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Portopulmonary Hypertension with Liver Cirrhosis

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Abstract A 49-year-old male with a past medical history of smoking (20 pack-years), 25 years of daily drinking, type 2 diabetes, alcoholic liver cirrhosis, hyperlipidemia, and hypertension presented with a complaint of weakness, shortness of breath and dizziness. Three days prior the patient completed an echocardiogram that revealed tricuspid regurgitation with right ventricular dilation and a right ventricular systolic pressure of 116 mmHg. Cardiology was consulted and completed right heart catheterization, VQ scan, as well as autoimmune and infectious investigations. The patient was diagnosed with Portopulmonary Hypertension (PPHTN) complicated by liver cirrhosis. Treatment was initiated with sildenafil, amlodipine, aspirin and continuous oxygen with scheduled outpatient appointments for both IV prostacyclin treatment and pre-liver transplant (LT) evaluation.

Keywords: *Portopulmonary, hypertension, cardiology, cirrhosis, pulmonary*

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1. Background

Portopulmonary hypertension is a division of pulmonary arterial hypertension accompanied by portal hypertension (with or without liver disease). Recently (PPHTN) has been a leading cause of pulmonary arterial hypertension and carries a poor prognosis for the patient if concomitant liver disease is present. Standard of treatment is vasodilator therapy along with LT, which may improve survival of patients. [1]

2. Case Presentation

Our patient is a 49-year-old male with a past medical history of smoking (20 pack-years, quit 10 years ago), 25 years of daily drinking (quit 18 months ago), type 2 diabetes, alcoholic liver cirrhosis, hyperlipidemia, and hypertension. The patient had been experiencing generalized weakness for over a month before seeing his PCP. At this appointment, an echocardiogram showed severe tricuspid regurgitation, right ventricular systolic pressure (RVSP) of 116 mmHg and right ventricular dilation. He was informed to go to an emergency department if any of his symptoms progressed. One week later, the patient became short of breath and experienced dizziness, prompting him to visit the closest ED at Owosso Memorial Hospital. On arrival, he was hypertensive and tachypneic with oxygen saturations dropping into the low 80s, he appeared to be fluid overloaded with lower extremity edema an ascites on exam. He was placed on 2L oxygen via nasal cannula and oxygen saturation subsequently improved. EKG demonstrated right axis deviation; however, troponin levels were within normal limits and the patient denied chest pain. CTA and chest x-ray were unremarkable. After review of the patient's recent echocardiogram, the patient was transferred to Sparrow Hospital for further management. Upon arrival, evaluation was concerning for pulmonary arterial hypertension. The patient was continued on oxygen and placed on aspirin 81 mg and amlodipine 2.5 mg. Cardiology was consulted at admission.

3. Investigations

Previous Echocardiogram- severe tricuspid regurgitation, RVSP 116, right ventricular dilation
EKG- Right Axis Deviation
Chest X-ray- Unremarkable
CTA- no pulmonary embolism
BNP 292 pg/mL
HDL 22 mg/dL
Direct Bilirubin 1.4 mg/dL
Hepatitis A, B, C panel negative
ANCA Panel negative
dsDNA ab, Anti-SCL-70, ANA, Anti-RNP, RF negative
HIV negative
VQ with low PE probability
Alpha 1 Anti-Trypsin 219 mg/dL
PFTs with DLCO- FEV1 57% of predicted, FVC 55% of predicted, FEV1/FVC ratio 75% of predicted However,

the total lung capacity is normal. Diffusion capacity 88% of predicted. Features of obstruction and restriction impairments.

Right heart catherization with nitric oxide challenge-precapillary pulmonary hypertension; mean pulmonary artery pressure 53 mmHg, normal pulmonary arterial wedge pressure 6 mmHg

4. Differential Diagnosis

There are many possible etiologies for PPHTN and there are 5 identified groups for pulmonary hypertension. The patient's normal pulmonary arterial wedge pressure (6 mmHg) refutes group 2 for left heart disease. Lung disease was of concern, however PFTs and DLCO showed features of obstruction and restriction not meeting criteria for COPD, eliminating group 3. The CTA and VQ were unremarkable, ruling out group 4. Our patient was found to not have HIV, schistosomiasis, congenital heart disease, drug or toxin induced PAH, or a family history of heritable pulmonary arterial hypertension, covering most of group 5. The patient's normal pulmonary arterial wedge pressure and increased mean pulmonary artery pressure are most consistent with group 1.

5. Outcome and Follow-up

Our patient is undergoing IV prostacyclin therapy and awaiting liver transplant.

