HEPATOPULMONARY SYNDROME : A RARE ENTITY

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SUMMARY : Hepatopulmonary syndrome is a rare entity for cirrhosis. We try to discuss this rare syndrome for a new case.

Key Words : Hepatopulmonary Syndrome, Spider Naevi, Cirrhosis.

INTRODUCTION

Central cyanosis in cirrhosis of liver was first described by Fluchiger in 1884 (1). The term “hepatopulmonary syndrome” (HPS) was first suggested by Kennedy and Knudson in 1977 (2) and has received attention recently because of the potential complications associated with liver transplantation in patients with severe hypoxemia (3). Hepatopulmonary syndrome is a triad of abnormal arterial oxygenation, hepatic dysfunction, and pulmonary vascular dilatations (3, 4).

We report here a case with hepatopulmonary syndrome.

CASE REPORT

An 18-year-old man referred to our hospital on June 17, 1994 for evaluation of liver dysfunction and cyanosis. He had a history of blood transfusion 15 years ago due to splenectomy. After 3 years from this operation, cyanosis occurred. He had no history of tobacco, alcohol abuse or hypertension. He complained of grade 1-2 effort intolerance for the last 3 years. On admission, marked cyanosis of the lips and nail beds, and clubbing of fingers were present. Many vascular spiders were also noted in the anterior and posterior wall of the chest. The liver was not palpable and the midline splenectomy scar was present. No ascites was exhibited and the remainder of the examination was negative. Blood pressure was 110/70 mmHg.

Laboratory data were as follows: erythrocyte count 5.86 x 10^{12} / L; hemoglobin 17.2 g/dl; leukocyte count 6.9 x 10^{9} / L; platelet count 203 x 10^{9} / L; serum total bilirubin 2.44 µmol / L; aspartate aminotransferase 69 U/L; alanin aminotransferase 101 U/L; lactate dehydrogenase 143 U/L; alkaline phosphatase 284 U/L; glutathione transferase 254 U/L; prothrombin time 12 seconds, albumin 3.7 g/dl and globulin 3 g/dl. Serum hepatitis B surface antigen was negative and antibody to hepatitis C virus was positive. Abdominal ultrasound and computed tomography revealed nodular liver and no evidence of liver tumor. Liver biopsy specimen disclosed established liver cirrhosis.

Arterial blood gas analysis revealed \( \text{PO}_2 \) 53 mmHg, \( \text{PCO}_2 \) 29.3 mmHg, and \( \text{O}_2 \) saturation 89.1%. Chest roentgenogram was normal. (Fig 1). The electrocardiogram, spirogram, and flow volume curve were normal. Echocardiography showed nor-
normal intracardiac anatomy, normal wall movement, and no evidence of atrial or ventricular septal defects.

Intravenously injected $^{99m}$Tc-macroaggregated albumin particles passed through the lungs and whole body imaging revealed a significant uptake not only in both lungs but also in the liver, brain and both kidneys (Fig 2a, b, c). The shunt ratio was 60%, as estimated by the quantitative radionuclide method (5).

Bone marrow specimens obtained by needle aspiration disclosed normocellular marrow with pronounced erythroid series hyperplasia, and the myeloid/erythroid ratio was 0.55. The number and size of megakaryocytes was normal.

Serum erythropoietin and vitamin B$_12$ levels were 15 mU/ml (normal: 5–30 mU/ml) and 440 pg/ml (normal: 200–1100 pg/ml) respectively. Leukocyte alkaline phosphatase score was 60. Routine hemoglobin electrophoresis was normal. Methemoglobin level was in normal limit (0.1%). These findings denied complications of polycythemia vera and hemoglobin abnormalities. Endoscopy revealed grade 2/4 esophageal varices. Thorax CT findings were normal.

**DISCUSSION**

In the present case, severe hyperoxemia without
hypercapnea was noted. The pulmonary functions were within normal limits. Intravenously injected $99m$ Tc- macroaggregated albumin particles passed through the pulmonary vasculature into the systemic circulation and lodged in endorgan capillary beds, such as in the kidneys, spleen and brain. This finding is consistent with those of hepatopulmonary syndrome (1, 4, 5, 9, 11, 18, 19, 20).

Hepatopulmonary syndrome was diagnosed according to published criteria and was based on contrast-enhanced two-dimensional echocardiography or $99m$ Tc- macroaggregated albumin lung perfusion scanning (21-22). Our observations indicate that $99m$ Tc- macroaggregated albumin lung perfusion scanning is a valuable tool in the diagnosis of this entity.

In patients with the hepatopulmonary syndrome, pulmonary angiography may appear normal or may suggest either diffuse or focal abnormalities. Our patient did not accept angiography. Besides, normal doppler echocardiographic findings hindered us from performing this invasive examination (7).

Recent data has confirmed that there are no particular biochemical indicators and clinical associations of liver disease that strongly correlate hepatopulmonary syndrome are very few (13, 17).

For reasons which are yet unclear, the pulmonary arterial vascular bed can dilate in the setting of acute and chronic liver disorders. The pulmonary vascular dilatations are most commonly associated with those patients that has also spider angiomas of the skin and conditions resulting in portal hypertension. These dilatations result in diagnostic findings that are quite characteristic for hypoxemia associated with this acquired pathophysiology (12).

Although specific correlations between hepatic pathology and pulmonary dysfunction are not clear according to the literature, the cutaneous spider naevi and the existence of extremity clubbing corresponded with hepatopulmonary syndrome. Our present case also had a lot of spider naevi. There may be a relation between pulmonary and skin capillary dilatation in hepatopulmonary syndrome (1, 3, 7, 8, 14, 15, 16). On the other hand, one study has suggested a correlation between the severity of esophageal varices and HPS (12).

Finally, a group of patients with clinically stable chronic liver disease may develop progressively deteriorating arterial oxygenation. Further studies in this syndrome are needed to address:

1- The identification of possible biochemical factors responsible for the pulmonary vascular dilatations.

2- The role of hepatic parenchymal dysfunction versus the degree of portal hypertension.

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