ELSEVIER

European Journal of Radiology 61 (2007) 44–56

EJR
EUROPEAN JOURNAL OF RADIOLOGY

www.elsevier.com/locate/ejrad

Review

Budd-Chiari syndrome: A review of imaging findings

Ayşe Erden *

Ankara University, School of Medicine, Department of Radiology, Talatpaşa Bulvarı, Sıhhiye 06100, Ankara, Turkey

Received 16 October 2006; accepted 2 November 2006

Abstract

Budd-Chiari syndrome is an uncommon, often fatal disorder resulting from an obstructed hepatic venous outflow tract. The obstructive lesion is situated in the main hepatic veins, in the inferior vena cava or in both. The nature, location and extention of the obstruction can be displayed on diagnostic imaging techniques. In addition to this direct evidence, the indirect findings of venous obstruction such as the presence of intra- and extrahepatic collateral veins, when combined with the altered morphology and enhancement pattern of the liver enables one to arrive at a confident diagnosis. In patients with suspected Budd-Chiari syndrome, gray-scale sonography with complementary support of color and pulsed Doppler examinations is the first step in approaching the diagnosis. It is followed by a contrast-enhanced cross-sectional technique, preferrentially by MR angiography. The patients with a high clinical suspicion of Budd-Chiari syndrome may undergo hepatic venography or venacavography directly so that a potential of recanalization (e.g. percutaneous transluminal angioplasty with or without stent placement or TIPS) of the obstructed segment under the guidance of these techniques would not be delayed.

Keywords: Budd-Chiari syndrome; Hepatic veins; Diagnostic imaging

Contents

1. Introduction

Budd-Chiari syndrome is a rare entity whose diagnosis is usually suspected during radiologic investigations. Imaging findings of this syndrome have an important role in directing the clinician to appropriate management algorithm.

In this article, it is aimed to define an approach to the diagnosis of Budd-Chiari syndrome based on abnormal imaging findings.

2. Definition

According to the European Group for the Study of Hepatic Vascular Diseases, Budd-Chiari syndrome is defined as hepatic

* Correspondence address: Hafta Sokak, 23/6, Gaziosmanpaşa 06700, Ankara, Turkey. Tel.: +90 312 362 30 30/67 73; fax: +90 312 310 08 08.
E-mail address: ayse.erden@medicine.ankara.edu.tr.

0720-048X/$ – see front matter © 2006 Published by Elsevier Ireland Ltd.
doi:10.1016/j.ejrad.2006.11.004

venous outflow obstruction at any level from the small hepatic veins to the junction of the inferior vena cava and the right atrium, regardless of the cause of the obstruction. Cardiac etiologies of hepatic congestion or veno-occlusive disease (sinusoidal obstruction syndrome) are not included in this definition [1].

3. Classification

At present, no standardized classification is available for Budd-Chiari syndrome [1]. Several classifications have been proposed based on pathogenesis (primary or secondary), location of the venous occlusion (Types I–III), or clinical and morphologic features (fulminant, acute, subacute and chronic) [2–4]. Budd-Chiari syndrome is classified as *primary* when it is caused by an intrinsic luminal web or thrombus, and *secondary* when it is caused by an extraluminal compression or tumoral invasion [1–5].

4. Pathogenesis and pathologic findings

Membranous obstruction of suprahepatic inferior vena cava, which is also known as primary Budd-Chiari syndrome, is due to fibromuscular membrane (web) or an acquired lesion. Membrane or web arises from the wall of the vessel and may obliterate the lumen completely or partially [6]. This type of lesion is believed to be a sequela of long-standing thrombosis [7].