6. Discussion: *Include a very Brief Review of Similar Published Cases*

The pathophysiology of PPHTN is still not well described. It is seen in both non-cirrhotic and cirrhotic portal hypertension and it not related to the severity of liver disease. [2] Female sex and autoimmune disease are risk factors. [3] There are 2 diagnostic criteria for PPHTN: 1. Portal hypertension (>15 mmHg, or portocaval gradient > 5 mmHg) and 2. Mean pulmonary artery pressure > 25 mmHg and mean pulmonary artery occlusion pressure < 15 mmHg, transpulmonary gradient > 10 mmHg, pulmonary vascular resistance > 240 dyn.s.cm-5 = 3 UI WOOD. (PPHTN comprises of 6% of all LT candidates. [4]

To avoid worsening of hemodynamics, patients with PPHTN should not be given beta-blockers. Anticoagulants should be avoided as well due to an increased risk of bleeding in these patients. [5] Medical treatment of PPHTN includes prostacyclin pathway agonists, endothelin receptor antagonists, nitric oxide-cyclic guanosine monophosphate enhancers, phosphodiesterase inhibitors and guanylate cyclase stimulators. Definitive treatment is LT, which may allow some patients to wean off medical treatment entirely.[6]

Onoe et al reported a 63-year-old female with alcoholic cirrhosis who was later diagnosed with PPHTN and treated with tadalafil and LT. Their patient experienced a rapid improvement after LT and was weaned off phosphodiesterase type 5 enzyme inhibitor therapy. [7]

Zopey and coauthors completed a three patient case series illustrating patients transitioning from hepatopulmonary syndrome to portopulomnary hypertension. Their first patient was a 57-year-old female with cirrhosis secondary to nonalcoholic steatohepatitis. She presented with a two-year history of shortness of breath, and during the same hospitalization the patient was diagnosed with hepatopulmonary syndrome. Seven years later, the patient underwent a routine transthoracic echocardiogram showing RVSP of 62 mmHg after a right heart catheterization (PPHTN) was diagnosed and began treprostinil treatment. The second and third cases describe a 63-year-old and 56-year-old men with hepatitis C and later diagnosed with PPHTN. The second case received a LT with subsequent resolution of symptoms, while the third patient was not a candidate for LT due to PPHTN severity and was treated with ambrisentan in hopes of improving PPHTN for LT reevaluation. [8]

7. Learning Points/Take Home Messages *3-5 Bullet Points*

1. Portopulmonary Hypertension is a type of pulmonary arterial hypertension along with portal hypertension with or without liver disease. 10% of PPHTN patients have portal hypertension without cirrhosis.
2. Female sex and autoimmune disease are risk factors for PPHTN.
3. PPHTN comprises 6% of liver transplant candidates.
4. Right Heart catheterization is recommended to confirm diagnosis of PPHTN.

References

[1] Saleemi S. Portopulmonary hypertension. Ann Thorac Med. 2010; 5(1): 5-9.

[2] Aldenkortt F, Aldenkortt M, Caviezel L, Waeber JL, Weber A, Schiffer E. Portopulmonary hypertension and hepatopulmonary syndrome. World J Gastroenterol. 2014; 20(25): 8072-8081.

[3] Kawut SM, Krowka MJ, Trotter JF, et al. Clinical risk factors for portopulmonary hypertension. Hepatology. 2008; 48(1): 196-203.

[4] Colle, I. O., Moreau, R., Godinho, E., Belghiti, J., Ettori, F., Cohen‐Solal, A., Mal, H., Bernuau, J., Marty, J., Lebrec, D., Valla, D. and Durand, F. (2003), Diagnosis of portopulmonary hypertension in candidates for liver transplantation: A prospective study. Hepatology, 37: 401-409.

[5] Provencher S, Herve P, Jaı¨s X, Lebrec D, Humbert M, Simonneau G, Sitbon O. Deleterious effects of beta-blockers on exercise capacity and hemodynamics in patients with portopulmonary hypertension. Gastroenterology 2006; 130: 120-126.

[6] Does Portopulmonary Hypertension Impede Liver Transplantation in Cirrhotic Patients? A French Multicentric Retrospective Study. Reymond M, Barbier L, Salame E, Besh C, Dumortier J, Pageaux GP, Bureau C, Dharancy S, Vanlemmens C, Abergel A, Woehl Jaegle ML, Magro P, Patat F, Laurent E, Perarnau JM Transplantation. 2018; 102(4): 616.

[7] Takashi Onoe, Asuka Tanaka, Kohei Ishiyama, Kentaro Ide, Hirotaka Tashiro & Hideki Ohdan Surgical Case Reports volume 4, Article number: 15 (2018).

[8] Radhika Zopey, Irawan Susanto, Igor Barjaktarevic, and Tisha Wang, "Transition from Hepatopulmonary Syndrome to Portopulmonary Hypertension: A Case Series of 3 Patients," Case Reports in Pulmonology, vol. 2013, Article ID 561870, 5 pages, 2013.

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