



Heptatopulmonary Syndrome- A Cause of Cyanosis Beyond Cardiopulmonary

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Abstract

The hepatopulmonary syndrome is a rare lung complication of liver disease. When liver is not functioning properly, blood vessel in lung may dilate, if this is severe enough, the lungs can lose their ability to effectively transfer oxygen to the body. This is called as hepatopulmonary syndrome and it occurs in approximately 5-32% of patients with scarring of liver. The following case highlights complication of liver disease leading to pulmonary compromise, leading to hypoxemia in a known case of disseminated tuberculosis. Child had come with complains of fever and respiratory distress since 4 days. On examination child was cachexic, neck veins pulsatile, central cyanosis present, pallor present, grade 3 clubbing present. Spo₂- 62% on room air, CVS-s1s2+, grade 4 murmur with thrill, P/A- left of liver palpable 3cm below RCM, non tender, with sharp margins. Child was evaluated thoroughly, pulmonary angiography was done s/o small AV fistula. CECT abdo s/o liver parenchymal disease with dysplastic nodules with portal hypertension with narrowing of splenic artery aneurysm. The cause of chronic liver disease was a mystery until HIV had come out to be positive. Liver transplant of the patient was planned and child was discharged after explaining grave prognosis to the mother about the condition of child.

Keywords: Hepatopulmonary syndrome, hypoxemia, chronic liver disease, HIV.

Introduction

Hepatopulmonary syndrome is an important complication of liver disease characterised by intrapulmonary vascular dilatation and hypoxemia. Prevalence of hepatopulmonary syndrome and its outcome in children is unknown. Hepatopulmonary syndrome is defined as a triad of liver disease with or without portal hypertension, abnormality in gas exchange, pao₂<80%, alveolar-arterial difference > 15 mmhg and pulmonary capillary vascular dilatation. Orthodeoxia test is done in hepatopulmonary, it is characterised by decrease in Pao₂> 4 mmhg or to 5% from supine decubitus

position to standing position. Arterial blood gas shows hypoxemia, respiratory alkalosis. It is due to hyperventilation intended to compensate for hypoxemia. Alveolar-arterial Pao₂ difference > 15 mmhg is an early marker of hepato-pulmonary syndrome. Hepatopulmonary syndrome should be searched in children with liver disease having dyspnea and hypoxemia who are in a liver transplantation protocol. Its diagnosis will get priorities in transplant waiting list.

Case Report

11 years old male child, born of non consanguineous marriage was brought by mother

with complaints of fever since 4 days and increased respiratory activity since 4 days. Child had past history of admission in department of cardiology in view of increased respiratory activity and diagnosed as pulmonary AV fistula. Child was diagnosed as disseminated TB in 2019. Child had history of Koch's contact with mother who was a diagnosed case of pulmonary TB. There is family history of father death due to some liver disease as per mother. On examination child was cachexia, afebrile, pr-88/min, good in volume, character, regular with no radio-radial and radio-femoral delay. RR- 36/min with mild suprasternal retraction, blood pressure was 94/62 mmhg, Spo2-RU-62%, RL-68%, LU-60%, LL-70%, neck veins pulsatile, pallor present, central cyanosis present, clubbing present, b/l cervical lymphadenopathy present, cvs-s1s2+, grade 4 murmur present, thrill present, 3cm firm left of liver palpable, sharp margins, non tender, 5 cm of spleen palpable, firm, CNS – GCS – 15/15. RS- air entry b/l equal and clear, child was already started on cat 1 AKT which was continued, arterial blood gas was done s/o PO2-68%, hyperoxia test was done which showed difference of 18% of PO2., fever profile was sent which came out to be negative. Child was started on antibiotics empirically, 2d echo with bubble contrast was done s/o? pulmonary AV fistula advised to do CT angiography to confirm the diagnosis. Liver function was deranged, sgot-134, sgpt - 100, APTT-34.4, PT-17.9, INR-1.4, ALP-247, GGT-74, LDH-549, TOTAL BILI – 2, in view of deranged LFT usg abdomen was done s/o borderline hepatosplenomegaly with raised echotexture with portal vein dilated measuring 18 mm, hence ct abdomen was done s/o liver parenchymal disease with dysplastic nodules with portal hypertension with narrowing of splenic artery aneurysm, pulmonary angiography was done s/o small AV fistula. Child continued to have fever spikes, repeat fever profile also came out to be negative. Antibiotics were stepped up as per sensitivity of intensive care unit, child did not respond. HIV test was sent, which came out to be

negative, on repeating the test, it came out to be equivalent, was advised to confirm by western blot test which came out to be positive on confirming, hence the cause of chronic liver disease was found. Hence the triad of hepatopulmonary syndrome was fulfilled and child was diagnosed as hepatopulmonary syndrome. Mother and other siblings were screened for HIV, mother came out to be positive for HIV. Both mother and child was started on ART. Gradually child responded. Child always had baseline tachypnea, central cyanosis and clubbing, mother was explained about medical illness and guarded prognosis of the child and was discharged as per seniors advice.



Case Discussion

Hepato pulmonary syndrome is characterised by hypoxemia with presence of alveolar arterial difference of > 15 mmhg. Depending upon alveolar arterial difference there is classification of severity of hepatopulmonary syndrome. Mild – $\text{Pao}_2 > 80$ mmhg, moderate – Pao_2 60-80 mmhg, severe – $\text{Pao}_2 < 50$ mmhg. Based on these criteria patient is classified as moderate hepatopulmonary syndrome. This classification is carried out to identify liver transplant candidate with poorest prognosis. Child was initially thought of some TOF physiology, hence 2 d echo was done s/o AV fistula, hence echo was repeated with bubble contrast s/o bubble seen in left side of heart after 5 cycles. This pulmonary pathology was secondary to some liver disorder as per usg abdomen and ct abdomen report. The cause of liver pathology was yet to be diagnosed. Chronic hepatitis was suspected hence TRIPLE H markers were sent, out of which HIV has come to be positive. As the criteria for hepatopulmonary syndrome was full filled, hence child was labelled as a case of hepatopulmonary syndrome in known case of disseminated tb. As per the Krowha classification, hepatopulmonary syndrome is classified into 2 groups, TYPE –I [diffuse], patients with pre capillary dilatation, who respond to 100% O_2 , achieving $\text{PaO}_2 > 200$ MMHG. Type II [focal fistulas], patients with localised dilatation, similar to arterio-venous malformation who have a poor response to administration of 100% O_2 , these patients present with true shunts, who are unable to attain $\text{Pao}_2 > 200$ mmhg. It is less common than type 1.

There is no effective medical management of hepatopulmonary syndrome. Liver transplantation is only treatment which reverses hepatopulmonary syndrome. Its success rate is 70-80%, resolution of hepatopulmonary syndrome post transplantation is seen after 3 months. Home oxygen therapy is considered for those patients with $\text{Pao}_2 < 60$ mmhg, it improves circulatory complications of hypoxemia and improves quality of life while waiting for transplant.

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