An Unusual Case of Cirrhosis

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1. Introduction

Constrictive pericarditis is a rare but severely disabling consequence of the chronic inflammation of the pericardium, leading to an impaired filling of the ventricles and reduced ventricular function [1]. The timely diagnosis of a cardiac etiology of liver dysfunction is important because such dysfunction is potentially reversible if the underlying cardiac disease is treated before the development of frank cirrhosis [2, 3].

Below, we present a case of a 49-year-old female who was incidentally found to have cirrhosis. Initial workup was negative. Thoracic imaging showed pericardial calcifications which ultimately led to the diagnosis of constrictive pericarditis. We will briefly discuss the literature on cardiac causes of liver cirrhosis.

2. Case Report

49-year-old white female with remote h/o sarcoidosis was referred to GI when her liver was noted to be nodular during laparoscopy for an ovarian cyst. She denied fatigue, vomiting-up blood, abdominal distension and pain, ankle swelling, itching, yellow discoloration of skin and eyes, and episodes of confusion or sleepiness. She denied alcohol abuse. Physical examination revealed normal vital signs and no icterus, spider nevi, clubbing, ascites, hepatosplenomegaly, or ankle edema. LFTs, hepatitis serologies, ANA, AMA, ASMA, Ferritin, Ceruloplasmin, and α1-AT, level were unremarkable. Liver biopsy showed cirrhosis. She developed worsening baseline SOB and was hospitalized. She was eventually diagnosed with constrictive pericarditis. A diagnosis of cardiac cirrhosis was made.

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pericarditis. We believe that constrictive pericarditis resulted from sarcoidosis. She is now being evaluated for pericardiectomy.

3. Discussion

Our case illustrates the importance of considering a cardiac etiology in the work-up of cirrhosis especially when the most common causes are not found. Classically, serum albumin is normal unless frank cirrhosis has developed; ascitic fluid analysis reveals an elevated serum albumin-ascites gradient (>1.1 g/dL) typical of portal hypertension and demonstrates an elevated total protein level (>2.5 g/dL) [4]. In our case, the LFTs were only mildly abnormal with a mildly elevated PT and mildly elevated alkaline phosphatase. The finding of a nodular liver was incidental and no primary liver disease was found. Sinusoidal dilatation indicating congestive hepatopathy helped steer us towards a cardiac etiology; other findings on liver biopsy that may be found include sinusoidal degeneration, variable degrees of hemorrhagic necrosis in zone 3, fatty change, and variable degrees of cholestasis [4]. In general, patients with passive hepatic congestion do not have stigmata of portal hypertension such as spider angiomata or evidence of portosystemic shunts such as caput medusa [4]. Patients with constrictive pericarditis, however, may develop ascites. Nevertheless, such findings were absent in our patient.

Constrictive pericarditis is a rare but severely disabling consequence of the chronic inflammation of the pericardium, leading to an impaired filling of the ventricles and reduced ventricular function [1].

Establishing the diagnosis of constrictive pericarditis and secondary congestive hepatopathy/cirrhosis remains a challenge. One study reported a median delay in diagnosis of greater than 10 years [5]. In another case, the diagnosis of constrictive pericarditis was made only after liver transplantation was performed for presumed cryptogenic cirrhosis [6].

Much of the difficulty in diagnosing constrictive pericarditis can be attributed to its insidious course and the fact that some of the symptoms and signs resulting from it, that is, dyspnea and ascites, can be mistakenly thought to result from primary liver dysfunction. Dyspnea in a patient with liver dysfunction usually makes one suspects hepatopulmonary syndrome, portopulmonary hypertension, cardiomyopathy, hepatic hydrothorax, ascites, or anemia, whereas ascites itself is a hallmark of liver disease complicated by portal hypertension. Constrictive pericarditis is curable and should be considered in all cases of unexplained cirrhosis, regardless of atypical hepatic histology [7].

Pericardial involvement is uncommon in sarcoidosis even in the presence of extensive myocardial infiltration. It is observed in fewer than 10% of patients with cardiac sarcoidosis [8]. Pericardial calcification should provide a clue to the diagnosis of constrictive pericarditis (see Figure 1). One case study demonstrated the importance of this finding on low-cost imaging to help point towards this diagnosis [9]. The major differential of constrictive pericarditis is restrictive cardiomyopathy which is more common in patients with sarcoidosis. Although echocardiography and cardiac CT/MR might distinguish one from the other, the most specific finding differentiating constrictive pericarditis from restrictive cardiomyopathy is demonstrated on heart catheterization. Simultaneous measurement of the left and right heart pressures demonstrates respiratory variation in ventricular filling and increased ventricular interdependence along with increased atrial pressures and equalization of end-diastolic pressures [10].

