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Case Report

Budd-Chiari syndrome mimicking autoimmune hepatitis ☆,☆☆

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ABSTRACT

Budd-Chiari syndrome is a rare disorder characterized by obstruction of hepatic venous outflow involving either the hepatic veins or the inferior vena cava. Risk factors include thrombophilia and myeloproliferative disorders, and symptoms are often unspecific. We present a case of a 60-year-old woman admitted to our medical unit for ascites and abdominal pain. Although her medical history significant for a mixed connective tissue disease and a mild increase of the transaminases had raised the suspicion of autoimmune hepatitis, the demonstration on computed tomography of the absence of outflow in the terminal tract of the supra-hepatic veins allowed a correct diagnosis. The contribution of radiological imaging is fundamental in the diagnosis of this rare and elusive pathology.

Introduction

Budd-Chiari syndrome (BCS) is a rare disorder characterized by obstruction of hepatic venous outflow involving either the hepatic veins or the inferior vena cava [1]. While its prognosis is good if an early therapy is started, the mortality is up to 90% in the cases left untreated [2,3]. Unfortunately the diagnosis is often not easy due to the nonspecificity of the symptoms [2]. We present a case of BCS mimicking an autoimmune hepatitis, in which a computed tomography (CT) allowed a correct diagnosis.

Case presentation

A 60-year-old woman was referred to our unit for the suspicion of autoimmune hepatitis. Her medical history was significant for a mixed connective tissue disease, diagnosed 10 years earlier, and for retinal vein thrombosis occurred 1 year earlier. In that occasion, a G20210A pro-thrombin gene mutation was identified. Two months before the admission, she had started to complain of abdominal tenderness and asthenia. Laboratory exams had showed an increase of alanine amino-transferase (ALT) and aspartate amino-

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transferase (AST) (about 3 times the upper limit of normal values) and a positivity of antinuclear antibodies (ANA). An abdominal ultrasonography had revealed a diffuse peritoneal ascites. At the admission, the physical examination of the patient revealed distended abdomen and edema of the lower extremities. Her reported medications were telmisartan, nebivolol, and low-dose aspirin. Laboratory exams showed mild increase of the transaminases (AST 103 U/L, ALT 96 U/L, normal values for both 5-50) and hypo-albuminemia (2.8 g/dL, normal values 3.5-5). Other routine tests were unremarkable. ANA were positive (1/160) whereas extractable nuclear antigens were not. Serology for hepatitis B and C was negative, as well as negative were antiliver and kidney microsome (anti-LKM), antisoluble liver antigen (anti-SLA), antismooth muscle antibodies, and antimitochondria antibodies.

A contrast-enhanced CT of the abdomen was ordered. Besides diffuse ascites, the exam showed mild hepatomegaly with inhomogeneous liver as for ischemic areas. The study of the hepatic vascular structures revealed narrow supra-hepatic veins without outflow in the tract near to the vena cava (Figs. 1-3). A thrombus was suspected in the vena cava at the confluence with the supra-hepatic veins, but not confirmed by a subsequent magnetic resonance imaging (MRI).

Based on the presence of pro-thrombotic factors, compatible symptoms and appropriate radiological findings [2,4], a diagnosis of BCS was made, and anticoagulation was started (full-dose low molecular weight heparin in the in-hospital phase and then apixaban 5 mg twice daily). Diuretics were also prescribed, and discontinued after three weeks. Serology for antiphospholipid syndrome and search for Janus Kinase 2 gene were negative.

Three months later, a color-Doppler ultrasonography of the abdomen documented normal flow in the supra-hepatic veins and complete disappearance of the ascites. At 6 months of follow-up, the patient is going well and continues to take anticoagulants.

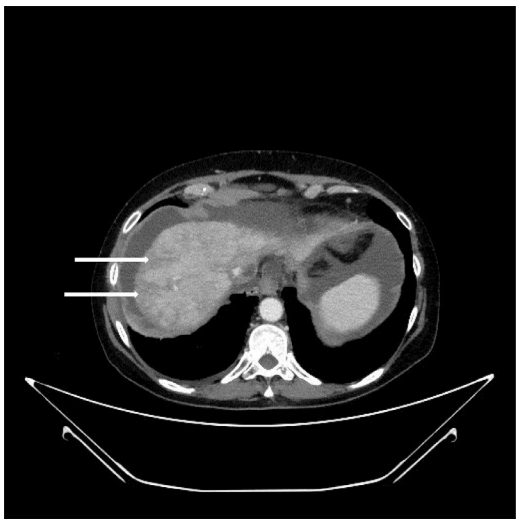

Fig. 1 – Axial contrast-enhanced CT image showing diffuse ascites and inhomogeneous liver for hypo-perfusion areas suggestive for ischemia (white arrows).

