Effect Of Ultrasound-Doppler On Diagnosis Of Budd-Chiari Syndrome in Behçet's Disease

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Özet: BUDD-CHIARI SENDROMU TANISINDA DOPPLER ULTRASONOGRAFİNİN ROLÜ

Bu çalışmada Behçet Hastalığı'na bağlı Budd-Chiari Sendromlu bir hastada, non-invasiv bir test olan Doppler-ultrasonografinin tanısal değeri araştırılmıştır. Doppler-sonografi ile intrahepatik, portal, hepatik ven ile vena kava inferiorun hemodinamik yapıları değerlendirilmiştir. Doppler-sonografi orta ve sağ hepatik venin parsiyel tıkalı olduğunu göstermiştir. Aynı bulgular anjiografi ile de teyid edilmiştir.

Sonuçlar, Budd-Chiari Sendromu'nun tanısında Doppler-Sonografinin hepatik ven ve vena-kava inferior hemodinamiğini değerlendirmede oldukça hassas bir test olduğunu göstermektedir. Böylece non-invasiv bir yöntem olan Doppler-Sonografik çalışma ile hepatik venografi veya inferior venacavagrafi gibi invasiv bir teste başvurulmadan Budd-Chiari sendromu tanısında önemli bir kolaylık sağlanabileceği sonucuna varılmıştır.

Anahtar Kelimeler: Budd-Chiari Sendromu, Behçet hastalığı, Tanı, Doppler ultrasonografi

Summary: This study was designed to evaluate Doppler-sonography imaging as potential non-invasive methods of diagnosis patient with Budd-Chiari Syndrome caused by Behçet's Disease. Doppler-sonography precisely defined intrahepatic, portal and inferior vena caval circulatory dynamics. These were confirmed by vena cavagram.

These results suggest that Doppler-sonography is excellent technique for the initial evaluation of patient suspected of having Budd-Chiari Syndrome in Behçet's Disease.

Key Words: Budd Chiari Syndrome, Behçet's Disease, Diagnosis, Ultrasound-Doppler Studies.

The Budd-Chiari syndrome is a rare disorder with a poor prognosis due to the occlusion of the hepatic veins or inferior vena cava, and it may be classified as primary or secondary on the basis of its pathophysiology (1,2). The primary type is due to congenital obstruction of the hepatic veins or inferior vena cava by membranous rings or webs. The secondary type refers to thrombotic obstruction of the aforementioned vessels and occurs often in patients with an underlying disease that predisposes them to thrombosis, such as hematologic disorders prolonged use of oral contraceptives, pregnancy and postpartum infections, trauma. In 25-30 %of cases the syndrome is classified as idiopathic (1,3).

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Originally described as a clinical triad of oral and genital ulcerations with iridocyclitis (4), Behçet's disease in now recognized as a multi systemic disorder (5-7). Vasculitis is thought to be the mechanism common to most Behçet's disease manifestations (8). Venous occlusion is frequent (5,6,9,10), but involvement of the hepatic ven is uncommon.

Many imaging techniques have been used to evaluate patients with suspected hepatic vein thrombosis, including liver scintigraphy, ultrasonography, angiography, computed tomography, and magnetic resonance imaging (11). Ultrasonography is certainly useful in the diagnosis of this syndrome(12,13), and it is considered the procedure of choice for emergency diagnosis(14).

Angiography has been the mainstay of diagnosis in patients with Budd-Chiari syndrome, both before and after surgery (15-17). Angiography, in fact, remains the definitive technique. Unfortunately, angiography is invasive and frequently the hepatic veins cannot be opacified beyond their ostia(18). Numerous less-invasive techniques, including nuclear medicine (2), sonography (12-14,19) CT (12,13,19), and most recently MR imaging (20), have had various degrees of success in characterizing the underlying vascular lesions in Budd-Chiari syndrome.

These radiographic techniques may all contribute important information about patients with Budd-Chiari syndrome. However, a portable, noninvasive examination would be desirable both before and after surgery. Doppler imaging is, therefore, an appealing technique in patients with Budd-Chiari syndrome. Duplex Doppler has been used with considerable success in the evaluation of the hepatic vasculature(21-24). Color-flow Doppler, only recently available for use in the abdomen, would also seem to have considerable potential.

