AN INTERESTING CASE OF HEPATOPULMONARY SYNDROME

MALARVIZHI
Department of Medical Gastroenterology,
MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract:
Here we report a case of 15 year old male presented with recurrent episodes of upper GI Bleed with dyspnea on sitting past history of jaundice on examination Moderately built, nourished, central cyanosis pandigital clubbing, no pedal edema, no lymphadenopathy, spider naevi , no jaundice Systemic examination Examination of AbdomenLivspan10cmsSplenomegaly, no free fluidCVS S1S2 heard no murmurRS NVBS no added sounds CBC HB 19.8 gmsdl PCV 60.5 T.B 0.8mgdl SGOT 32 IU SGPT 45 IU T.P 6.3gmsdl USG ABD Doppler Both lobes of Liver small 8.8 cms portal vein -13mm no respiration variations splenic vein 7.6 mm splenoportal collaterals UGIE Esophageal varices Grade 1-11 columns , PHTG Arterial blood gas Analysis PH 7.47 Pco2 24.5 Pao2 48 Sao2 81.3 Na 141 K 3.7 Hco3 52 CECT chest normal ECHO Situs Levocardia ,Normal AVVA concordance Septum intact, mPAP 17 mmhg no PHT, normal LV function

ContrastEchocardiogram Peripheral intravenous injection of agitated saline to produce micro bubbles(10-15micron) appeared in Left ventricle Delayed after 3 to 6 beats suggestive of intrapulmonary shunting

Keyword :
Hepatopulmonary Sy drome, ContrastEchocardiogram,Intrapulmonary Shunting, Platypnea.

INTRODUCTION:
Hepatopulmonary syndrome can occur in 10 to 30 in patients with Cirrhosis, NCPF, Chronic hepatitis with Portal Hypertension. Characterised by hypoxemia Arterial alveolar gradient,Age corrected A-aO2 < 15mmhg It is a triad of hepatic dysfunction,PaO2<70 mmHg(at an inspired oxygen 0.21) , intrapulmonary vasodilation. The disease course is progressive with increased mortality and morbidity. The Occurrence of Hepato Pulmonary Syndrome does not correlate with stage of Liver disease
Definitive treatment is Liver Transplantation

CASE REPORT:

**Case summary:** 15yr old boy presented with breathlessness for 5yrs - Platypnea. History of recurrent UGI bleed for 3mths. H/o jaundice 2yrs ago. **Physical Examination** Mod Built & nourished, Central cyanosis Pandigital clubbing Abdomen: Liver span-10cms, moderate Splenomegaly UGI scopy- Esophageal varices Gr1-11.

CECT-ABD: Splenomegaly, dilated Spleni&c Portal vein, plenorenal, Gastric collaterals ECHO revealed Normal AV/VA concordance **Contrast ECHO**: injection of agitated saline microbubbles {10-15micron} Delayed appearance of microbubbles [3-6beats] in left side of heart - **Intrapulmonary shunting Characteristic of Hepatopulmonary syndrome.**

Patient was treated symptomatically and listed for Liver transplantation.

**Discussion:** Pathophysiology of Hepatopulmonary Syndrome

**Vasodilatation:**
Overproduction of the vasodilators, Nitric Oxide from injured hepatobiliary system.
Decrease in their clearance by the liver.

*Normal sensitivity of the pulmonary vessels to vasoconstrictors in response to hypoxemia is blunted in HPS.*
Hypoxemia:
Pulmonary capillary diameter is normally about 8-15 micrometer (µm) and this could rise up to 500 µm in HPS which causes hypoxemia.

These changes lead to the following:
- a. Ventilation perfusion (V/Q) mismatch:
- b. Right to left shunting of the blood:
- c. Diffusion impairment:

Treatment Liver Tx if PaO2 50-60 mmHg
Intrapulmonary Embolization Coil Embolisation of the pulmonary AV shunt prior Liver Transplantation TIPS Palliation as patient awaits Liver Transplantation

Conclusion:
Liver transplant priority consideration In patients with HPS exists in the pediatric population. Hence, high index of clinical suspicion and investigations to diagnose Hepatopulmonary syndrome becomes quintessential in management of these patients.

This case is presented for its rarity and characteristic findings in contrast ECHO.

References:
Hepatopulmonary Syndrome — A Liver-Induced Lung Vascular Disorder


