

Constrictive Pericarditis as a Cause of Refractory Ascites

Zurabi Lominadze, MD¹, Leila Kia, MD², Sanjiv Shah, MD³, Keyur Parekh, MD⁴, and Josh Levitsky, MD, MS^{2,5}

¹Department of Medicine, Northwestern University Feinberg School of Medicine, Chicago, IL

²Division of Gastroenterology and Hepatology, Department of Medicine, Northwestern University Feinberg School of Medicine, Chicago, IL

³Division of Cardiology, Department of Medicine, Northwestern University Feinberg School of Medicine, Chicago, IL

⁴Department of Radiology, Northwestern University Feinberg School of Medicine, Chicago, IL

⁵Division of Organ Transplantation, Department of Surgery, Northwestern University Feinberg School of Medicine, Chicago, IL

Abstract

We report a 43-year-old man who presented for evaluation of ascites, varices, and hepatosplenomegaly. Initial labs were notable for normal platelets, mild liver synthetic dysfunction, and disproportionately elevated alkaline phosphatase. He was presumed to have underlying cirrhosis, and diuresis was attempted without success. A transjugular liver biopsy showed marked sinusoidal dilation without cirrhosis. Diagnostic paracentesis revealed fluid studies suggestive of cardiac ascites. Further cardiac evaluation confirmed constrictive pericarditis. The case highlights the importance of considering a broad differential in the evaluation of ascites.

Introduction

The diagnosis of constrictive pericarditis as a cause of ascites requires a high degree of suspicion because cardiac etiologies of ascites are rare and easily overlooked. There is often a delay in diagnosis, and there are reported cases describing patients treated for juvenile cryptogenic cirrhosis, hepatitis B virus-induced cirrhosis, and tuberculosis for up to 10 years before constrictive pericarditis was ultimately diagnosed.^{1,2}

Case Report

A 43-year-old healthy man presented with abdominal distention. The patient recalled having a severe upper respiratory tract infection preceding the onset of abdominal distention. Initial evaluation revealed elevated alkaline phosphatase (407 U/L) and bilirubin (2.5 mg/dL), ascites and hepatosplenomegaly on computed tomography (CT), and portal hypertensive gastropathy on upper endoscopy. Evaluation for underlying liver disease, including transaminases, anti-nuclear antibody, anti-smooth muscle antibody, anti-mitochondrial antibody, alpha-1 anti-trypsin, and iron studies, was normal. INR was 1.4 and the platelet count was 157,000/ μ L. He had previously been a heavy binge alcohol drinker (a case of beer per day on weekends), but had recently quit.

He began treatment for presumed end-stage liver disease related to alcohol use. Physical exam was notable for muscle wasting, abdominal distention due to ascites, lower extremity edema, and palmar erythema without spider angiomas. Diuretics were continued and evaluation was begun for possible liver transplantation. Abdominal MRI showed sequelae of portal hypertension, but no liver lesions, steatosis, or cirrhosis. The patient continued to have refractory ascites despite adherence to a low-sodium diet and diuretics, requiring frequent large-volume paracenteses. A transjugular liver biopsy showed marked sinusoidal dilatation without fibrosis, steatosis, or granulomas.

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Correspondence: Josh Levitsky, Northwestern University Feinberg School of Medicine, Division of Gastroenterology and Hepatology, Department of Medicine, 676 N. St. Clair Street, 19th Floor, Chicago, IL, 60611 (j-levitsky@northwestern.edu).



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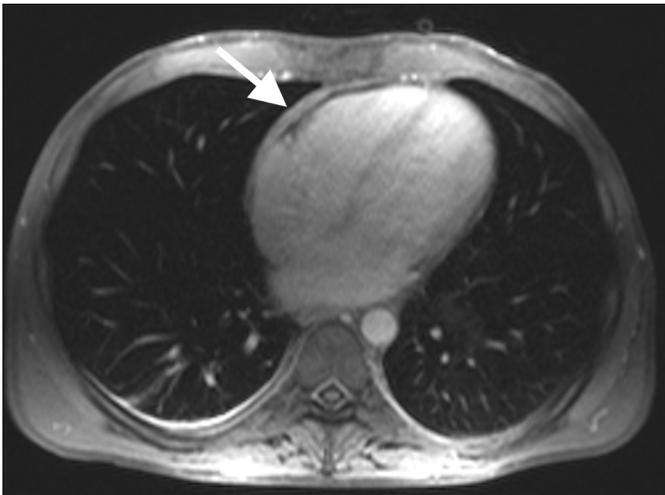


Figure 1. Cardiac MRI axial view of the heart with thickened pericardium.

