Case Report

A case report of constrictive pericarditis: a forgotten cause of refractory ascites

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ABSTRACT

Constrictive pericarditis is well known but rare and commonly forgotten cause of ascites. Early diagnosis of constrictive pericarditis is difficult due to absence of typical cardiopulmonary signs and multiple vague symptoms and its insidious course. In this case report, we present, a 61-year-old male referred for liver transplantation vs transjugal intrahepatic portosystemic shunt work-up for presumptive diagnosis of nonalcoholic steatohepatitis cirrhosis and refractory ascites. Comprehensive work-up before liver transplantation including liver biopsy, liver ultrasound, and Doppler, magnetic resonance imaging was not consistent with liver cirrhosis. Echocardiographic was suggestive of constrictive pericarditis, further work-up with right heart catheterization, cardiovascular magnetic resonance and multidetector cardiac computed tomography confirmed the diagnosis of constrictive pericarditis. Patient underwent surgical pericardiectomy, he remained stable after surgery and did not require further paracentesis and discharged in stable condition.

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Introduction

Constrictive pericarditis characterized by the presence of rigid, thick noncompliant, fibrotic, and/or calcific pericardium. Clinically constrictive pericarditis characterized by congestive heart failure, advanced stage hepatic congestion leads to congestive hepatomegaly and formation of ascites [1], constrictive pericarditis is well known but rare and commonly forgotten cause of ascites. Early diagnosis of constrictive pericarditis is difficult due to absence of typical cardiopulmonary signs and symptoms and its insidious course. Patient with constrictive pericarditis can presents with nonspecific symptoms related to heart failure, lung disease, and liver disease. The difficulties in differentiating primary liver disease from pericardial disease have been described over several decades ago [2]. Diagnosis delay of constrictive pericarditis to 10 years has been reported, and patients have undergone liver transplantation before the diagnosis was established [3,4].

Case presentation

A 61-year-old male referred to our hospital for liver transplantation vs transjugal intrahepatic portosystemic shunt for
The presumptive diagnosis of nonalcoholic steatohepatitis cirrhosis and refractory ascites. He was diagnosed with liver inflammation and cholestasis suggestive of stage 3 liver fibrosis 3 years before his current referral. Patient presented with severe refractory ascites, he underwent multiple large volume paracentesis. He is known to have other medical problems including hypertension, hypothyroidism, and chronic kidney disease. At our institution, multiple routine work-up before liver transplantation including repeat liver biopsy, ultrasound of the liver and portal vein duplex, and liver magnetic resonance imaging, excluded liver cirrhosis, no collaterals vessels, nor focal hepatic lesions, liver biopsy revealed severe cholestasis with portal inflammation and fibrosis and perisinusodial fibrosis with no evidence of cirrhosis. Echocardiography was performed due to severe shortness of breath and revealed several signs of constrictive pericarditis such as diastolic septal bounce, exaggerated left ventricle-right ventricle interdependence with inspiratory shift of interventricular septum, and other signs of constrictive pericarditis (Fig. 1A and B). Multidetector cardiac computed tomography (MDCT) of the heart without intravenous iodinated contrast revealed abnormal pericardial thickening with maximum thickness of 5.4 mm (maximum thickness of 2 is considered the upper limit of normal) with scattered foci of calcification (Fig. 2). For further confirmation of constrictive pericarditis, cine and black blood imaging of the heart cardiovascular magnetic resonance (CMR) of the heart were consistent with constrictive pericarditis (Fig. 3A and B). Right side heart catheterization revealed square root sign and ventricular interdependence with respiration. Subsequently, patient underwent surgical pericardectomy, at surgery, heart was found to be constrained inside the layers of thick fibrotic, calcific pericardium, the pericardium was successfully and completely dissected, the central venous pressure went from 25 cm H2O before surgery to 9 cm H2O immediately after surgery. He was transferred to surgical intensive care unit for postoperative management, there were no complications and subsequently patient was transferred to medical floor. The patient remained stable and did not require any further paracentesis and discharged home in stable condition on regular medications for his other medical problems.

