Spiral Computed Tomography and Magnetic Resonance Angiography Evaluation in Budd Chiari Syndrome

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Abstract

Budd-Chiari syndrome is caused by the obstruction of the hepatic venous outflow at the level of the hepatic venules, large hepatic veins, and inferior vena cava up to the confluence with the right atrium. When it is untreated, the mortality rate for patients is high. Because the clinical presentation of this syndrome is nonspecific, imaging investigation - computed tomography and magnetic resonance - are important diagnostic steps. Contrast-enhanced multiphase spiral computed tomography (CT) and magnetic resonance (MR) angiography permits morphologic and functional assessment of parenchymatous liver changes in this particular entity.

In this review, we present the spectrum of vascular and hepatic parenchymal abnormalities in Budd-Chiari syndrome observed on multiphase contrast enhanced spiral CT and MR angiography.

Keywords

Budd Chiari syndrome – spiral computed tomography – magnetic resonance angiography

Introduction

Budd-Chiari syndrome (BCS) is a heterogeneous group of disorders induced by thrombotic or non-thrombotic obstruction of hepatic venous outflow (congenital-web, diaphragm, interruption of the inferior vena cava (IVC); injury and/or inflammation, liver tumor) located at the level of the hepatic venules, large hepatic veins, and IVC up to the confluence with the right atrium [1]. Idiopathic BCS is present in one third of cases.

The syndrome has been classified according to the site of the obstruction, as follows: type I: occlusion of the IVC with or without secondary hepatic vein occlusion; type II: occlusion of major hepatic veins; type III: obstruction of the small centrilobular venules (considered as veno-occlusive disease) [1, 2] .

Men and women are equally affected, but an acute presentation is more common in females. Most patients present in the third and fourth decade. Morbidity and mortality are largely dependent on the underlying cause.

Usually, the clinical presentation of this syndrome is nonspecific. It can present in a fulminant, acute, subacute or chronic form [1-3].The presentation is dependent on the onset and extent of venous occlusion [3-5]. Hepatic venous thrombosis leads to an elevation of sinusoidal pressure. Histologically, the increase in pressure leads to centrilobular congestion followed by necrosis and atrophy [6]. In the acute form, venous collaterals have not developed and hepatocellular necrosis rapidly occurs. In most patients with the subacute and chronic form of BCS a variety of accessory hepatic veins drain into the IVC above or below the site of obstruction. The collateral venous drainage helps to relieve hepatocellular congestion but does not prevent peripheral atrophy [6-8]. Obstruction of the hepatic venules induces shunting of the flow from the hepatic arteries into the portal veins. Due to the accessory venous drainage and preservation of the portal venous supply, the caudate lobe and central liver is often spared and present compensatory hypertrophy [9-11].

Sectional imaging evaluation is an important diagnostic step also in the assessing of BCS complications [8, 10-12].

Imaging technique

Patients with clinical and ultrasonographic suspicion of BCS underwent our routine spiral CT protocol for the examination of liver tumors. CT sections were collimated to 5 mm at a pitch of 1. Images were obtained during an arterial phase (25-30 sec) and portal-phase (50- 60 seconds) after initiation of i.v. administration of a 100 ml bolus of nonionic contrast material at a rate of 3 ml/sec. MR imaging

J Gastrointestin Liver Dis
June 2008 Vol.17 No 2, 223-226
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was performed with a 1.5-T superconducting system using: Fast spin-echo T2- and FSPGR breathold T1-weighted MR images. In all patients we used a bolus (0.1 mmol/kg) of paramagnetic contrast media before acquisition of a multiphase 3D gradient-recalled echo MR angiography, in arterial, portal and parenchymal phase.

Imaging findings

CT abnormalities

In the *acute BCS*, CT signs reveals hepatomegaly, ascites and lack of opacification of the hepatic veins (Fig.1). The liver appears inhomogeneous with a mottled appearance on contrast multiphase imaging (Fig.2). There is delayed enhancement in the periphery of the liver and around the hepatic veins [3, 4]. Peripheral atrophy of the liver with

