Diagnosis of Budd-Chiari syndrome (SBC) by pulsed Doppler ultrasound and computed tomography: a retrospective study

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Authors: M. Dridi\textsuperscript{1}, S. Jendoubi\textsuperscript{2}, L. Ben Farhat\textsuperscript{3}, I. Marzouk Moussa\textsuperscript{2}, A. Manamani\textsuperscript{2}, L. Hendaoui\textsuperscript{3}; \textsuperscript{1}Bab saadoun, Tu/TN, \textsuperscript{2}Tunis/TN, \textsuperscript{3}Sidi Daoued/TN

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Aims and objectives

Budd-Chiari syndrome (BCS) is the manifestation of hepatic venous outflow obstruction the exact frequency is unknown. The classic clinical presentation varies from fulminant form with severe portal hypertension, acute form with the clinical triad of: ascites, abdominal pain, hepatomegaly and chronic forms [1]

**Acute:** results from an acute thrombosis of the main hepatic veins or the IVC; patients may present with rapid onset ascites

**Chronic:** the chronic form is related to fibrosis of the intrahepatic veins, presumably related to inflammation [2]

The aim of our study was To determine the clinical characteristics of SBC and illustrate the performance of imaging in the positive and etiologic diagnosis.

Methods and materials

From July 2009 to March 2014, 25 symptomatic patients followed for SBC explored in the radiology department of university hospital Mongi Slim were retrospectively and consecutively included in this trial. There were 18 women, 7 men, with an average age of 32 years (range 6-62). Clinically, four patients showed evidence of advanced BCS, including abdominal ascites, coagulopathy and hepatomegaly.

All patients were investigated by abdominal pulsed Doppler ultrasound. Nineteen patients were explored by a multi-slice computed tomography (MSCT)

Two patients underwent MRI with axial cuts T1-weighted sequences, T2 and a dynamic T1 gadolinium.

Results

The etiology can be divided into primary and secondary cause. Primary BCS results from the direct occlusion of hepatic veins from an intraluminal source of the thrombus. Secondary BCS is caused by an extrinsic compression of either the IVC or the hepatic veins. The primary form of BCS is the most common, with an underlying hypercoagulable state present in most patients. This can include mutations in Factor V Leiden, Protein C and S deficiencies, and the presence of antithrombin[3]. A number of other primary causes of BCS have been identified and covered in a number of excellent articles.
Secondary BCS can be related to hepatic cystic disease or some other external compression of the local vasculature, most commonly malignant disease of various abdominal organs. Budd-Chiari syndrome can be classified according to etiology, site of obstruction, manifestations, and duration of the disease. In our study, Budd-Chiari syndrome was considered primary in 14 cases when obstruction of the hepatic venous outflow tract is the result of an endoluminal venous lesion (thrombosis)

A thrombogenic field was found in 8 these patients:

- One patient had polycythemia vera.
- Five patients had a protein C and S deficiency.
- Two patients had a hypercoagulable state in post partum.

For the other 6 cases no risk factor was found,

It was considered secondary in 11 cases when the obstruction results from the presence in the lumen of material not originating from the venous system or from extrinsic compression by a neighboring tumor.

In practice, BCS is regarded as primary when no causes of secondary obstruction are found. BCS can also be classified according to the level of obstruction.

**Sonographic Findings in BCS:**

In our study, Doppler ultrasound confirmed the diagnosis in 17 cases showing an hepatic vein thrombosis (n = 4), spindly veins (n = 12), dilated (n = 3), permeable (n = 9), ostial stenosis (n = 4), extrinsic compression (n = 6).

Spectral analysis showed a reverse flow (n = 20) was absent (n = 4), normally modulated (n= 2).

Doppler sonography of the liver, with sensitivity and specificity of 85% or greater, is the technique of choice for initial investigation when BCS is suspected.

There are different categories of signs, specific signs : hepatic vein involvement and suggestive signs such as intrahepatic collateral circulation (fig 1, fig 2) , a caudat vein (3mm or larger) ,benign regenerative nodules (fig3) , caudate lobe hypertrophia portal thrombosis , ascites (fig 4).

Sonographic features of hepatic vein involvement include a partial or complete inability to see the hepatic veins, stenosis with proximal dilatation, intraluminal echogenicity, thickened walls, and thrombosis . 9 except for the paraumbilical vein, which is also seen in portal hypertension are a specific diagnostic criterion for BCS. Collaterals are either
intrahepatic, which communicate with systemic vessels via subcapsular collaterals or from an occluded hepatic vein to a non occluded hepatic vein or to the caudate lobe. Rarely, collaterals are also seen from hepatic veins to the suprahepatic IVC close to the right atrium [5,6].

Abdominal CT showed an enlarged segment I (n = 12), a heterogeneous enhancement mosaic (fig5) (n = 12) Segment I was typically enhanced in all patients.

Images for this section:

**Fig. 1:** a-Thrombus in the middle (M) and right hepatic veins (D) seen as echogenic areas (arrows) in the transverse subcostal image. b-Collateral vessel connecting occluded (or stenotic) hepatic vein to neighboring patent hepatic vein

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Fig. 2: Images through the epigastric presence of fibrous cords replacing veins and showing an intrahepatic collateral between hepatic veins (arrow) seen as an abnormal vessel communicating between the 2 hepatic veins

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Fig. 3: Helical CT after IV contrast injection displays enhancing nodule (arrows) surrounded by low-attenuating area related to vascular disorders. On helical CT, benign regenerative nodules are homogeneously hyperattenuating on arterial phase and remain slightly hyperattenuating on portal vein phase.

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Fig. 4: 56-year-old woman with enlargement of abdominal diameter image show enlargement caudate lobe and increased contrast enhancement compared with the remainder of the liver

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Fig. 5: Image showing hydatid cyst compressing VCI and heterogeneous enhancement mosaic of the liver parenchyma with presence of hypodense bands of the fibrosis

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Conclusion

Budd-Chiari syndrome is an uncommon disorder the etiology is often misdiagnosed or underdiagnosed. Therefore, a successful diagnostic and therapeutic approach is of vital importance. Color Doppler sonography offers noninvasive hemodynamic and effective modality for diagnosis of BCS.

Personal information

References