Pulmonary Veno Occlusive Disease Presenting as Failure to Thrive

Divij Pasrija¹ • Shilpi Gupta² • Ryan Breuer¹ • Jeffrey Herendeen¹

Received: 26 August 2020 / Accepted: 4 January 2021 / Published online: 14 January 2021
© Dr. K C Chaudhuri Foundation 2021

To the Editor: We had a 6-mo-old infant, born at term (weight - 3.3 kg), who presented because of not feeding well and vomiting for a few days before admission. There was no history of fast breathing, accessory muscle use, edema noticed on any part of the body. No rash, fever or diarrhea in the past 2 mo. Workup for inadequate weight gain included complete blood count (CBC), liver function test (LFT), and kidney function test (KFT), urine electrolytes, which were within normal range. Respiratory polymerase chain reaction (PCR) panel for respiratory syncytial virus (RSV), influenza A/B, adenovirus, and other viruses was negative. Chest radiograph showed perihilar nonspecific opacification, chest CT was done which was suggestive of interstitial lung disease. Echocardiography revealed mildly elevated pulmonary artery pressure with no abnormality in function or structure of the heart. Differential diagnosis included pulmonary alveolar proteinosis, hypersensitivity pneumonitis, and bronchiolitis obliterans.

Patient was planned for lung biopsy and developed signs of acute heart failure post procedure, with elevated serum lactate (> 4 mmol/L) and pro-brain natriuretic peptide (BNP) levels (> 6000 pg/mL – Normal < 100 pg/mL). Repeat echocardiography at this time showed bowing of ventricular septum to left side of heart, suggestive of severe pulmonary hypertension despite optimization with milrinone, 100% oxygen and nitric oxide with minimal response. Parents wished to stop further care based on the ominous prognosis. Lung biopsy (received

postmortem) revealed near complete obliteration of small veins with fibrinous debris, septal thickening, which suggested the diagnosis of pulmonary veno occlusive disease (PVOD).

Key clinical message: PVOD is an extremely rare cause of lung disease in childhood and presenting symptoms include, shortness of breath, chronic cough, and rarely exertional syncope. Diagnosis should be considered in patients with pulmonary hypertension not responding to medical treatment [1]. Based on a registry, the disease is likely underdiagnosed with prevalence being described as 1.6% in a cohort with pulmonary hypertension in childhood [2]. Most patients present at an advanced stage and the interval from diagnosis to death or lung transplantation is in months [3].

As per a case series of 9 children with PVOD by Woerner et al. the diagnosis was made by biopsy at the time of lung transplantation in 2/9 patients and in 3/9 at autopsy [4].

Compliance with Ethical Standards

Conflict of Interest None.

References

1. Harch S, Whitford H, McLean C. Failure of medical therapy in pulmonary arterial hypertension. Is there an alternative diagnosis? *Chest*. 2009;135(6):1462–9.
2. Berger RM, Beghetti M, Humpl T, et al. Clinical features of paediatric pulmonary hypertension: a registry study. *Lancet*. 2012;379(9815):537–46.
3. Montani D, Lau EM, Dorfmueller P, et al. Pulmonary veno-occlusive disease. *Eur Respir J*. 2016;47(5):1518–34. <https://doi.org/10.1183/13993003.00026-2016>.
4. Woerner C, Cutz E, Yoo SJ, Grasemann H, Humpl T. Pulmonary venoocclusive disease in childhood. *Chest*. 2014;146(1):167–74. <https://doi.org/10.1378/chest.13-0172>.

✉ Divij Pasrija
divijpasrijamamc@gmail.com; divijpas@buffalo.edu

¹ Department of Pediatric Critical Care, Oishei Children's Hospital, University of Buffalo, 9th Floor, 818 Ellicott Street, Buffalo, NY 14203, USA

² Department of Internal Medicine, Mercy Hospital of Buffalo, Buffalo, NY, USA