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Commentary

Improve the Awareness of Pulmonary Vascular Disease Associated with Liver Disease

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DESCRIPTION

Portopulmonary Hypertension (PoPH) and Hepatopulmonary Syndrome (HPS) are opposite pulmonary vascular complications of liver disease. Though once thought mutually exclusive, evidence now shows they can coexist. In China, the same liver disease can cause both conditions *via* unclear mechanisms. PoPH involves pulmonary remodeling and hypertension, while HPS causes intrapulmonary dilatation and shunting. Recent guidelines have raised awareness, but underdiagnosis persists. HPS requires transplantation, while PoPH uses pulmonary hypertension drugs with promising results. Further research on pathophysiology and multidisciplinary collaboration is key to improving diagnosis, enabling treatment access, and safely managing these diseases alongside liver transplantation.

Liver disease-related pulmonary vascular disease mainly refers to Portopulmonary Hypertension (PoPH) and Hepatopulmonary Syndrome (HPS), both of which are pulmonary vascular complications on the basis of portal hypertension and chronic liver disease respectively. The former is manifested as pulmonary small vessel remodeling and increased resistance, while the latter is characterized by Intrapulmonary Capillary Dilatations (IPVDs). Because the pathophysiological features of the two are completely opposite, it was thought that they could not coexist in the past. However, more and more clinical evidence confirms that they can co-exist in the same patient [1]. IPVDs, a fundamental feature of HPS, are common in PoPH. Fusnner found that IPVDs were detectable in 59% of patients with PoPH, while most patients had fewer pulmonary shingles and no significant hypoxemia [2].

In China, portal hypertension mostly occurs in patients with chronic liver disease, and the same disease can lead to opposite pathophysiological responses of pulmonary vessels, the exact mechanism of which is not clear. In 1990, the academic conference on pulmonary vascular diseases associated with liver disease was held at Mayo Clinic, Florida, USA, which was the first international conference involving liver disease and pulmonary vascular diseases. However, pulmonary vascular diseases associated with liver disease have not been paid sufficient attention. With the development of liver transplantation, the International Liver Transplant Society published guidelines for the diagnosis and management of HPS and PoPH in 2016 [3]. Since then, attention was gradually paid to pulmonary vascular complications related to liver disease.

At present, there is no effective treatment for HPS except liver transplantation. There are three main types of drugs for the treatment of pulmonary hypertension, which are endothelin receptor antagonists, 5-phosphodiesterase inhibitors, and prostacyclin analogs. Due to the background of chronic liver disease, there are few studies on the above three classes of drugs in patients with PoPH. The first randomized controlled phase IV clinical trial for PoPH was Portico Re-sheathable Transcatheter Aortic Valve System (PORTICO) study [4]. All three types of drugs relieve pulmonary hypertension by dilating the pulmonary artery, which, in theory, may aggravate pulmonary vascular shunt or hypoxemia [1]. But in the practice of our cases, it happens in the opposite way [5]. We hypothesized that the appearance of PoPH may promote the progression of IPVDs to relieve pulmonary artery pressure, and after PoPH is relieved, IPVDs are also relieved. If this hypothesis is true, then PoPH should have appeared before HPS. However, it is currently believed that the sequence of occurrence of HPS and PoPH is uncertain. In many case reports, it is more common for HPS to be diagnosed before PoPH or at the same time, and it is rare for PopH to be diagnosed before HPS [1].

Pulmonary vascular complications occur only when liver disease develops to a more severe degree. Therefore, pulmonary vascular diseases related to liver disease are often considered as an aggravation of the liver disease. They are very easily neglected by medical workers. As we reported in this case, the patient was hospitalized repeatedly for a long time. Each time, the patients were after supplemented with albumin and diuresis. After treatment, the hospitalization interval of the patients was significantly prolonged and the daily living endurance was significantly improved.

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Therefore, when patients with chronic liver disease have symptoms such as chest tightness, shortness of breath, edema, and syncope after activity, they should not simply be diagnosed as aggravated liver disease. Necessary physical and laboratory-based examinations (cardiac examination, arterial blood gas analysis, cardiac ultrasound, and electrocardiogram) should be conducted to avoid missed diagnosis of complications.

Due to the special liver disease background of PoPH patients, pulmonary arterial hypertension-related studies have mostly excluded patients complicated with liver injury or even PoPH patients. For patients with severe liver injury complicated with PoPH, the need for liver transplantation is more urgent. However, POPH is associated with increased perioperative risk at the time of LT, and PoPH is treated similarly to idiopathic PAH with PAH therapy. These therapies are associated with improved pulmonary hemodynamics and facilitation of safe LT [6]. Therefore, in the subsequent clinical experiment, we hope that PoPH patients with severe liver damage can be incorporated into clinical trials, so that they may have the opportunity to reduce pulmonary vascular resistance and liver transplantation.

Pulmonary vascular diseases associated with liver disease are two opposite complications. Whether the occurrence and development of PoPH and HPS promote each other and whether they are within the same pathological pathway are not clear at present. More research is needed to explore this intricate mechanism. The diagnosis and treatment of them require the multidisciplinary collaboration of the liver disease department, respiratory department, cardiovascular department, and interventional department. As a general hospital with a liver disease center, there are many patients with liver cirrhosis and portal hypertension in our hospital. At present, we are collecting relevant data on these patients and screening their pulmonary vascular-related complications. And in the near future, we can have a deeper understanding of pulmonary vascular diseases related to liver disease.

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REFERENCES

1. DuBrock HM, Krowka MJ. The myths and realities of porto-pulmonary hypertension. Hepatology. 2020;72(4):1455-1460.
2. Fussner LA, Iyer VN, Cartin-Ceba R, Lin G, Watt KD, Krowka MJ. Intrapulmonary vascular dilatations are common in portopulmonary hypertension and may be associated with decreased survival. Liver Transpl. 2015;21(11):1355-1364.
3. Krowka MJ, Fallon MB, Kawut SM, Fuhrmann V, Heimbach JK, Ramsay MA, et al. International liver transplant society practice guidelines: Diagnosis and management of hepatopulmonary syndrome and portopulmonary hypertension. Transplantation. 2016;100(7): 1440-1452.
4. Sitbon O, Bosch J, Cottreel E, Csonka D, De Groote P, Hoeper MM, et al. Macitentan for the treatment of portopulmonary hypertension (PORTICO): A multicentre, randomised, double-blind, placebo-controlled, phase 4 trial. Lancet Respir Med. 2019;7(7): 594-604.
5. Li N, Wu Q, Meng J, Feng C, Jiang S, et al. Macitentan treatment of portopulmonary hypertension with hepatopulmonary syndrome: A case report and literature review. ESC Heart Fail. 2023;10(4): 2718-2721.
6. DuBrock HM. Portopulmonary hypertension: Management and liver transplantation evaluation. Chest. 2023.