Pressure measurements showed a portosystemic gradient of 1 mm Hg, with markedly elevated right atrial and inferior vena cava pressures (25 mm Hg). Ascitic fluid revealed an elevated serum-ascites albumin gradient (SAAG) of 1.6 g/dL, with a fluid albumin level of 2.7 g/dL. The fluid total protein level was also elevated at 4.4 g/dL. Cytology was negative for malignancy. Based on the fluid studies, a cardiac etiology for his ascites was suspected. Repeat physical exam demonstrated an elevated jugular venous pressure (JVP) of 14-16 cm H₂O, along with Kussmaul's sign (a paradoxical increase in JVP with inspiration). An echocardiogram showed a left ventricular ejection fraction of 50-55%, a prominent diastolic interventricular septal bounce, elevated right atrial pressure, and biatrial enlargement.

Cardiac MRI demonstrated pericardial thickening and a diastolic interventricular septal bounce that became exaggerated during inspiration, consistent with constrictive pericarditis (Figure 1 and Video 1). Right heart catheterization showed equalization of right ventricular (RV) and left ventricular (LV) diastolic pressures, a dip-and-plateau pattern in the RV and LV pressure tracings, and discordance of the peak RV and LV pressures during inspiration, consistent with constrictive physiology. The patient underwent pericardiectomy, his ascites dramatically improved, and he no longer required diuretics. Pericardial histological examination was notable for fibrosing pericarditis with chronic perivascular lymphoplasmacytic inflammation with scattered reactive lymphoid follicles. Cultures for bacteria, fungi, and mycobacteria were all negative. The patient remained symptom-free, and at last follow-up over 1 year following his procedure, he denied recurrent ascites.

Video 1. Diastolic interventricular septal bounce that became exaggerated during inspiration, consistent with constrictive pericarditis. Please view the video at: <http://acgcasereports.gi.org/2015/04/09/constrictive-pericarditis>.

Discussion

Our patient's history of alcohol use and the marked ascites and muscle wasting on physical exam led to a nearly 18-month delay in diagnosis of constrictive pericarditis. The SAAG was high, consistent with both possible liver disease and ascites praecox. However, the isolated elevated alkaline phosphatase, relatively intact liver synthetic function, and liver biopsy consistent with congestive hepatopathy suggested a cardiac cause of his illness. The most suggestive feature of an underlying cardiac etiology was the elevated ascites fluid total protein level. With a high SAAG, this finding is strongly correlated with cardiac ascites, but is usually absent in cardiac cirrhosis, which is associated with lower ascites fluid protein levels.³ Assessment of JVP in such patients is critical; careful examination of the patient's neck veins in the upright position during respiration and the finding of Kussmaul's sign may have led to an earlier diagnosis of constrictive pericarditis.

The differential diagnosis for cardiac ascites includes right-sided heart failure (constrictive pericarditis, restrictive cardiomyopathy, pulmonary arterial hypertension, or primary causes of RV failure such as arrhythmogenic RV dysplasia), right atrial myxoma, tricuspid valve dysfunction, or inferior vena cava obstruction.⁴ The most challenging of these to differentiate are constrictive pericarditis and restrictive cardiomyopathy. A history of previous pericarditis, trauma, cardiothoracic surgery, chest irradiation, or connective tissue disease is common in constrictive pericarditis, but rare for isolated restrictive cardiomyopathy.⁵ Using echocardiography, one may see biatrial enlargement, biventricular hypertrophy, abnormal ventricular relaxation, and absence of respiratory variation in mitral and tricuspid inflow in restrictive cardiomyopathy. In constrictive pericarditis, ventricular relaxation is normal or exaggerated, and a prominent diastolic septal bounce (due to ventricular interdependence) and increased respiratory variation in the mitral and tricuspid inflow are present.⁴ Cardiac MRI and CT allow visualization of pericardial thickening, inflammation, and calcification that cannot be reliably visualized by transthoracic echocardiography.⁵ Simultaneous right and left heart catheterization remains the gold standard for confirmation of constrictive physiology.

Once constrictive pericarditis is diagnosed, urgent pericardiectomy is the treatment of choice. Fortunately, our patient had an uneventful recovery, though prolonged delays in treatment can lead to myocardial atrophy or dysfunction that is irreversible even after pericardiectomy.⁵ In a cohort study of 163 patients, Bertog et al reported an approximate 6% perioperative mortality, with patients diagnosed with idiopathic or post-viral constrictive pericarditis having the best prognosis. The 7-year survival for these patients is about 88%.⁶

Disclosures

Author contributions: Z. Lominadze wrote the manuscript. L. Kia edited the manuscript. S. Shah and K. Parekh edited the manuscript and interpreted the images. J. Levitsky is the article guarantor.

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Informed consent was obtained for this case report.

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