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CASE REPORT | LIVER

Acute Liver Failure Due to Budd-Chiari Syndrome in the Setting of Cardiac Synovial Sarcoma

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Abstract

Primary malignant tumors of the heart, specifically cardiac sarcomas, are rare and mainly diagnosed at autopsy. Acute Budd-Chiari syndrome is a recognized cause of acute liver failure and has been associated with several rare cardiac tumors: atrial myxoma, caval rhabdomyosarcoma, and primary cardiac adenocarcinoma. We present the first case of a fatal, highly differentiated cardiac synovial sarcoma that presented as acute liver failure from Budd-Chiari syndrome.

Introduction

Primary malignant tumors of the heart are rare, with an estimated frequency of 0.0017–0.33%.[1] While 95% of malignant cardiac tumors are sarcomas, only 300 cases have been reported to date.[1,2] These tumors typically have an unfavorable prognosis, with 1-year mortality reported to be as high as 50%, as there is often a delay in diagnosis until the patient is symptomatic from systemic embolization, myocardial infarction, valvular heart disease, or new-onset heart failure.[1,3,4] There is no consensus on therapeutic modalities for these tumors.[4]

Budd-Chiari syndrome (BCS) is a disorder in which the major hepatic veins or inferior vena cava (IVC) are occluded secondary to clot burden resulting in acute liver failure (ALF). BCS typically occurs in the setting of thrombophilia, myeloproliferative disorders, congenital webs, and malignancy.[5] Acute BCS has been described in the setting of several rare cardiac tumors, including atrial myxoma, caval rhabdomyosarcoma, and primary cardiac adenocarcinoma, but not synovial sarcoma.[5]

Case Report

A 60-year-old white man with hypertension and Parkinson's disease presented with acute liver failure, lactic acidosis, acute renal failure, altered mental status, and acute-on-chronic lower extremity edema for 3 months. Initial examination revealed a somnolent, disoriented man with symmetric lower extremity edema, hepatomegaly, and ascites, but no other stigmata of chronic liver disease. Admission laboratory tests were remarkable for AST 3877 unit/L, ALT 2939 unit/L, total bilirubin 4.5 mg/dL, indirect bilirubin 2.9 mg/dL, INR 3.8, white blood cell 19.4 k/μL, platelets 63 k/μL, lactic acid 13.9 mmol/L, BUN 67 mg/dL, and creatinine 3.3 mg/dL, with a negative acetaminophen level and toxicology screen. A comprehensive hepatitis viral serology panel, Epstein-Barr virus, and Cytomegalovirus DNA-PCR, and HIV testing were unrevealing. Diagnostic paracentesis was negative for spontaneous bacterial peritonitis or malignancy. All other microbiological work-up revealed no evidence of infection. Testing for alpha-1 antitrypsin deficiency, hemochromatosis, and Wilson's disease were negative.

