuosity of the inferior caval vein (***c***).

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tortuosity, and distal chamber pressure.[1] A circuitous drainage can result in redistribution of pulmonary flow into the healthier lung leading to negligible left-to-right shunt. No active interventions were planned in this asymptomatic child.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951123004468.

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Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the Indian Council of Medical Research and with the Helsinki Declaration of 1975, as revised in 2008, and have been approved by the institutional committee of Madras Medical Mission, Chennai, India.

Reference

1. Wang C-C, Wu E-T, Chen S-J, et al. Scimitar syndrome: incidence, treatment, and prognosis. Eur J Pediatr 2008; 167: 155–160.