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IMAGE | LIVER

Severe Hepatopulmonary Syndrome in an Adolescent Patient with Non-Cirrhotic Portal Fibrosis

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CASE REPORT

A 17-year-old male presented with progressive splenomegaly since age 7 years, recurrent epistaxis and skin bleeds for the last 5 years, and worsening dyspnoea (initially on exertion progressing to symptoms at rest) for the last 3 years. Examination showed evidence of hypoxemia (blood oxygen saturation on room air, 70%; on 3 L/min oxygen, 92%, with orthodeoxia), clubbing, peripheral cyanosis, petechiae, and massive splenomegaly. Investigations revealed evidence of hypersplenism and normal hepatic synthetic functions. Work-up for known etiologies of chronic liver disease was negative. Liver biopsy showed maintained liver architecture with obliterative portal venopathy suggestive of non-cirrhotic portal fibrosis (NCPF). There was evidence of severe hepatopulmonary syndrome (HPS), including severe shunting on saline contrast echocardiography, arterial hypoxemia of PaO_2 49 mm Hg, alveolar-arterial PO_2 gradient 60 mm Hg, and macro-aggregated albumin perfusion lung scan showing brain uptake of 25.4% (Figure 1). Pulmonary angiography revealed increased vessel density in the bilateral pulmonary parenchyma with hypertrophied and tortuous bronchial arteries but no obvious intrapulmonary shunting (Figure 1). He was managed conservatively with continuous oxygen therapy and pentoxiphylline. The need for liver transplantation as a definitive management was explained to the family.

NCPF has been classically described as a disease of adulthood with a mean age of onset in third to fourth decade.[1] Though reported in more than one-third of cirrhotic patients, HPS is a rare entity in non-cirrhotic portal hypertension and is usually asymptomatic, especially in younger patients.[2-4] Very limited pediatric data are available for HPS prevalence, especially in non-

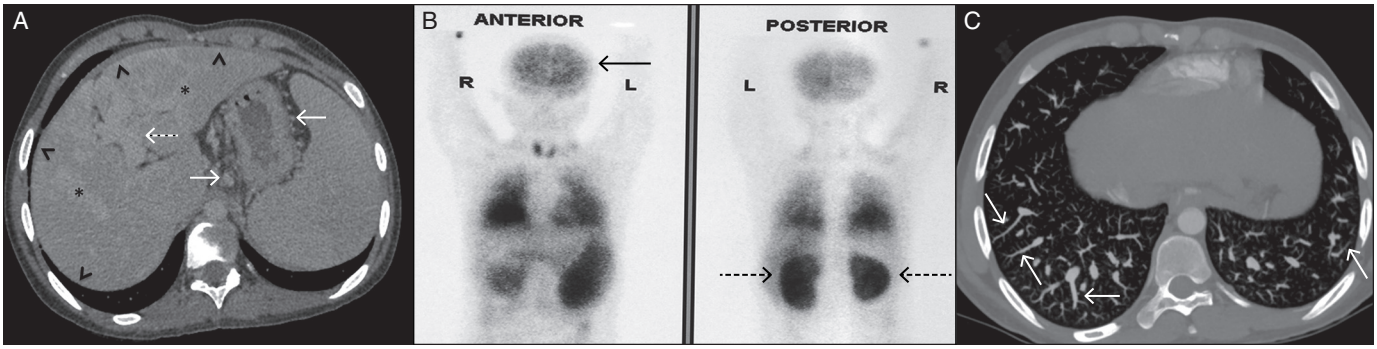

Figure 1. (A) Axial abdominal contrast-enhanced computed tomography showing multiple perigastric portosystemic collaterals (arrows) with splenomegaly consistent with portal hypertension. Hepatic contour is smooth (arrowheads) with interlacing areas of fibrosis (asterisks), suggesting the possibility of non-cirrhotic portal fibrosis. Portal vein is patent (interrupted arrow). (B) 99m-Technetium macro-aggregated albumin perfusion lung scan showing significant extrapulmonary uptake of tracer over the brain (solid arrow) and kidneys (interrupted arrows), suggesting shunting through the lung caused by an intrapulmonary shunt. (C) Axial maximum intensity projection image showing an increased number of visible peripheral pulmonary artery branches with dilatation of distal peripheral lower lobe pulmonary arteries that do not taper normally and extend out to the pleural surface (arrows).

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cirrhotic subjects. In the study by Sari et al, out of 16 non-cirrhotic pediatric cases, none of them fulfilled the diagnostic criteria of HPS.[5] Another study of a pediatric non-cirrhotic population showed the prevalence of HPS was 13%.[3] This suggests that HPS in a pediatric population with NCPF is a rare phenomenon. Ours is likely among the few reports in the literature of symptomatic HPS in a pediatric subject with NCPF.[4]

Clinical presentation in our case is unique for a NCPF patient. NCPF patients usually have an underlying HPS on investigation but without any symptoms at diagnosis. Symptomatic HPS with severe hypoxia (severe HPS) and cyanosis at a young age (<18 years) is unique even as per adult standards. This has prognostic complications, especially for liver transplant considerations in the perioperative period. Though options like medical therapies (e.g., antibiotics, nitric oxide inhibitors, etc.) and interventional therapies (e.g., shunts, etc.) have been tried, they have remained largely unsuccessful, and liver transplantation remains the only definitive cure.[4,6]

DISCLOSURES

Author contributions: V. Sood, S. Rajesh, and B. Bihari Lal contributed equally to drafting the manuscript. D. Rawat and S. Alam supervised and edited the manuscript. V. Sood wrote the article and is the article guarantor.

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