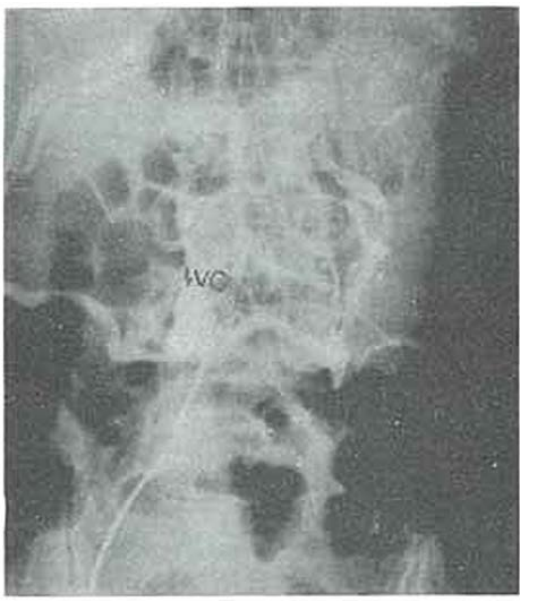

Figure 1: Inferior venocavagram confirms occlusion of inferior vena cava.

The purpose of this study is to describe in patient with Budd-Chiari syndrome caused by Behçet's disease, diagnosed by ultrasound and Doppler sonography.

CASE REPORT and METHOD

A 30 year-old man admitted for ascites. He suffered from recurrent buccal ulcerations and erythema nodosum. On admission, hepatomegaly, massive ascites, oral ulceration and scrotal scars were found. Liver function tests were normal. Angiography showed thrombosis of the inferior vena cava with extension into the right atrium (Fig. 1). Histological examination of a liver tissue specimen showed congested centrilobular sinusoids and hepatocyte necrosis.

In the patient, the diagnosis of Budd-Chiari syndrome was based on the existence of massive, predominantly centrilobular, sinusoidal congestion at histological examination of the liver and marked abnormalities of inferior vena cava as demonstrated by angiography,

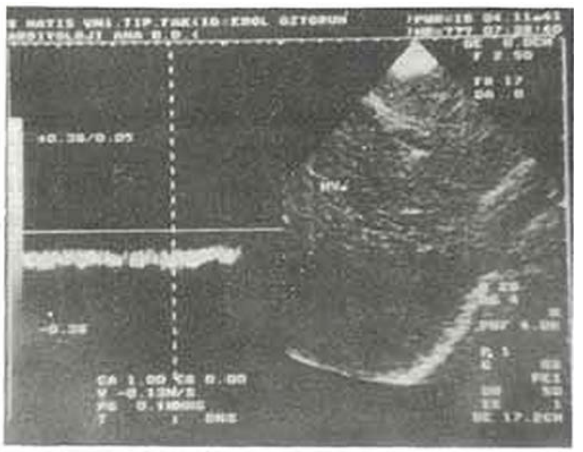

Figure 2: Doppler sonographic study demonstrates a continuous and slightly undulating signal pattern; peak velocity is 13cm/s in the middle hepatic vein.

Figure 3: Doppler sonographic study demonsrates a continuous slow without oscillations; peak velocity is 11 cm/s arrow in the right hepatic vein.

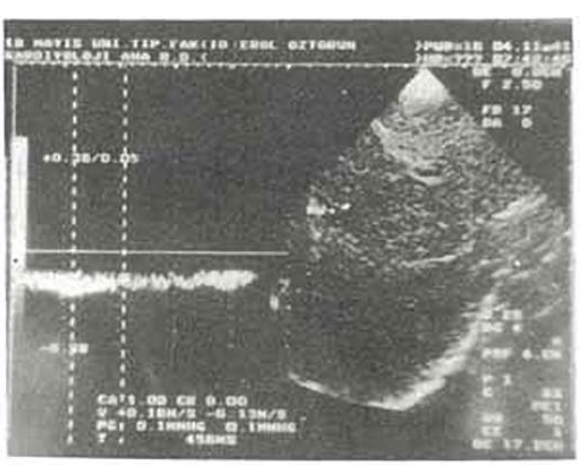

Figure 4: Doppler sonographic study shows normal flow pattern in the portal vein.

ultrasonography, computed tomography. Diagnosis of the cause of Budd-Chiari syndrome included the following investigations: 1) The search for a primary myeloproliferative disorder with blood cell count, total erythrocyte volume measurement, bone marrow biopsy, 2) Hemolysis tests performed for paroxysmal nocturnal hemoglobinuria, 3) an assay for antibodies to detect the lupus anticoagulant.