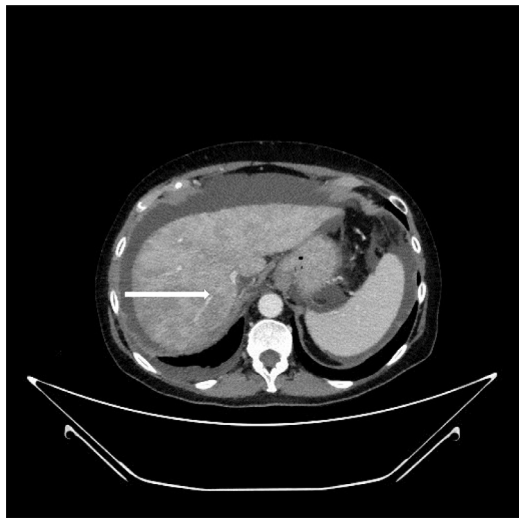

Fig. 2 – Axial contrast-enhanced CT. Narrow right supra-hepatic vein without outflow in the terminal segment (white arrow). Intermediate and left supra-hepatic veins are not visible.

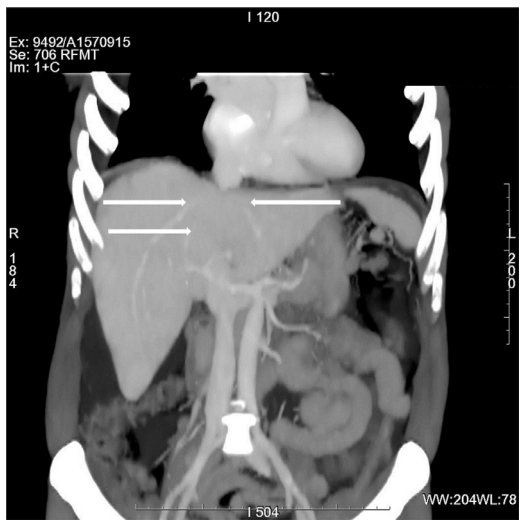

Fig. 3 – Coronal maximum intensity projections contrast-enhanced CT. Narrow supra-hepatic veins without outflow in the tract near the vena cava (white arrows).

Discussion

BCS is rare, with an incidence of about 0.4-0.8 per million person-years [5]. For several years, its etiopathogenesis has been unclear, but now it is widely accepted that BCS, at least in its primary form, more frequent in the western world [2], is basically a deep vein thrombosis of an unusual site [2,4]. The secondary BCS is instead caused by compression or infiltration of the supra-hepatic veins due to several causes such as neoplasms, abscesses, parasitosis, or trauma [2]. The primary BCS shares several risk factors with other vein thromboses. In a prospective observational study, almost 90% of patients had

at least one thrombophilic disorder (Factor V Leyden or prothrombin gene mutations, antiphospholipid syndrome, others), about 50% two or more, about half had a mieloproliferative disorder (Janus Kinase 2 gene positive or negative), and almost 25% suffered from an auto-immune disease [4].

The clinical presentation ranges from asymptomatic cases to acute liver failure [2,6]. The more frequent symptoms are ascites and abdominal pain [4,6]. In an appropriate anamnestic and clinical context, the diagnosis relies basically on imaging procedures. Doppler-ultrasonography is usually able to identify the absence of outflow in the supra-hepatic veins (sensitivity and specificity about 85%) and is the first-line imaging study for evaluation of patients with suspected BCS. CT or MRI is equally useful without the limit of operator dependence. The most common sign of BCS on CT is nonvisualized supra-hepatic veins [2,4,7,8].

In our case, the previous diagnosis of an autoimmune disease, the female sex of the patient, and the increase of the transaminases had raised the suspicion of autoimmune hepatitis. However, the CT performed was able to demonstrate the absence of outflow in the terminal tract of the supra-hepatic veins and this, together with the anamnestic and clinical findings, allowed a correct diagnosis.

The cornerstone of the treatment for primary BCS is full-dose anticoagulation, which should be started as soon as possible and continued indefinitely if no contraindications exist [2,4,6]. In inveterate cases, or in case of failure of anticoagulant therapy, transjugular intrahepatic portosystemic shunting, or less frequently, percutaneous transluminal angioplasty, are used [9–12]. Orthotopic liver transplantation should be considered in the most serious cases, when other options have failed [9–12].

The prognosis of primary BCS has changed in the recent years. While before the use of the anticoagulation the vast majority of patients died from BCS [1,2], nowadays the overall survival approaches 90% at 2 years [4,11]. The prognosis of the secondary BCS depends instead on the underlying cause. Our patient is going well at the 6-month follow-up.

Ethics approval and informed consent

This case report has obtained approval from the head of the Internal Medicine unit at the Department of Internal Medicine of the Azienda Ospedaliera di Cosenza (Italy).

Author contributions

Bova C, De Bartolo T, Pellegrini R, De Vuono A, Fiaschi E: Conception and design of study. Bova C: drafting the manuscript. All authors read and approved the final manuscript.

Patient consent

A written informed consent for the publication of this case has been obtained from the patient.

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