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A case of unexplained exertional breathlessness diagnosed as Hepatopulmonary syndrome: a case report

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Abstract: *18 years old thin built girl from delhi was admitted to Indraprastha Apollo hospitals, New delhi with chief complaint of insidious onset breathlessness on exertion for 4-5 months. She also had low grade intermittent episode of fever, dry cough, weakness and fatigue for same duration. On physical examination, she was conscious, alert; Pulse-118; BP-90/60; Respiratory rate-26/min; Spo2-71% (On room air); digital clubbing present; Chest-bilateral vesicular breath sound equal; Abdomen- splenomegaly present. On investigations: arterial blood gas analysis showed type-1 respiratory failure. Liver function was deranged. Abdominal ultrasound and CT angiography report were suggestive of cirrosis of liver with splenomegaly. Pulmonary CT angiography showed pulmonary arterial dilatation. Diagnosis of hepatopulmonary syndrome was made. So, in any patient having unexplained exertional breathlessness with liver disease, this syndrome has to be thought.*
Keywords: *breathlessness, cirrhosis, angiography, hepatopulmonary syndrome*

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I. Introduction

Hepatopulmonary syndrome is the result of a complication of chronic liver disease seen in 16-24% of patients[1]. Its prevalence among cirrhotic patients varies from 4–47 percent. Prevalence depends on which diagnostic criterion is used. But the exact pathogenesis of this syndrome is still unknown. Patient presents with features of chronic liver disease and dypsnea of different types of severity[2].

II. Case report

18 yrs old thin built girl, student, thin build from delhi was admitted to Indraprastha Apollo hospitals, New delhi with chief complaint of insidious onset breathlessness on exertion for 4-5 months, mMRC grade-3 with no postural relationship. She also had low grade intermittent episode of fever, dry cough, weakness and fatigue for same duration. She denied any episode of chest pain, wheeze, hemoptysis, hematemesis, weight loss or any other problem. She had past history of pulmonary tuberculosis in childhood and enteric fever 1yr before. On physical examination, she was conscious, alert; Pulse-118; BP-90/60; Temperature-98.4; Respiratory rate-26/min; Spo2- 93% (on oxygen by nasal prong @3-4lit/m) and 71% (On room air); no significant palpable lymph node; digital clubbing present; Chest-bilateral vesicular breath sound equal; Abdomen- soft, scaphoid, non tender, splenomegaly present. On investigations: Arterial blood gas analysis (on oxygen by nasal prong @3lit/m)- 7.48/26.5/47/19.5/86.5%; Hemoglobin-13.2 gm%, WBC count-5300, Platelet-1.32 lakh/mm^3; Creatinine-0.4; Bilirubin [total/direct]-2.6/1; SGOT/SGPT-58/35; Hepatitis markers-negative; Chest Xray-normal (figure-1); USG abdomen was suggestive of chronic liver disease; fibroscan value- 8.1; 2D echocardiography: EF-60%. Abdominal CT angiography report was suggestive of cirrosis of liver with splenomegaly, large collaterals in paraoesophageal, retroesophageal and spleno renal regions. Pulmonary CT angiography showed pulmonary arterial dilatation with peripheral pulmonary artery branches reaching till the lung periphery. Based on these all findings, she was diagnosed to have hepatopulmonary syndrome.

III. Discussion

The unique pathological features that are viewed in autopsy are much dilatation of the pulmonary precapillary and capillary vessels and absolute increase in the number of dilated vessels[3]. Endothelin-1 is released from injured liver cells. As a result, expression of endothelin-B receptors is increased in pulmonary endothelial cells. Activation of these receptors upregulates the endothelium nitric oxide synthase enzyme and increases the production of nitric oxide. It causes vasodilation of vascular smooth muscle. In addition, increased phagocytosis of bacterial endotoxin in the lung activates nitric oxide synthase and increases nitric oxide production. Bacterial translocation and subsequent monocyte aggregation may also promote pulmonary angiogenesis in this disorder[4].

Hepatopulmonary syndrome is diagnosed based on a clinical triad of liver disease, increased alveolar-arterial oxygen gradient and evidence of intrapulmonary vascular dilatations[5]. Our patient had deranged liver function test, features of cirrhosis of liver in ultrasound plus CT abdomen and abnormal fibro scan report. Alveolar arterial oxygen gradient for our patient was abnormally high (Normal value: 5-20). Intrapulmonary vascular dilatations were seen in chest CT angiography findings. Hypoxaemia was due to an increase in alveolar-capillary gradient as a result of intrapulmonary vasodilatation.

Pulmonary angiography is an invasive procedure. It generally excludes alternative causes of hypoxemia, such as pulmonary embolism, pulmonary hypertension, large direct arteriovenous communications. Octavio et al case was diagnosed on echocardiography with agitated saline contrast which showed the patient having extra cardiac shunt. Scintigraphy using albumin macro-aggregates showed uptake in brain[6]. But we have found intrapulmonary vascular dilatations on CT pulmonary angiography. So, all of these three methods are equally useful for diagnosis.

IV. Conclusion

If any unexplained extertional breathless patient has associated liver disease, diagnosis of hepatopulmonary syndrome has to be placed in mind. CT pulmonary angiography is one of the effective methods to be useful for the diagnosis of this syndrome.

References

[1]. De Santis M, Vaz Rodrigues L, Matos F. *Hepatopulmonary syndrome: case report and literature review. Rev Port Pneumol. 2010 Jul-Aug;16(4):679-85.*

[2]. Jorge M. Hurtado-Cordovi, Seth Lipka, Jaspreet Singh, Ghulamullah Shahzad, and Paul Mustacchia. *Diagnostic Challenge of Hepatopulmonary Syndrome in a Patient with Coexisting Structural Heart Disease. Case Reports in Hepatology Volume 2011 (2011), Article ID 386709, 3 pages*

[3]. Rodríguez-Roisin R, Krowka MJ. *Hepatopulmonary syndrome--a liver-induced lung vascular disorder. N Engl J Med 2008; 358:2378.*

[4]. Josephine A Grace and Peter W Angus. *Hepatopulmonary syndrome:Update on recent advances in pathophysiology, investigation, and treatment. Journal of Gastroenterology and Hepatology 28 (2013) 213–219*

[5]. Michael J. Krowka, Michael J. Krowka. *Hepatopulmonary Syndrome. CHEST journal. May 1994Volume 105, Issue 5, Pages 1528–1537.*

[6]. Octavio Germán Muñoz Maya, Oscar Mauricio Santos, Paula Granda et al. *Case report of hepatopulmonary syndrome in patients with non-cirrhotic portal hypertension. 2012 Asociaciones Colombianas de Gastroenterología, Endoscopia digestiva, Coloproctología y Hepatología*

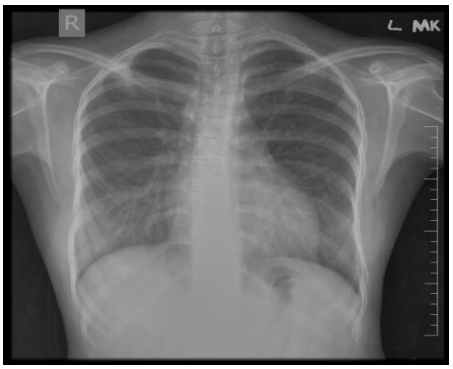

Figure: 1-Chest Xray

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