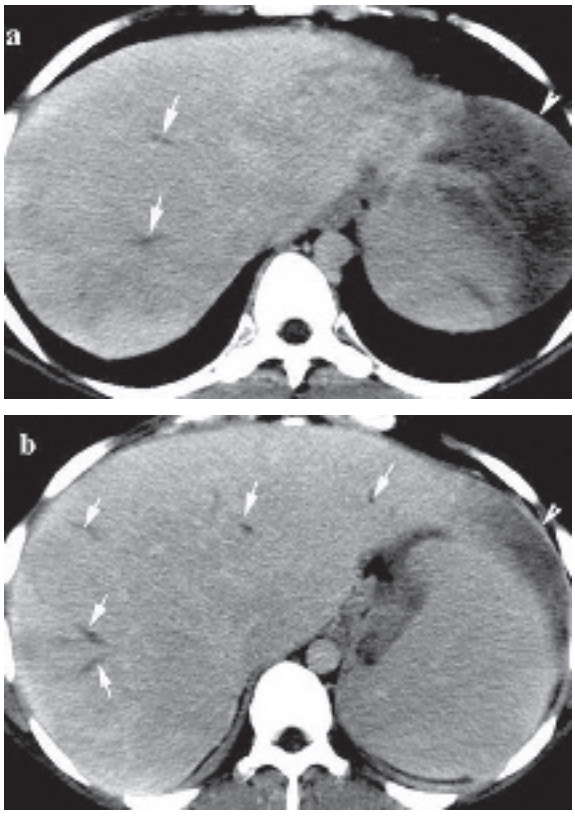

Fig 1. Budd-Chiari syndrome in the context of antiphospholipid syndrome. Enhanced spiral CT: thrombosis of hepatic veins (arrows) and left liver ischemia (arrow head).

enlargement of the caudate lobe is evidenced (Fig.2). The peripheral zones of the liver usually appear hypoattenuating because of the reversed portal venous blood flow, which results from the increased postsinusoidal pressure produced by hepatic venous obstruction. The caudate lobe is enlarged and demonstrates increased contrast enhancement compared with the remainder of the liver.

In the *chronic BCS,* CT signs are: atrophic liver with exception of the caudate lobe, collateral vessels within the liver and related to portal hypertension, large, multifocal regenerative nodules with enhancement due to their

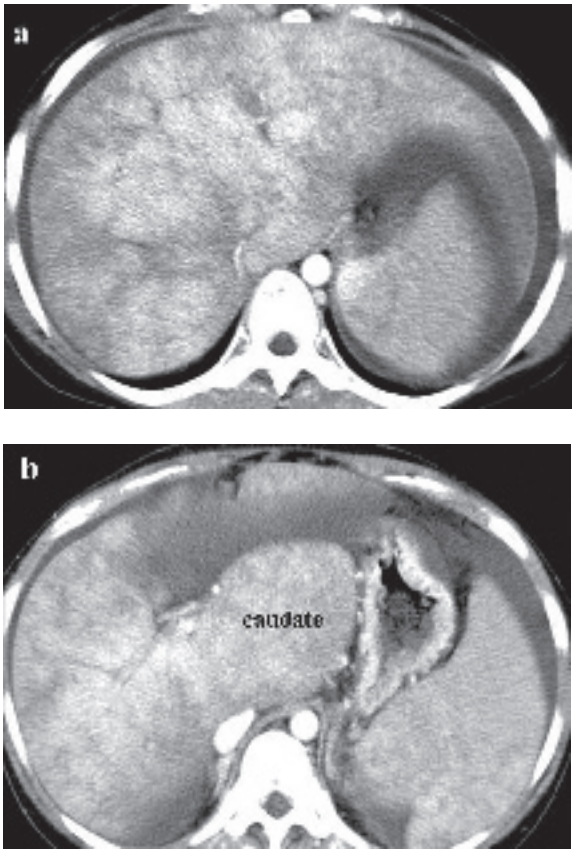

Fig 2. Budd-Chiari syndrome in the context of thrombocytosis. Enhanced spiral CT: hypoattenuating of the peripheral liver parenchyma, inhomogeneous enhancement of liver parenchyma with a mottled appearance (a); hypertrophy of the caudate lobe (b), ascites.

hypervascularity - best seen on arterial and portal phase imaging [13]. The porta hepatis may be displaced anteriorly; portal vein thrombosis found in 9–20%; thrombosis within the hepatic veins and IVC in 18–53% [8].

