**Budd-Chiari syndrome as an initial presentation of hepatocellular carcinoma: a case report**

Lavinia Alice Bălăceanu¹, Camelia Cristina Diaconu², Gheorghiţa Aron¹

¹Internal Medicine Department, “Sf. Ioan” Clinical Emergency Hospital, ²Internal Medicine Department, Clinical Emergency Hospital Bucharest, “Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania

**Abstract**

We report the case of a 84-year-old admitted with symptoms of congestive heart failure. Ultrasonography revealed a hyperechoic nodule in the left lobe of the liver, with a peripheral hypoechoic rim, multiple irregular hypoechoic nodules in both hepatic lobes, portal vein, inferior vena cava, and right atrium thrombosis. On ultrasonographic and alpha-fetoprotein criteria the case was interpreted as hepatocellular carcinoma with Budd-Chiari syndrome. The particularity of the case is the initial presentation of the hepatocellular carcinoma as Budd-Chiari syndrome. The inferior vena cava and right atrium thrombosis, as a cause of secondary Budd-Chiari syndrome in a patient with hepatocellular carcinoma, has been rarely reported.

**Keywords:** hepatocellular carcinoma, Budd-Chiari syndrome, ultrasonography.

**Introduction**

Budd-Chiari syndrome is a very rare disease. Its incidence is not well reported in the literature. It is even more uncommon that Budd-Chiari syndrome appears as a complication of hepatocellular carcinoma.

**Case report**

A 84-year-old man presented to the Emergency Department for abdominal pain, leg swelling, shortness of breath, weight loss. He had a long history of chronic hepatitis B (HBV) and arterial hypertension, permanent atrial fibrillation, stroke, and ischemic heart disease.

The physical examination on admission revealed a blood pressure of 110/90 mmHg, irregular heart rate of 88 bpm, dyspnea at rest, decreased vesicular murmur in the right pulmonary base, firm hepatomegaly, bilateral peripheral edema, cachexia, and impaired clinical status.

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Blood tests showed normal hemogram values, cholestasis syndrome (total bilirubin 2.4 mg/dL, direct bilirubin 1.6 mg/dL, alcaline phosphatase 293 IU/L, gamma-glutamyl-transferase 360 IU/L), hepatic cytolysis, positive inflammatory tests (erythrocyte sedimentation rate 70 mm/h), hypocholesterolemia (131 mg/dL), mild alterations of liver function values (total protein 7.6 g/dL, albumin 3.5 g/dL, hypergammaglobulinemia 34%, INR 1.93, prothrombin activity 40%), positive D-dimers (400 ng/mL), elevated alpha-fetoprotein (957 ng/mL), and normal CA 19-9 and CA 125. ECG showed atrial fibrillation, 143 bpm. Transthoracic echocardiography revealed mild dilatation of the left atrium, decreased left ventricular systolic function with ejection fraction of 35%, and a 22x19 mm thrombus in the right atrium (fig 1). Abdominal ultrasonography revealed a 58 x 45 mm hyperechoic nodule in the left lobe of the liver, with a peripheral hypoechoic rim (fig 2), other multiple irregular hypoechoic nodules in the both hepatic lobes, normal spleen, right pleural effusion, mild ascites, portal vein thrombosis (fig 3) and inferior vena cava thrombosis (fig 4). The chest X-ray showed right pleural effusion and single pulmonary nodular opacity in the right inferior lobe. The patient was discharged at the family
Fig 1. Transthoracic echocardiography, 4-chambers apical view, showed a 22x19 mm hyperechoic thrombus in the right atrium.

Fig 2. Abdominal sonogram evidenced a 6 cm hyperechoic inhomogeneous node, with a less defined capsule and a peripheral hypoechoic rim, in the left lobe of the liver, segment II.

Fig 3. Longitudinal sonogram through the main portal vein demonstrated echogenic thrombus filling the lumen of vein.

Fig 4. Longitudinal sonogram showing inferior vena cava thrombosis.

Discussion

The Budd-Chiari syndrome implies thrombosis of the hepatic veins and/or the intrahepatic or suprahepatic inferior vena cava [1]. Malignant neoplasms are reported in the literature as being responsible for about 10% of Budd-Chiari syndromes, by direct compression or invasion of vascular structures [1]. Major hepatic and portal venous invasion occurs in approximately 10% of patients with hepatocellular carcinomas [2]. Caval and atrial extension of the tumor can rapidly trigger Budd-Chiari syndrome [2]. The thrombus can be tumoral or a blood clot in the context of coagulopathy. The imaging diagnosis of Budd-Chiari syndrome is based on three categories of signs: hepatic vein involvement, intrahepatic collateral circulation, and other coexistent conditions such as inhomogeneous parenchymal structure or benign regenerative nodules, caudate lobe hypertrophy, ascites, portal thrombosis, recanalized umbilical vein [3]. The combination of ultrasound signs of “altered hepatic and/or caval veins” and “caudate lobe hypertrophy” is the best strategy for diagnosing Budd-Chiari syndrome [4]. The acute form of Budd-Chiari syndrome with thrombus filling the lumen is uncommon [3,4] and the clinical presentation had the classical triad of ascites, abdominal pain, and hepatomegaly [3-5]. Also, it has been described in the literature the step-by-step occlusion of one vein followed by two or more veins, with development of venous collaterals, without significant symptoms [3]. Inferior vena cava and right atrium thrombosis, with secondary Budd-Chiari syndrome, has been rarely reported as the presenting feature of HCC [6,7]. Patients with Budd-Chiari syndrome secondary to hepatocellular carcinoma have a poor prognosis [5].
The particularity of our case is the initial presentation of hepatocellular carcinoma as Budd-Chiari syndrome. Right atrium, inferior vena cava, and portal vein thrombosis appeared in a patient with a long history of asymptomatic chronic HBV infection, despite the hypocoagulable state, with spontaneously elevated INR. According to the American Association for the Study of Liver Diseases (AASLD) guidelines (updated in 2010), a mass found incidentally or at screening, in a patient with known hepatitis B or cirrhosis of other etiology, is likely to be HCC [8]. Although we did not have a histopathological exam for confirmation or other imaging evaluation (CT scan, MRI or CEUS), as recommended by AASLD and EASL (European Association for the Study of the Liver) [8], we interpreted the hepatic lesions as HCC. The diagnosis of HCC is sustained by the ultrasonographic appearance of hepatic lesions and markedly elevated alpha-protein in a patient with a long history of chronic hepatitis B. The largest inhomogeneous hyperechoic hepatic nodule, of 6 cm diameter, with positive Doppler color flow, and the other small hypoechoic nodules had the classical ultrasound appearance of HCC. Abdominal ultrasonography did not show tumoral portal vein invasion. The simultaneous presence of the thrombus in three different venous locations raised the probability of clot thrombus formation. From the tumor markers, serum alpha-fetoprotein greater than 500 ng/mL in a high-risk patient is diagnostic for HCC [8]. In our case, markedly elevated serum alpha-fetoprotein associated with the ultrasonographic aspect led to the diagnosis of HCC with Budd-Chiari syndrome. Negative results of other tumor markers (CA 19-9, CA 125) and the result of abdominal ultrasonography made unlikely the possibility of kidney, pancreatic or digestive tract cancer in our case. The patient was discharged at family request, without waiting for the CT scan evaluation.

Conclusions

Budd-Chiari syndrome is an uncommon complication of hepatocellular carcinoma. The inferior vena cava and right atrium thrombosis, as a cause of secondary Budd-Chiari syndrome in a patient with hepatocellular carcinoma, has been rarely reported.

References