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Case report

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Hepatopulmonary syndrome : a rare diagnosis or easily missed entity?

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Learning Point for Clinicians

The prevalence of hepatopulmonary syndrome (HPS) is not clear yet. The diagnosis of hepatopulmonary can be masked by other co-morbidities and the non-specific presentation. Although its presence is associated with high mortality, this condition is reversible after liver transplant. Awareness of diagnostic criteria for HPS is crucial amongst physicians.

Background

The diagnosis of hepatopulmonary syndrome (HPS) is often missed due to lack of suspicion. Patients with a diagnosis of HPS can be allocated extra points on the liver transplant wait list to enhance the possibility of receiving an organ. We present a 64-year-old male presented for evaluation of worsening ascites and shortness of breath. The oxygen saturation was 89% in the supine position and 86% when sitting up or standing. After ruling out all the obvious causes of worsening dyspnea, ABG, PFTs and Echo bubble study was performed which confirmed diagnosis of HPS. Established diagnosis of HPS in these patient advocates bad prognosis but at the same time also gives them extra points in order to prioritize them on liver transplant list. This progressive phenomenon with ominous prognosis has become an indication for urgent liver transplantation, given its reversible nature. Therefore, awareness among healthcare providers and early diagnosis are crucial.

Case

We present a 64-year-old male patient with a known history of chronic obstructive pulmonary disease (COPD), sarcoidosis and liver cirrhosis secondary to non-alcoholic steatohepatitis (NASH). He came in for worsening ascites and dyspnea. Physical examination revealed blood pressure 100/64 and respiratory rate of 22 with a 2L baseline O_2. The oxygen saturation was 89% in the supine position and dropped to 86% when sitting up or standing. He had reduced air entry at the right lung base and a moderately distended abdomen secondary to ascites. Chest x-ray showed right-sided pleural effusion. Thoracocentasis was performed and a diagnosis hepatic hydrothorax was made after fluid analysis. Despite improvement in chest X-ray the next day the patient, however, remained dyspneic with increasing oxygen requirements. The patient's pulmonary function test (PFT) showed marked reduction in diffusion capacity of 36% of predicted. However, FEV1/FVC ratio was 70% and FEV1 was 58% which was unchanged from PFTs done 3 years before. Arterial blood gases in the sitting position showed primary respiratory alkalosis with arterial oxygen (PO_2) of 57.8 and A-a gradient of 60.05. Further evaluation included an echo bubble study, which revealed bubbles crossing from right to left

side during the fifth cycle suggesting right to left intrapulmonary shunt. By this, the diagnostic criteria for hepatopulmonary syndrome (HPS) were fulfilled including presence of portal hypertension with chronic liver disease, PaO_2<60, Increased A-a gradient > 15 and a positive bubble study and the diagnosis was made. He was discharged home with an increase in home oxygen therapy to 4L, and is instructed to follow-up with the GI clinic for further care and evaluation for liver transplant.

Discussion

HPS is characterized by a defect in oxygenation induced by pulmonary vascular dilatation in patients with liver disease.[1] This condition is diagnosed in 15–30% cirrhotic patients, but again, studies which derived the prevalence were mostly done at transplant centers.[2] Prospective multicenter prevalence studies have not been reported till date.

Our patient had a broad differential diagnosis possible for hypoxia. Hepatic hydrothorax, massive ascites, H/O COPD could be possible reasons for hypoxia. However, after ruling out all these causes, HPS was considered as a possible diagnosis. Certain signs which are very sensitive and specific for HPS include platypnea (dyspnea increases in upright position) and orthodeoxia (drop in saturation >4 mm hg in upright position)[4] can also point toward diagnosis of HPS.

The severity of liver disease as reflected by Model for End-stage Liver Disease (MELD) and Child–Pugh scoring system are not predictive of the occurrence of HPS.[3] In liver cirrhosis patients with HPS, the median survival is 24 months as compared with 87 months in patients without HPS. Also, 5-year survival rate in HPS patients is 23% as compared with 63% in patients without HPS in similar MELD stage liver disease.[4] Currently, the only proven treatment of HPS is Orthotopic Liver Transplant (OLT) and complete resolution of HPS occurs in most cases after OLT.[5] However, the time frame depends on the severity of the disease pre-transplant. The patients with pre-transplant PaO_2 <50 have shown to be having high post-OLT mortality.[6] Patients with clinical evidence of HPS and PaO_2 of <60 mm Hg on room air with no underlying lung disease can receive enhanced prioritization for organ allocation (UNOS).

Conclusion

It is imperative to have a high index of suspicion for HPS in a patient presenting with liver disease and hypoxia. This progressive phenomenon with ominous prognosis is rapidly reversible after liver transplantation. Awareness among healthcare providers and early diagnosis are crucial.

Conflict of interest: None declared.

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