A Budd Chiari Syndrome Case with A Thrombus in A Strategic Place of Inferior Venacava

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Özet: *INFERİOR VENA KAVANIN STRATEJİK BİR BÖLGESİNDEKİ TROMBUSE SEKONDER GELİŞEN BUDD-CHIARİ'Lİ BİR OLGUNUN TAKDİMİ*

Budd-Chiari sendromu genellikle kötü prognozla gittiği bilenen bir hastalıktır. Hastalık hepatik venöz dönüşte herhangi bir şekilde veya lokalizasyonda engellenme sonucu oluşmaktadır. Myeloproliferatif hastalıklar, Behçet hastalığı, paroksismel nokturnal henoglobunüri, inferior vena cava (IVC)'nın membranöz obstruksiyonu sayılabilecek etyolojik sebepler arasındadır. Hastalar genellikle karın ağrısı, asit ve hepatosplenomegali ile başvururlar (9). Genelde tedavide cerrahi ön planda tutulmakla beraber, bazı son çalışmalarda antlagreganlar, antikoagulanlar ve diğer tıbbi tedavilerle hastalara oldukça faydalı olulnduğu bildirilmektedir (1). Bu hastada ilginç yan inferior vena cavanın tam atrıuma dökülmleden önce bir trombus tarafından tam tıkalı olmasıdır ve beraberinde bütün hepatik ven dallarının ultrason (US) da dilate olduğunun tespitidir.

Anahtar kelimeler: Budd-Chiari sendromu, trombus,l inferior vena cava.

Summary: *Budd-Chiari syndrome (BCS) is seen as a disease with a poor prognosisi. The syndrome is considered to be the result of any kind of oblstruction involving hepatic venous outflow. One can encounter myeloproliferative disorders, Behçet's disease, paroxismal nocturnal hemoglobinuria, membranous obstruction of inferior vena cava (IVC) among the etlologic factors. The patient usually presents with abdominal pain, ascitis, hepatosplenomegaly (9). Tle general attitude to these patients in wlew of treatment is generaliy surgery like portocaval shunts, but antiagregants, anticoagulants and other nedical measurements have been shown to bebeneficial in some recent studies (1). The interesting point in this patent is thet the cause of the syndrome is a thrombus just at the entry of inferior vena cava to right atrium, and accordingiy all hepatic veins were dilated on ultrasonography (US).*

Key words: , Budd-chiari syndrome, thrombus, inferior vena cava.

A thirty-seven years old man came to us with a right upper quadrant pain for approximately 11 years. This pain had been coming in various times, there had been no association with meals or effort, but sometimes it had been increasing after runnig. One year ago, Patient applied to another haspital andy hospitilized there for 38 days. The patient was referred to our hospital.

On admision, on physical exam: liver was 6 cm palpable under costal margin at mid claviculer line, its total vertical lenght was 15 cm. On US, hepatomegaly, left lob hypertrophy, non homogen parencyme, hepatic venous diatation in the there main branches were detected (Figure 1 hepatic venous dilatation in the three main branches). A mild splenomegaly was among the findings on US. Looking at the blood biochemistry; BUN: 31 mg(dl Creatinin : 1.9 mg/dl, Urinalysis: normal, Hb: 15.1 WBC: 6200 Platelet count: 291000. Alkaline Phosphotase: 262 and 289 ALT:45 AST:47 Total protein: 7.9 Albumin: 3.9 Bilirubin:0.3 (indirect) /0.1 (direct) Parasite: negatif Serum iron: 14u g/lt Total iron bindingğ capacity: 48 ug/lt Transferrin saturation: 29% PTZ: 15, HBs (+), Echocardigraphy: Normal Peripheric venous pressure : 5.7 cm H_2O Liver biopsy: Parancymal bazic structure was intact. Sinusoidal dilatation, congestion and dilatation in central vein and pericentral fibrosisi were present. Iron stainig was negatif. Staining for seticulin showed increased steining.

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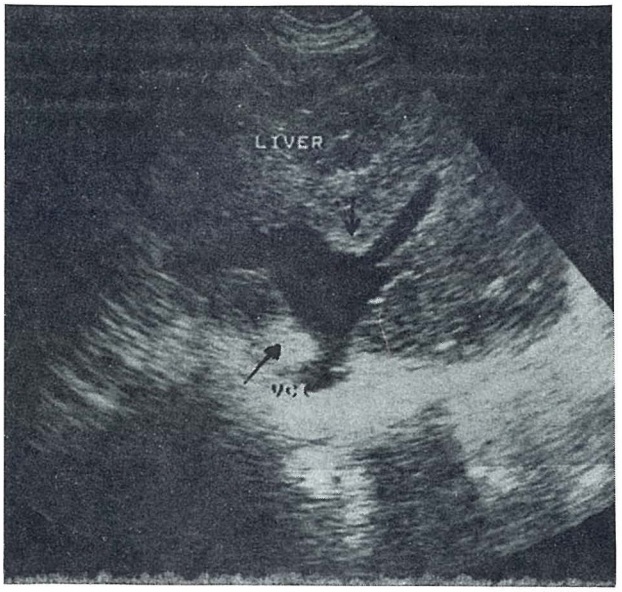

Figure 1: US shows dilatation of the hepatic veins (arrows) and obstruction of the inferior vena cava at the level of hepatic portion.