MRI abnormalities

Regional differences in signal intensity because of varying perfusion, atrophy, hypertrophy, necrosis, and differences in the amount of intracellular fat or iron may be found. Other findings are hepatosplenomegaly, an enlarged caudate lobe, a cirrhotic liver, and ascites [13, 14]. The liver parenchyma appears usually inhomogeneous. MRI in acute form: the liver presents in the periphery areas of low SI on T1weighted images and high SI on T2 weighted images, lack of enhancement of the hepatic veins (Fig.3); reduced enhancement of peripheral liver on delayed phase [15, 16]. IVC abnormalities shown on MRI include diffuse narrowing (Fig.4) and focal thrombosis. In subacute form, the liver periphery presents low signal on T1, high signal on T2 and heterogeneous enhancement. In chronic form, large regenerative nodules present increased signal intensity on T1 and low-to-intermediate SI on T2 [10, 14]; no significant difference in the signal between the central and peripheral liver on T1 and T2 images; accessory hepatic veins drain into the IVC above or below the site of obstruction (Fig.5).

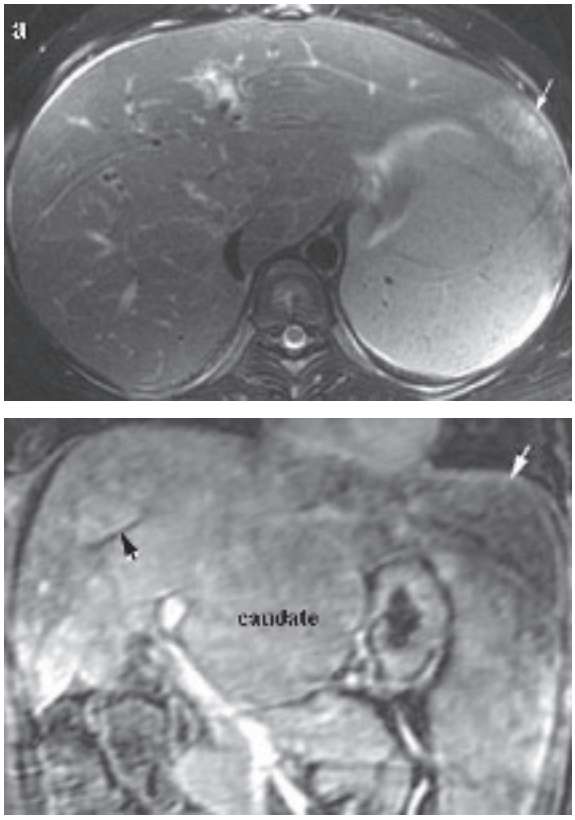

Fig 3. Budd-Chiari syndrome in polycytemia vera. MRI evaluation - T2 wi (a), T1 postGd, venous phase: dysmorphic hepatomegaly with caudate enlargement and presence of ascites, inhomogeneous mottled enhancement on the central part of the liver, hepatic venous thrombosis (black arrow), left liver ischemia (white arrow).

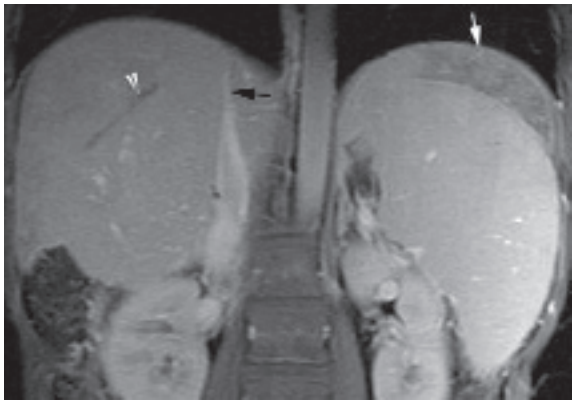

Fig 4. Acute Budd-Chiari syndrome. MRI T1 wi postGd evaluation in parenchymal phase, coronal plane: narrowing of the IVC (black arrow); right hepatic vein thrombosis (white arrow), left liver ischemia (arrow head).

Common CT and MRI abnormalities in Budd-Chiari syndrome

Changes in liver parenchyma. In BCS the most common hepatic change is hypertrophy of the caudate lobe. It is found in 82–91% of all cases [10,15,16] and is related to independent drainage of this lobe. In the acute stage, the liver may be globally enlarged due to vascular congestion [16-18]. In the chronic stage, atrophy of the right lobe of the liver, hypertrophy of the left lobe, irregularities of liver contours, and presence of regenerative nodules are prominent features [14, 16, 18]. Nodularities of the hepatic surface may show the progression to cirrhosis. Regional enhancement differences reflect the hemodynamic disturbance in the liver in patients with BCS. Nonenhancement is an indicator of hypoperfusion. A patchy pattern of hepatic enhancement is thought to be produced by regional stagnation of portal flow [13]. Normal or increased enhancement of the caudate lobe is usually seen in acute forms of BCS because the caudate lobe has separate venous drainage from the remainder of the liver [16]. At multiphasic spiral CT or MRI evaluation, large regenerative nodules are markedly and homogeneously hyperattenuating (hyperintensity) on arterial dominant phase images and remain slightly hyperattenuating (hyperintensity) on portal venous phase images [11, 14].