The timely diagnosis of a cardiac etiology of liver dysfunction is important because such dysfunction is potentially reversible if the underlying cardiac disease is treated before the development of frank cirrhosis [2, 3]. Moreover, early treatment of underlying cardiac disease might also prevent the development of hepatocellular carcinoma as suggested by an interesting case study in which a patient with negative hepatitis serologies and cirrhosis secondary to constrictive pericarditis developed hepatocellular carcinoma confirmed by biopsy [11].

4. Conclusion

This case study illustrates to gastroenterologists the need to consider a cardiac etiology in the work-up of cirrhosis especially when the most common causes are not found and if liver biopsy shows evidence of congestive hepatopathy and is inconclusive for primary liver disease. The presence of pericardial calcification in a patient with cirrhosis should suggest the possibility of constrictive pericarditis and cardiac cirrhosis and prompt cardiac MR/CT imaging and cardiac catheterization. Simultaneous invasive measurement of the pressures in the right and left heart chambers by an experienced cardiologist is the key to making the diagnosis. Constrictive pericarditis is a rare but well-known manifestation of sarcoidosis. Prompt referral to a thoracic surgeon for pericardiectomy is important once the diagnosis is made to prevent progression to cirrhosis.

Conflicts of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

References

[1] B. Maisch, P. M. Seferović, A. D. Ristić et al., “Guidelines on the diagnosis and management of pericardial diseases executive
summary; The Task force on the diagnosis and management of pericardial diseases of the European society of cardiology," European Heart Journal, vol. 25, no. 7, pp. 587–610, 2004.

[2] F. Heureux, L. Frankart, B. Marchandise, M. Buche, J. P. Martinet, and J. Donckier, “Recurrent ascites: two case reports,” Acta Clinica Belgica, vol. 52, no. 3, pp. 176–181, 1997.

[3] A. A. Sheth and J. K. Lim, “Liver disease from asymptomatic constrictive pericarditis,” Journal of Clinical Gastroenterology, vol. 42, no. 8, pp. 956–958, 2008.

[4] C. C. Giallourakis, P. M. Rosenberg, and L. S. Friedman, “The liver in heart failure,” Clinics in Liver Disease, vol. 6, no. 4, pp. 947–967, 2002.

[5] S. van der Merwe, J. Dens, W. Daenen, V. Desmet, and J. Fevery, “Pericardial disease is often not recognised as a cause of chronic severe ascites,” Journal of Hepatology, vol. 32, no. 1, pp. 164–169, 2000.

[6] P.-H. Bernard, P. L. Metayer, B. L. Bail, C. Balabaud, J. Saric, and P. Bioulac-Sage, “Liver transplantation and constrictive pericarditis,” Gastroenterologie Clinique et Biologique, vol. 25, no. 3, pp. 316–319, 2001.

[7] M. Kirsch and B. Fleshler, “Deceptive liver histology delays diagnosis of cardiac ascites,” Southern Medical Journal, vol. 85, no. 11, pp. 1151–1152, 1992.

[8] V. Sekhri, S. Sanal, L. J. DeLorenzo, W. S. Aronow, and G. P. Maguire, “Cardiac sarcoidosis: a comprehensive review,” Archives of Medical Science, vol. 7, no. 4, pp. 546–554, 2011.

[9] M. Toledano and A. Bhagra, “Pericardial calcification in constrictive pericarditis,” International Journal of Emergency Medicine, vol. 5, no. 37, 2012.

[10] M. H. Khandaker, R. E. Espinosa, R. A. Nishimura et al., “Pericardial disease: diagnosis and management,” Mayo Clinic Proceedings, vol. 85, no. 6, pp. 572–593, 2010.

[11] P. S. Song, K. C. Koh, B. C. Yoo et al., “A case of hepatocellular carcinoma complicating cardiac cirrhosis caused by constrictive pericarditis,” The Korean Journal of Gastroenterology, vol. 45, no. 6, pp. 436–440, 2005.