# A remarkable presentation of a massive Budd-Chiari syndrome

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## Introduction

A 59-year-old female presented to the emergency department with malaise, significant weight loss, abdominal discomfort, dyspnoea and severely swollen peripheral extremities. She had a past medical history of sickle cell anaemia, a latent tuberculosis infection and a chronic hepatitis B that was treated with PEG-interferon alpha-2 in 2008. Since 2018, she was lost to follow-up.

The patient was critically ill, sarcopenic and lethargic. Clinical examination revealed icteric sclerae and a markedly distended and diffusely tender abdomen and peripheral oedemas. Her vital signs included a blood pressure of 105/64 mmHg, tachycardia of 130 bpm, hypothermia of 35.6°C and a SpO2 of 100% in ambient air.

Laboratory workup demonstrated a total bilirubin of 12.9 mg/dL, AST 500 U/L, ALT 218 U/L, ALP 178 U/L and GGT 126 U/L, a thrombocytopenia of 34 x10E3/µL, 16,01 x10E3/µL leukocytes, a CRP of 94.8 mg/L and a lactate of 10,12 mmol/L. The serum creatinine was 1.04 mg/dL, eGFR 54.1 ml/min. The INR was 2.77 and there was a severe hypoalbuminemia (22 g/L). D-dimers were >20 000 ng/ml. The patient presented also with spontaneous hypoglycaemia.

A multiphasic contrast-enhanced thoracic and abdominal computed tomography (CT) was performed (figure 1). Given the results of the CT, a 2D echocardiogram was urgently requested (figure 2).

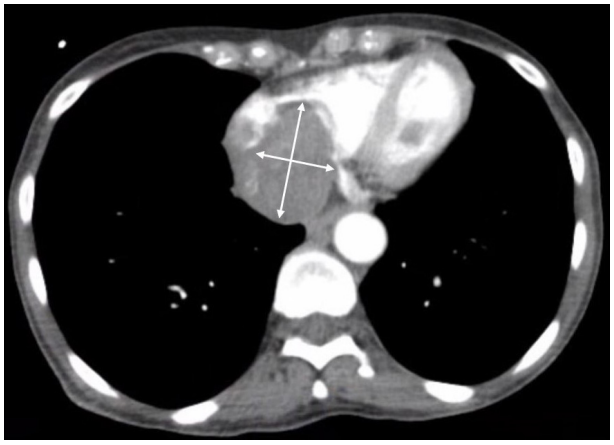

Figure 1a. — Cross-sectional Computed Tomography showed tumoral thrombus in the IVC extending into the right atrium (arrow).

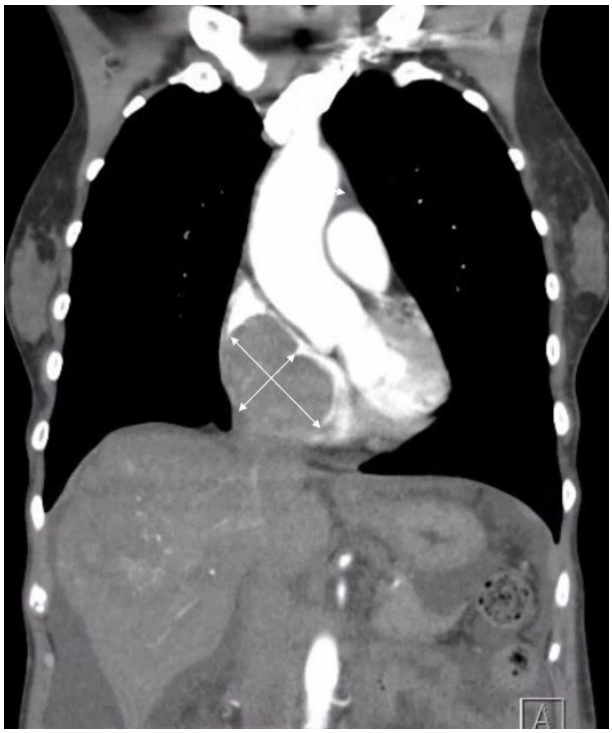

Figure 1b. — Coronal section of the Computed Tomography showed tumoral extension through the middle hepatic vein into the right atrium (arrow).

## Question

What is the most likely diagnosis based on clinical presentation, laboratory results and imaging?

