A Case of Hepatopulmonary Syndrome as First Manifestation of Liver Cirrhosis in Hypopituitarism.

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Abstract

Background: Hepatopulmonary syndrome (HPS) is a vascular complication of liver cirrhosis and/or portal hypertension causing pulmonary microvascular dilatation and angiogenesis resulting in a ventilation-perfusion mismatch that induces hypoxia. The prevalence of HPS depends on the diagnostic criteria and methodology used ranging from 4% to 47% in patients with cirrhosis affecting 10 to 30% of patients with liver transplant indication. The pathophysiology is not completely understood however different physiological processes related to an increased production of nitric oxide and pro-angiogenic factors has been described. The association between the development of non-fatty liver disease (NFLD) secondary to the metabolic syndrome triggered by hypopituitary dysfunction has also been reported. We describe a case of HPS as first manifestation of liver cirrhosis in a patient with history of hypopituitarism.

Case presentation: 39-year-old Hispanic male with history of craniopharyngioma status post resection and radiation 19 years ago who complained of dyspnea for two months. On physical examination, oxygen saturation 97% on 3L nasal canula, digital clubbing, platypnea and orthodeoxia (SO2 84% on room air in upright position improving to 93% lying down) were found. Blood work revealed normal liver chemistry, arterial gases pH of 7.46, pCO2 29, pO2 49 HCO3 20 and A-a gradient 64.5mmHg suggestive of shunting. Transthoracic contrast echocardiogram revealed delay appearance of bubbles in the right-side chambers suggestive of extracardiac shunt. Liver biopsy revealed stage III bridging fibrosis, mild degree of portal chronic inflammation with ductular reaction and vascular dilatation. The underlying etiology of shunt was attributed to HPS in the setting of newly diagnosis of chronic liver disease. Shunting confirmed with nuclear medicine study (R>L shunt 18%). The patient was transferred to high care level institutions for liver transplant evaluation.

Conclusion: Diagnosis of HPS is important due to its significant impact in mortality and quality of life. As most of the patients are asymptomatic, a high clinical suspicion is crucial. Some procedures including transjugular intrahepatic portosystemic shunt and medications have been studied for HPS. However, liver transplantation is the only definitive treatment. Dysfunction of the hypothalamic – pituitary axis has been associated with NFLD secondary to metabolic syndrome.