The etiology of the venous thrombosis is obviously an important consideration. Hematologic abnormalities such as myeloproliferative disorders, paroxymal nocturnal hemoglobinuria, the antiphospholipid syndrome, inherited deficiencies of protein C, protein S and antithrombin III, factor V Leiden mutation, prothrombin-gene mutation, methylenetetrahydrofolate reductase mutation are responsible for the majority of cases of Budd-Chiari syndrome [5,8]. The other factors that contribute to the development of Budd-Chiari syndrome include pregnancy immediate postpartum period and use of oral contraceptives. The secondary Budd-Chiari syndrome is caused by an extraluminal compression of a space occupying lesion or luminal invasion of malignant tumor (renal cell carcinoma, hepatocellular carcinoma, adrenal carcinoma, hepatic metastasis, primary leiomyosarcoma of inferior vena cava) [5,8]. In some instances, the cause is difficult to establish, and in others, multiple etiologic factors can play role in the development of the venous occlusion [5].

As the hepatic veins constitute the sole efferent vascular drainage of the liver, obstruction or increased pressure within these vessels or in their radicles result in an increased sinusoidal pressure. Regardless of the etiology, elevation of hepatic sinusoidal pressure leads delayed or reversed portal venous outflow. The portal venous stasis and congestion cause hypoxemic damage in adjacent hepatocytes. Afterwards centrilobular fibrosis, nodular regenerative hyperplasia, and ultimately cirrhosis occur. Main alterations in hepatic morphology include atrophy of peripheral regions and hypertrophy of the caudate lobe and central portions of the liver [8,9].

Liver biopsy is an important procedure to confirm the diagnosis and assesses the degree of hepatocellular damage. Histopathologic clues of venous outflow obstruction are centrizonal congestion, hemorrhage and cell necrosis, sinusoidal dilatation with or without central vein obliteration and congestive pattern of cirrhosis [2].

5. Clinical findings

The clinical presentation of Budd-Chiari syndrome depends on the extent and rapidity of the hepatic vein occlusion and on whether a venous collateral circulation has developed to decompress the hepatic sinusoids. Rapid onset of abdominal pain due to liver congestion and insidious onset of intractable ascites are major manifestations of acute form. The chronic form is manifested as complications of cirrhosis [2,8]. Budd-Chiari syndrome may be quite indolent or even asymptpomatic in some patients [5].

6. Radiologic procedures

The diagnostic imaging is the starting point in the investigation for Budd-Chiari syndrome. The structural changes detected on imaging methods, when integrated with pertinent clinical findings and results of laboratory tests, enables one to arrive at a confident diagnosis. However, it should be noted that the imaging findings are usually overlooked despite they are rather typical. It is not unusual for patients with chronic forms of Budd-Chiari syndrome to be diagnosed with liver cirrhosis. Real time gray-scale sonography combined with color flow and duplex Doppler examination is considered by some to be the initial imaging technique [1,8,10,11].

6.1. Ultrasonography and color Doppler sonography

The most important goal in patients who refers with the suspicion of having Budd-Chiari syndrome is assessment of the hepatic vein patency and size. The state of the inferior vena cava is also noteworthy. The obstructive process in the hepatic veins or in the inferior vena cava may have diverse appearances. The presence of echogenic material in the lumen indicates a thrombotic process. Color and pulsed Doppler techniques as the complementary of gray-scale sonography is very beneficial to confirm the diagnosis (Fig. 1). The sensitivity of pulsed Doppler examination in Budd-Chiari syndrome is 87.5% [12]. In a recent study, it is found that contrast-enhanced sonography is superior to gray-scale and color Doppler imaging for the detection and characterization of hepatic vein thrombosis [13].

Sonography has been used for the demonstration of webs since late 1980s [14]. Membrane or web is seen as an echogenic crescent-shaped focus constricting the lumen (Fig. 2). A tiny calcification presumably at the site of the web can be demonstrated. In segmental obstruction of inferior vena cava, a hyperechogenic cord-like structure is seen to obliterate the hepatic portion of the vessel [6]. Similar appearance in hepatic veins is seen in cases of chronic thrombosis (Fig. 3).