Discussion

This case demonstrated the difficulty and delay in the diagnosis and management of patient with constrictive pericarditis presented with chronic severe refractory ascites initially misdiagnosed as ascites related to nonalcoholic steatohepatitis cirrhosis. The relationship between constrictive pericarditis and hepatic dysfunction and ascites is well recognized, typically clinical presentation of constrictive pericarditis related
to fluid overload evident by elevated jugular venous pressure, pleural effusions, lower extremity edema, ascites, and pulsatile liver. Prior cases series of constrictive pericarditis presented with, 75% of patients presented with peripheral edema, 60% with hepatomegaly, and 40% with ascites [5,6], the most important consistent finding in patients with constrictive pericarditis is elevation of jugular venous pressure (JVP) that reported to be present in more than 80% of cases, this is most important clinical sign in patients with constrictive pericarditis because it is not typically seen in patients with chronic liver disease.

Diagnosis of constrictive pericarditis is difficult because patients presented with vague and multiple symptoms. The most important key in the diagnosis is considering it as a differential diagnosis and undertaking careful history and comprehensive physical examination, as previously mention the most important consistent clinical finding is elevation of JVP, elevation of JVP can be missed if elevation more than the angle of the jaw, to avoid this pitfall patient should be placed in upright position to accurately determine the top of JVP. Plain chest radiography may reveal pericardial calcification, however, this finding is not specific nor does it confirm constrictive physiology, only 25% of patients with constrictive pericarditis will have pericardial calcification on chest radiograph [7]. Transthoracic echocardiography (TTE) should be the starting investigation if constrictive pericarditis is suspected [8], there is no, however, single pathognomonic finding to diagnose constrictive pericarditis, but there are several findings, common 2-dimensional echocardiographic findings include; right and left atrial dilation, dilated noncollapsing inferior vena cava and thickened pericardium, diastolic septal bounce and respirophasic septal shift over several cycles support the diagnosis of ventricular interdependent. The key benefit of echocardiography in assessment of suspected constrictive pericarditis relies on its ability to provide detailed hemo-

dynamic assessment, in constrictive pericarditis significant respiratory variation is evident in the inflow profile across the mitral and bicuspid valve [9], on inspiration, the peak mitral E wave velocity decreases by using pulsed-wave Doppler analysis, at the onset of expiration, the peak tricuspid E wave velocity decreases by ≥40%. Tissue Doppler imaging and advanced echocardiographic imaging such as strain imaging can be very helpful in diagnosis of constrictive pericarditis [10], echocardiographic proposed diagnostic criteria have diagnostic sensitivity 87% and specificity 91% [11]. MDCT should not be used as the first imaging modality for constrictive pericarditis, however, in certain clinical scenarios such as end-stage calcific pericardial constrictions, MDCT is essential in the evaluation of location and extent of pericardial calcification [12], also MDCT is very helpful in patients with prior cardiac surgery and radiation heart disease because it provides assessment of parenchymal lung disease and proximity of cardiovascular structures to the sternum. It should be emphasis that absence of pericardial thickening on MDCT does not not exclude constrictive pericarditis [13]. CMR is highly sensitive for di-
agnosis of constrictive pericarditis, anatomical findings such as pericardial thickening can be evaluated with T1-weighted black blood imaging [14]. T2-weighted short tau inversion recovery can be used to assess pericardial edema. CMR can provide useful complementary role of echocardiography to visualize early septal flattening which is a feature of constrictive physiology by real time cine imaging with free breathing [15], also, although not frequently performed, CMR utilizing real-time phase contrast imaging can be performed to assess hemodynamic assessment of constrictive pericarditis pathophysiology, when >25% variation in transmitial flow during respiration [16], in addition CMR can provide a helpful preoperative evaluation of the pericardium and cardiac anatomy.

In summary, this case demonstrated chronic unrecognized constrictive pericarditis as the cause of chronic severe ascites. Clinicians need to maintain a high index of suspicion for constrictive pericarditis when evaluating patients with unexplained ascites. The most consists clinical finding is detection of elevated JVP. Echocardiography is the initial diagnostic test of choice in evaluating patients with suspected constrictive pericarditis, even if echocardiography is reported being unremarkable, constrictive pericarditis may still be a possibility. The complementary role of MDCCT and CMR can often provide diagnostic certainty to confirm the diagnosis and initiate treatment.

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