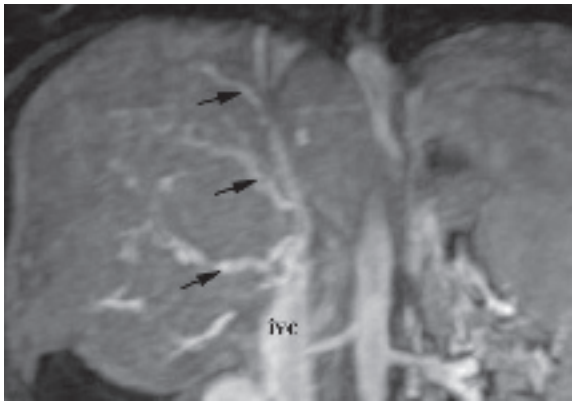

Fig 5. Chronic Budd-Chiari syndrome. MRI T1 wi postGd evaluation in parenchymal phase, coronal plane: accessory hepatic veins draining into the IVC.

Changes in the vascular system. The hepatic artery can be the major supplier of blood to the liver when the portal vein becomes the draining vein in BCS [13]. Secondly, hepatic artery anatomic variations must be shown in patients with BCS who are candidates for liver transplantation. Narrowing, stretching, or distortion of the hepatic artery on enhanced spiral CT and MR angiography may be the indirect signs of severe morphologic changes in liver parenchyma.

Portal vein system. The portal vein and its intrahepatic branches may be influenced by structural changes of the liver tissue. Because the hepatic veins constitute the efferent vascular drainage of the liver, obstruction or increased pressure in these vessels results in increased sinusoidal and portal pressure. In areas with complete hepatic vein obstruction, the resistance to portal flow is increased and may stop and even reverse the flow. It has been shown that after hepatic venous occlusion, the portal vein becomes the draining vein, and the occluded area is supplied with arterial blood alone [10]. In patients with BCS, stasis in the portal system may cause thrombosis [11]. The accurate evaluation of the portal system is particularly important when assessing the possible treatment options (shunt surgery or transjugular intrahepatic portosystemic shunt placement).

Hepatic vein system. Normal main hepatic veins are best visualized on subvolume targeted maximum intensity

projections obtained in the craniocaudal direction. The right hepatic vein can also be well depicted on coronal images because of its parallel orientation to the body axis [10, 13, 16]. However, the left and middle hepatic veins, which are shorter than the right hepatic vein, may be difficult to visualize, if they are compressed by the distorted liver parenchyma. They can be best identified near the caval confluence. Nonvisualization of hepatic veins may be suggestive of Budd-Chiari syndrome but also inadequate time delay after contrast material administration can be responsible for the nonvisualization of these veins.

Inferior vena cava. Contrast-enhanced multiphase CT and MR angiography can delineate the IVC in great detail along its course. Stenosis of the IVC may be associated with external compression of an enlarged caudate lobe. Contrast-enhanced three-dimensional (3D) MR angiogram replace the IVC cavograms.This method offers the advantage over conventional cavography to evaluate the nature of the obstruction (intrinsic or extrinsic in origin) at the same time as the surrounding soft tissue anatomy and also the cranio-caudal extension of the obstructive lesion [10, 11,13].

Collateral veins. The intrahepatic collateral vessels divert blood away from the occluded hepatic vein and drain into a patent hepatic vein or a systemic vein. The sites of extrahepatic collateral veins in BCS are generally different from those collaterals localized at the portosystemic communication sites in cirrhosis. Extrahepatic collateralized routes seen in BCS are the left renal–hemiazygos pathway, inferior phrenic–pericardiophrenic collaterals, and superficial collaterals of the abdominal wall [2, 11, 16, 18].

Conclusion

Cross sectional enhanced multiphase spiral CT and MR angiography allow a correct diagnosis of the Budd-Chiari syndrome and help to evaluate the liver status as well as its possible complications.

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