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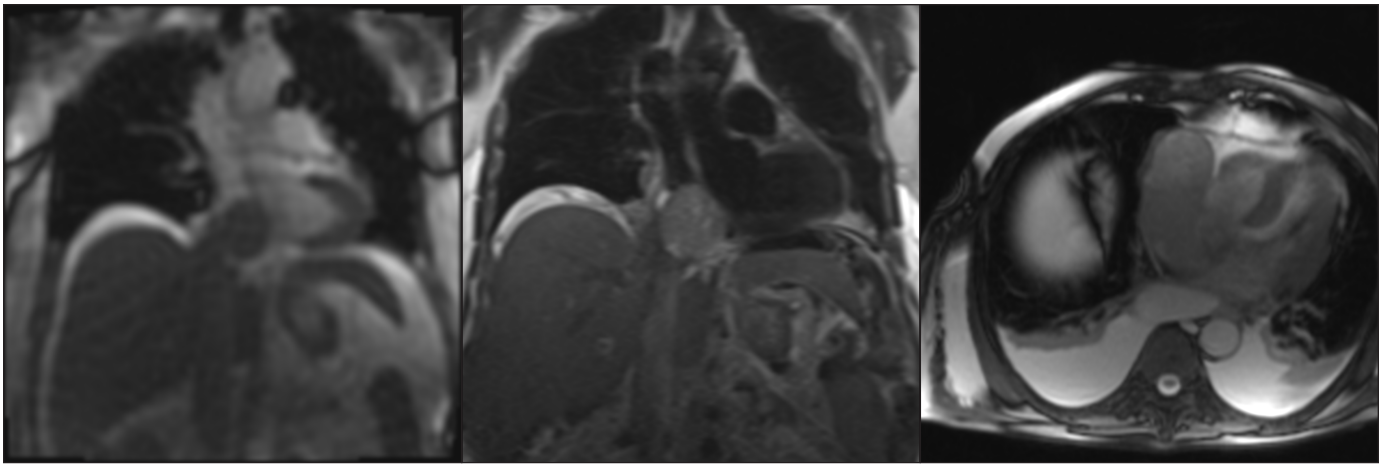

Figure 1. T2 weighted MRI image demonstrating an echodense right atrial mass concerning for malignancy.

The patient developed hepatorenal syndrome requiring continuous venovenous hemofiltration. Bilateral lower extremity Doppler ultrasonography showed extensive thrombosis of the femoral-popliteal system bilaterally with extension into both iliac veins. IVC filter placement was attempted, but aborted due to extensive clot burden. Liver ultrasonography with Doppler flow revealed hepatic vein thrombosis and reversal of portal venous flow. Cranial computed tomography (CT) was unremarkable. Abdominal, thoracic, and pelvic CT revealed further extension of thrombus into the right atrium. Transthoracic echocardiography showed normal left ventricular ejection fraction with a large echodense mass in the right atrium. Cardiac MRI demonstrated a 10.6 x 5.0 x 5.6-cm heterogeneous mass within the pericardium invading the right atrium, which was biopsied by cardiothoracic surgery (Figure 1). The patient continued to decline clinically and was transitioned to comfort care. Postmortum revealed a large lateral right atrial highly differentiated synovial sarcoma metastatic to the parietal and visceral pericardium. Extensive clots extending from the hepatic veins to the right atrium confirmed the diagnosis of BCS.

Discussion

While BCS has been described in association with atrial myxoma, IVC rhabdomyosarcoma, and primary cardiac adenocarcinoma,[5] this is the first report of association with cardiac synovial sarcoma. The clinical presentation of ALF should always prompt a search for BCS, and once diagnosed, a thorough work-up for thrombophilia, myeloproliferative disorders, congenital webs, and, in our case, malignancy, should be completed. Recent expert opinion by Mancuso suggested that in the presence of portal hypertension, transjugular intrahepatic portosystemic shunt (TIPS) should be performed as first-line therapy; angioplasty with stenting should be considered if TIPS is unsuccessful.[6] Liver transplantation should be used only as a rescue therapy. Due to the advanced presentation of liver disease and the clot burden, TIPS was not attempted in our patient, nor was transplantation an option with active malignancy. In retrospect, earlier investigation into either cardiac or hepatic etiologies for new-onset symmetric lower extremity edema 3 months prior to the patient's presentation to our hospital may have provided earlier treatment options with TIPS, stenting, or anticoagulation. A high index of suspicion for BCS is necessary for any patient demonstrating new onset features of portal hypertension, and we would recommend a right upper quadrant ultrasound with hepatic venous Doppler to evaluate for thrombosis. In the setting of large clot burden, malignancy should always be considered; we argue that, given the aggressive nature of cardiac tumors, synovial sarcomas should be considered in the differential diagnosis of BCS in the absence of an inherited thrombophilic state.

Disclosures

Author contributions: All authors contributed equally to manuscript creation. JG Stine is the article guarantor.

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