Hepatopulmonary Syndrome

G Agrawal*, N Kumar**, D Rosha***

Abstract

Hepatopulmonary syndrome (HPS) is defined as clinical triad of advanced liver disease, arterial deoxygenation and intra pulmonary vascular dilatation. It is a rare complication of liver disease of varied etiology and indicates a poor prognosis. Many theories have been put forward to throw light over its pathogenesis. The major clinical manifestations are arterial hypoxemia, clubbed fingers and spider naeovi. Orthodeoxia and platypnea are usual clinical features. A simple non invasive method to screen HPS is desirable. Contrast enhanced 2D ECHO cardiography is the preferred screening test. No effective medical treatment has been found. Although liver transplant seems feasible to reverse (at least partially) this situation, however it is associated with increased post operative morbidity and mortality. ©

INTRODUCTION

Hypoxemia is a common clinical manifestation in patient with liver cirrhosis. If hypoxemia and dyspnea develop in these patients in the absence of known intrinsic cardiopulmonary disorder, the hepatopulmonary syndrome must be considered. Relationship between cirrhotic liver disease and lung was first described by Fluckiger in 1884 in a women with liver cirrhosis, cyanosis, and digital clubbing. In the year 1977, Kennedy and Knudsen coined the term ‘Hepatopulmonary Syndrome’.

We present a case of a patient with hepatopulmonary syndrome, as well the current understanding of the pathogenesis and clinical management of hepatopulmonary syndrome.

CASE REPORT

A 52 year old female was shifted to neuro intensive care unit with history of fall in bathroom on 11/08/06 evening. She had severe headache, vomiting followed by sudden loss of consciousness on next morning. Her MRI brain done at local hospital revealed large right sided intra cerebral haemorrhage with significant midline shift. She was intubated and put on ventilator support in the view of hypotension, desaturation and decerebrate posture in the emergency department.

Her past history revealed ERCP done in year 2001 followed by cholecystectomy in year 2002. Upper gastrointestinal tract endoscopy done in the past revealed grade 3 oesophageal varices. There was a history of alcohol abuse. She had no known cardiac or pulmonary disease.

On physical examination she had temperature of 99.4 F, pulse of 82/min, blood pressure of 100/60 mm hg and respiratory rate of 24/min. The extremities revealed grade 3 clubbing but no edema and no sign of deep vein thrombosis. Neurological examination revealed unequal pupils (right 3.5 mm reactive to light and left 1.5 mm sluggishly reactive to light) and decerebrate posture with GCS scale of 3. Respiratory and cardiology examination were unremarkable.

Initially laboratory test were significant for high haemoglobin (18gm%) and normal WBC count. LFT’s were mildly deranged, chest X ray showed mild haziness in the right lower zone. ECG showed normal sinus rhythm and ECHO showed no pulmonary artery hypertension. ABG showed pH 7.52, paCO2 of 39 mm hg, paO2 of 56 mm hg, and 87% saturation on fio2 of 1.0.

She underwent craniotomy and urgent evacuation of intra cerebral haemorrhage. Tracheostomy was done in the view of long term airway protection and difficulty to wean off from ventilator after 3 days. USG abdomen showed hepatosplenomegaly with liver parenchymal disease and portal hypertension. Anti HBc were reactive. She showed significant improvement neurologically but was difficult to wean her from ventilator. Her saturation remained around 80% with high fio2 (about 70 %). Two probable diagnosis in this context were pulmonary embolism and hepatopulmonary syndrome. D dimer level done was 1100. A contrast micro bubble Echocardiography was done, as she could not be moved put for CT pulmonary angiography.

*DNB 2nd year; **Registrar; ***Senior Consultant, Department of Respiratory Medicine, Indraprastha Apollo Hospital, New Delhi-110076.

Received : 28.8.2007; Revised : 7.11.2007; Accepted : 18.2.2008

© JAPI • VOL. 56 • APRIL 2008 www.japi.org
Meanwhile her spO$_2$ showed drastic improvement when made to lie down in supine position rather than in propped up posture. Agitated saline appeared in left atrium and ventricle after 3-4 beat consistent with intrapulmonary shunt. Finally CT pulmonary angiography showed multiple dilated vasculature channels in lower lobes of both lungs with early venous filling (Figs. 1 and 2). Based on this result and clinical background a diagnosis of Hepatopulmonary syndrome was made.

**DISCUSSION**

Our patient had all four criteria of HPS 1) Chronic liver disease 2) Pulmonary gas exchange abnormalities with an increased alveolar arterial gradient 3) Evidence of intra pulmonary shunting and 4) Absence of other significant cardio pulmonary disease. Relationship between cirrhotic liver disease and lung was first described by ‘Fluckiger’ in 1884 based on observation of a women with cirrhosis, cyanosis and clubbed finger. In 1977, Kennedy and Knudson coined the term hepatopulmonary syndrome to describe this entity. Mild hypoxemia occurs in approximately 1/3rd of all patients with chronic liver disease and is often multifactorial. The hypoxemia of the HPS, however, is uncommon, and its exact incidence is not known. The prevalence in the setting of cirrhosis ranges from 4-17%. The correlation between the severity of liver disease and incidence of HPS remains controversial. However, it was reported that HPS occurs occasionally in non-cirrhotic cases of portal hypertension and could completely reverse after the causative agent is eradicated. Patient with HPS may be asymptomatic (only 18% have dyspnea). Platypnea is well described but not always present, while orthodeoxia is not unique to HPS, but is highly suggestive of it. Severity of hypoxemia does not correlate with the severity of underlying liver disease. Clubbing, cyanosis and spider nevi may also be present.

Etiology of this syndrome remains unknown. Most commonly accepted hypothesis postulates that there is in adequate synthesis or metabolism of pulmonary vasoactive substance by impaired liver, leading to functional vasodilatation of the pulmonary vasculature producing hypoxemia. To date, however, no particular substances have being implicated in causing this dilatation, but possibilities include prostaglandins, nitric oxide, vasoactive intestinal peptide, calcitonin, glucagons, substance P, and atrial natriuretic factor. Hypoxemia is postulated to be due to decrease oxygen diffusion into the dilated vessels along with decrease intrapulmonary blood transit time. The blood transit time is decreased due to the lower vascular resistance in the intra pulmonary dilatation and associated hyper dynamic circulation characteristic of liver disease. Therefore there is not a true shunt and PaO$_2$ can be significantly improved by supplemental oxygen. Finally, HPS patient has also being found to have decreased hypoxic pulmonary vascular constriction.

Intra pulmonary vascular dilatations have being classified into two types based on pulmonary angiography. More common type 1 lesion can be characterize by diffuse pulmonary vascular dilatation with good response to 100% oxygen, while type 2 lesion are more discrete and localized dilatation with poor response to oxygen. Three imaging techniques used to diagnose intra pulmonary vascular dilatation are 1)
INNOVATIVE PHYSICIANS CAN GET RICH IN FAME, OTHERS CAN DELAY THE OPPORTUNITY

CONCLUSION

The hepatopulmonary syndrome is under recognized complication of chronic liver disease. It can be treated with oxygen but it is potentially correctable only with reversal of underlying disease or with liver transplantation. It must be considered in every patient with advanced liver disease manifesting symptoms of dyspnea and hypoxemia. Further research is needed to establish its underlying etiology and pathogenesis.

REFERENCES


