ABSTRACT

Hepatopulmonary syndrome in a 22 year old gentleman with liver cirrhosis

Caporusso, J[1]; Alexander, B[1], Suri, P[2].

1. University of Texas Rio Grande Valley School of Medicine-Department of Internal Medicine, Doctors Hospital at Renaissance
2. Manipal College of Medical Sciences, Kathmandu University, Pokhara, Nepal.

BACKGROUND

Hepatopulmonary syndrome is a severe complication of end-stage liver disease characterized by triad of liver disease, intrapulmonary vascular dilatation, and arterial hypoxemia. Although the pathogenesis is not completely understood, pulmonary vascular dilatation occurs due to imbalance between vasodilators and vasoconstrictors. Liver injury is thought to increase endothelin production and cause bacterial translocation, causing increased nitric oxide production, causing vasodilation of pulmonary vasculature. History and physical examination are important in leading the physician to the correct diagnosis as the majority of these patients present with non-specific clinical manifestations and imaging. Identification of specific physical exam findings is important in not missing key features of a patient's physical exam which will give you direction to a diagnosis of hepatopulmonary syndrome.

CASE PRESENTATION

We are presenting a 22-year-old hispanic male with past medical history of nonalcoholic steatohepatitis-related liver cirrhosis with evidence of portal hypertensive gastropathy with esophageal varices who presented with intermittent dyspnea and desaturation. Physical exam did not have overt signs of volume overload and was positive for platypnea and orthodeoxia. ABG revealed hypoxemia with PaO2 of 66 and orthodeoxia. Diagnosis of hepatopulmonary syndrome was confirmed with contrast Echo ordered which revealed normal ejection fraction and showed R to L shunting by agitated saline contrast. The patient was managed with medical and oxygen therapy. He was discharged home on oxygen therapy as he improved. Patient evaluated by a hepatologist outpatient and placed on the liver transplant list.

DISCUSSION

Hepatopulmonary syndrome can often be missed due to its nonspecific presentation. Dyspnea is its most common presenting symptom. However, being aware of other presenting symptoms are key to diagnosis; such as platypnea and orthodeoxia, as present in our case. Obtaining

Echo with contrast is important as it will confirm the presence of an intrapulmonary shunt. Differentiating an intracardiac shunt vs intrapulmonary shunt is important. With an intracardiac shunt, contrast appears in the left heart within three heart beats after injection, however, with an intrapulmonary shunt, contrast appears in the left heart after three beats as in our patient. Once diagnosed, oxygen therapy is recommended. Liver transplantation is the only effective therapy.