CLINICAL VIGNETTE

Cardiac Cirrhosis: An Uncommon Cause of Cirrhosis in the Community

Scott Hahm, MD

A 73-year-old African American male presented with abdominal distension. He reported his abdomen became distended after a dual-chamber cardiac pacemaker was placed 2 months prior. He has severe peripheral edema prior to pacemaker placement. His edema was severe despite maximum dosages of diuretics with lower extremity skin ulceration, weeping fluid. The patient had wild-type transthyretin cardiac amyloidosis, TTR, congestive heart failure, and atrial flutter status post ablations x2 on chronic apixaban anticoagulation. He also has chronic kidney disease. He is a former football player. He denied alcohol. Abnormal labs at the time of his consultation included: Creatinine 1.76, AST 71, Alkaline phosphatase 148, total bilirubin 1.4, albumin 3.5. He underwent chronic liver disease evaluation which revealed only an anti-smooth muscle antibody titer of 1:40. Ultrasound guided paracentesis removed 4L of fluid, with ascitic fluid analysis supporting a diagnosis of cirrhosis. A subsequent liver biopsy showed hepatic congestion with extensive fibrosis and patchy regenerative change consistent with diagnosis of cardiac cirrhosis. There was no amyloidosis on the congo red stain of his liver biopsy. MELD score was 17 and his Child Pugh score was child class B. He was subsequently referred to transplant cardiology for evaluation.

Discussion

Cardiac cirrhosis is an uncommon form of liver cirrhosis that is caused by cardiac dysfunction, primarily when the chambers of the right side of the heart fail. Cardiac cirrhosis is more common in men and in patients with prior Fontan procedure for correction of heart defects. Right sided heart failure leads to venous congestion and an increase in venous pressure in the hepatic sinusoids. Increasing cardiac pressures, low cardiac output or impaired perfusion contribute to hepatic congestion. Hepatic congestion causes intrahepatic edema resulting in decreased perfusion and oxygen diffusion. Hepatic congestion also can result in hemorrhagic injury, hepatocyte atrophy with associated collagen deposition and ultimately fibrosis. The leading causes of cardiac cirrhosis include: valvular disease, severe pulmonary hypertension, cor pulmonale, biventricular heart failure, pericardial diseases, cardiac tamponade and constrictive pericarditis.[1] Liver biopsies of patients with cardiac cirrhosis reveal congestive hepatopathy, appearing as a nutmeg liver with reddish central areas which represent sinusoidal congestion and bleeding in atrophied regions contrasting with yellowish discoloration of the normal liver tissue.[1] Hepatocytes in zone 3 of the hepatic lobule are most commonly where sinusoids appear to be enlarged and with variable degrees of hemorrhage.[2] This results in collagen deposition which over time can develop into fibrosis. Fibrosis surrounding centrilobular veins and in the perisinusoidal space is referred to as "cardiac sclerosis."[2] These histologic findings are useful in differentiating cardiac cirrhosis from other forms of ischemic injury such as Budd-Chiari syndrome.

Patients with cardiac cirrhosis typically present with signs of heart failure rather than that of liver cirrhosis. Common symptoms include: dyspnea, paroxysmal nocturnal dyspnea and orthopnea. Jaundice and splenomegaly are uncommon in patients with cardiac cirrhosis and total bilirubin levels rarely exceed 3mg/dl.[1] However, when assessing a patient's prognosis, total serum bilirubin was the strongest predictor for all-cause mortality.[3] Hypoalbuminemia is a common laboratory finding, however the mechanism appears to be related to bowel edema causing protein losing enteropathy and malnutrition as opposed to loss of synthetic function. Transaminase levels tend to only be mildly elevated: 2-3x upper limit of normal. This is in stark contrast to acute ischemic liver injury (shock liver) where transaminase levels are 10-30x the upper limit of normal. Imaging can be useful in diagnosis. Simple ultrasound of the inferior vena cava showing a diameter >2.3cm can be diagnostic. When calculating MELD score on for patients on anticoagulants due to heart failure, the MELD-XI (excluding INR) has been validated as a useful predictive model in place of the traditional MELD formula.[4]

Patients with cardiac cirrhosis are managed with diuretics to decrease the preload their heart must work against. Beta blockers which are often used to decrease preload must be used with caution in patients with cardiac cirrhosis as beta blockers are hepatically metabolized and may decrease cardiac contractility. For patients for whom medical therapy is inadequate, left ventricular assist device (LVAD) or dual transplant of heart and liver may be necessary.[5] If liver cirrhosis is considered irreversible, heart transplant alone is considered a contraindication, requiring a patient to receive a dual transplant.

Conclusion

Cardiac cirrhosis is an uncommon cause of cirrhosis in most community GI practices, making understanding of the pathophysiology essential to properly manage patients. The patient presented is a classic example of TTR amyloidosis (cardiac amyloidosis). However, his amyloidosis did not involve the

liver, making the management of his cirrhosis standard of care for those with cardiac cirrhosis: diuretics and dual heart- liver transplant. This patient failed to adequately diurese and was referred to the transplant service for additional therapeutic interventions.

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