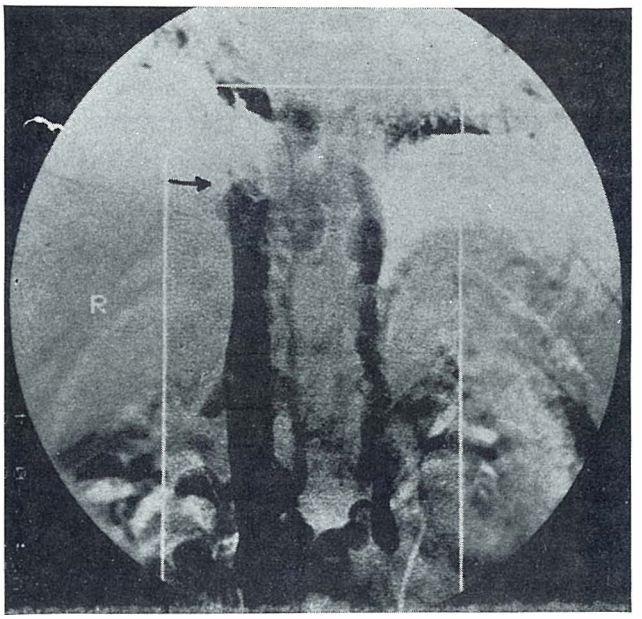

Figure 2: Inferior Cavagraphy with digital substraction angiography reveals a complete obstruction of inferior vena cava (arrow).

In inferior vena cavography, through femoral vein reaching appropriate area, we gave contrast material, observed thet just after the point where hepatic veins joins IVC, there is a total obstruction (Figure 2; the picture shows the total obstruction at the point just after the hepatic portion of inferior vena). We also saw the dilatation of the hepatic veins in the hepatogram taken at the same session.

At the clinical follow up, allthough there are literature mentioning about the utility of surgical produres inciude balloon angioplasty which was reported to be applied sucsesfully in some clinics together with local thrombolytic therapy and expandable metallic stents (5,6.7). we have prefered to be more conservative, and treated the patient medically. We gave the patient two antiaggregants, dipyridamol 75 mg tid and acetyi salycilic acid 250 mg each day. We followed the patlent for alimost two and a haif years. At the end of this period, There was not any further enlargment in liver. Left lob hypertrophy, caudat lob hypertrophy, dilatation of three main hepatiç veins and their branchers were importent findings. Almost one and a half year leter, some of the lab results were as ALT: 24, AST: 29, ALP: 219, total bilirubin: 1.2, direct bilirubin: 0.4. At last. one mount ago these parameters were alimost same.

DISCUSSION

This case is not approprite for the classic definition of BCS, which is considered to be composed of cases with involvement of hepatic veins. But according to the new definition (2), one can name this case BCS. The new definition is thet, a case is nemed BCS, if there is an obstruction to hepatic venous outflow in any place until it reaches right atrium.

Our case is diferent from classic BCS cases and amoneg the patients who are seen rarely. With the inferior venocavography we saw that there was an obstruction at a vary strategic place. During our research, we could'nt find a causative factör for the formation of thrombus. İn Turkey, one shouid think of Behçet's disease thet can present with this clinical symptoms and findings (1,4). But in our patient there was no arthritis, eye involvement, history of oral genital aphtous ulcerations. Also patergy test was negatif. Seeing the blood celis parameters we did'nt think of a myeloproliferatve disorder. Although it was reported in some recent literature that it couid be understood whether there was a latent myeloproliferative disease.

The way of doing this is by performing an eryroid colony cuiture and chromozomal studying. İt is reported that growing of erytroid colonles in

the absence of exogen erytropoetin indicates the presence of a latent polycytemia vera (3). Among suspicious causes is paroxismal noktturnal hemoglobuniria which also couid'nt be found in our patient. İts absence was decided after seeing the negativity of both acid ham test and sucrose lysis test. What we saw in inferior venacevvography was'nt considered to be a membranous obstruction because thet was a total obstruction. Bùt we should'nt be so sure about whether the obstuction was due to a thrombus, or a membrane with around a thrombus. Becuse in a recent study, M. Kage et al examined 17 autopsy cases of BCS with membranous obstruction. At the end of study they stated that although it was generally accepted thet membranes were congenital, this didn'nt explaine the late onset of the disease. They saw there had been thrombus around membranes in 7 of 9 cases. And in some cases they decided thet solely a thrombotic process and its sequelas were responsable from the occlusion, but not an indication of congenital malformation was present (8). These patients can be searched for constrictive periarditis and chronic liver disease for a long time, as in our case (10). Although the possibillity of finding of a thrombus in such a cause and web in IVC, one should do cavografy, especially if there is clinical situation suitable to BCS. As in our case ailthough one encouters thet a patient has been HBs Ag positive, got hepatomegaly, mildiy increased liver enzymes and bilirubin leveis, that's why can attribute this picture to chronic active hepatitis due to hepatitis B virus in the first place. But this case shows us the importance of making a biopsy first of all, even if patient is in a situation as described above.

We want to point out that in this case report, we presented an interesting case with respect to the place of thrombotic occlusion. it ıs in a very strategic place which was reported rarely. Secondly in patients taking medical therapy, we show that the expectancy of a good survival in a relatively healty manner is more then previousiy belleved. So we now seriousiy think of giving chance of medical treatment to such patients more then before.

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