## Answer

Additional lab testing showed an alpha-foetoprotein of 89 594 ng/ml and a HBV DNA of 5 400 000 IU/ml. The patient was diagnosed with decompensated cirrhosis with portal hypertension (MELD-Na 32 – Child-Turcotte-Pugh C13) due to a chronic uncontrolled hepatitis B with multifocal hepatocellular carcinoma (HCC) and massive tumoral thrombus, with Inferior Vena cava (IVC)

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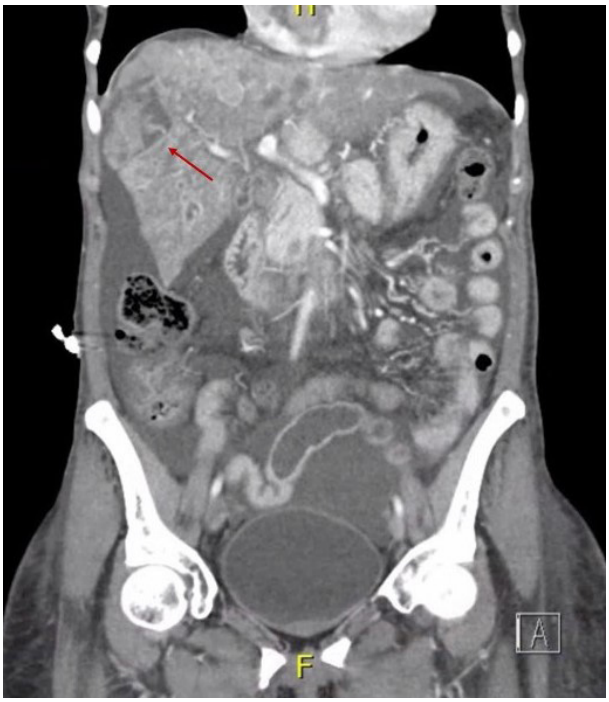


Figure 1c. — Coronal section of the Computed Tomography showed multifocal HCC (arrow).

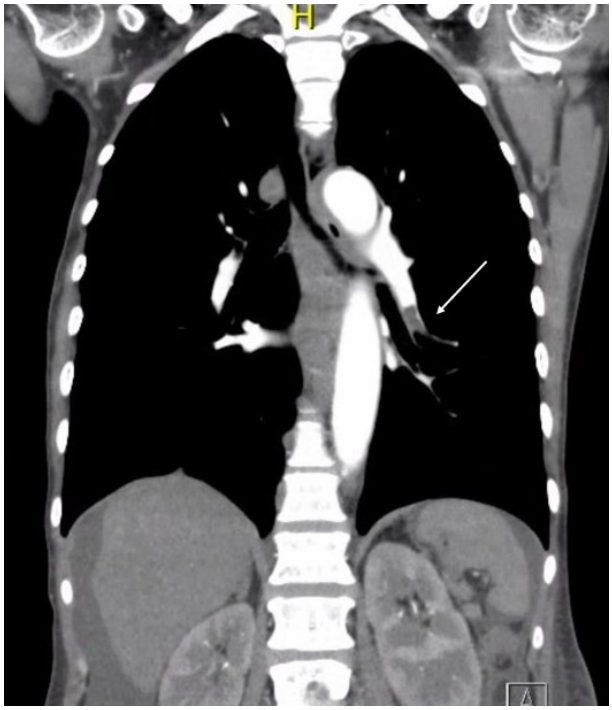


Figure 1d. — Coronal section of the Computed Tomography showed left-sided pulmonary embolism (arrow); occlusive segmental embolism in the antero- and medio basal segment of the left lower lobe.

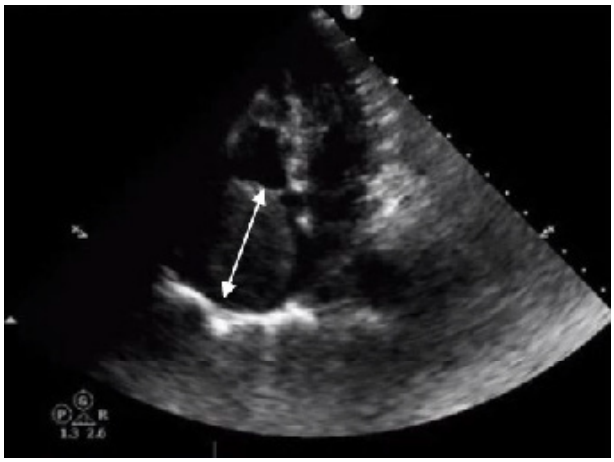

Figure 2. — 2D echocardiogram showed tumoral thrombus in the right atrium (arrow).

obstruction extending into the right atrium, resulting in a Budd-Chiari syndrome (BCS). She had also left-sided pulmonary embolism (1). This case is classified as terminal stage HCC, according to the BCLC staging system, for which best supportive care was offered (2).

*Teaching point/summary (extra)*

This clinical image illustrates a case of HCC developing on a background of chronic uncontrolled hepatitis B presenting with secondary BCS, with tumoral thrombus in the IVC extending into the right atrium and pulmonary embolism. The extent of this disease, portends an extremely poor prognosis.

**Keywords:** HCC, Secondary Budd-Chiari Syndrome, Hepatitis B.

**Conf ict of interest**

*None*

## References

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