The patient with Budd-Chiari caused by Behçet's disease was seen. In this patient no myeloproliferative or coagulation disorder was found after all investigations described above had been performed and were negative since no specific test is available to as certain Behçet's disease, or two major and two minor criteria were required for the diagnosis of Behçet's disease. Clinical features of the patient with Budd-Chiari syndrome caused by Behçet's disease are reported below.

In the case, ultrasound examination was performed using real time equipments (Toshiba Sonolayer 160 ultrasonography, JAPAN) and 3.5 MHz sector probe with pulsed Doppler flowmeter operating at center frequency of 3.5 MHz. Doppler examination was made with the sampling volume cursor brought in the center of the vessel and placed at an angle 60° the longitudinal axis of the vessel. The direction of blood flow was considered toward or away from the transducer (or the source of Doppler bean), depending on whether the doppler signal was positive or negative, that is, above or below the baseline.

Ultrasound examination of the abdomen showed an anlarged liver with a few hypoechoic areas with indented edges. The normal portal veins and ascites were demonstrated. Abdomen ccmputed tomography scan without contrast enhancement demonsrated inhomogeneous density of liver parenchyma; a thrombosis in the vena cava. Hepatic veins not be correctly evaluated.

In case, Doppler sonographic examinations was performed without knowledge of previous imaging/angiographic findings.

The patient underwend Doppler sonography, which showed in the middle and right hepatic veins a continuous slow with very slight variations of velocity (peak velocity was 13 cm/s in the middle an 11 cm/s in the right hepatic vein), directed towards the inferior vena cava. The flow patterns in the portal vein and hepatic artery were normal. Inferior vena cava could not be correctly evaluated. The Doppler findings were considered indicative of partial thrombotic obstruction of the middle and right hepatic and the patient was further evaluated to identify a possible underlying disorder (Figs. 2,3,4).

In the case occlusion of inferior vena cava was confirmed by venacavagram Fig. 1.

DISCUSSION

Ultrasonography is the diagnostic procedure when Budd-Chiari syndrome is suspected, and some authors believe that a definitive diagnosis can be made by sonography alone (12,14). In most chronic cases hepatic veins are not visualized, whereas in all acute cases at least one major hepatic vein is usually identified showing abnormalities suggestive of this syndrome: stenosis, partial dilatation, thick wall echoes, abnormal course, extrahepatic anastomoses, and thrombosis. The inferior vena cava may be seen compressed by the liver or obstructed by thrombotic material. Hepatic parenchymal changes include atrophy of the right lobe, enlargement of the caudate lobe, and hypoechoic focal areas due to intraparenchymal hemorrhage (11,13). The most important limitations of sonographic evaluation are the difficult visualization of the hepatic vein rings and webs and the underestimation of caval impairment (14). Duplex Doppler allows demonstration of possible alterations of the flow pattern in the portal vein, hepatic veins, and inferior vena cava; however, little attention has been given to its diagnostic usefulness in Budd-Chiari syndrome (26,27). Recently, duplex and color Doppler sonography have been shown to be promising in the evaluation of decompressive portacaval and mesoatrial shunts therapeutically used in this syndrome: normal patency is suggested by hepatofugal flow in the portal vein and triphasic Doppler signal at the anastomotic level (26,28).

In the case reported here, ultrasound examination and Doppler sonography allowed a correct diagnosis of Budd-Chiari syndrome. Ultrasound study showed abnormalities suggestive of this syndrome and duplex Doppler demonstrated in the middle and right hepatic veins a continuous slow flow directed towards the inferior vena cava. In the hepatic veins of normal subjects Doppler signal is normally triphasic and there are wide variations in flow velocity and direction in response to both respiratory and cardiac cycles: flow increases during, inspiration and decreases during expiration (26,29). In Budd-Chiari syndrome altered blood flow patterns may be shown in the hepatic veins: the pattern of continuous slow flow observed in this patient is probably indicative of partial obstruction; the absence of Doppler signals suggests subtotal or total occlusion, in the hepatic veins(11,30).

The demonstration of these qualitative changes, with or without directional canges of blood flow, is porbably diagnostic of Budd-Chiari syndrome.

In concsluison, our case of Budd-Chiari syndorme demonstrate that Doppler sonography show in the hepatic veins a flow pattern highly suggestive of partial thrombotic obstruction. Doppler sonography can bu used to diagnosis Budd-Chiari syndrome without hepatic venography or inferior cavography.

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