ESSENTIAL THROMBOCYTOSIS AND BUDD-CHIARI SYNDROME A CASE REPORT AND A REVIEW OF THE LITERATURE

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SUMMARY

A 30 year-old female with a delayed diagnosis of essential thrombocytosis who developed Budd-Chiari syndrome following a healthy baby delivery is reported in this article. The patient had two stillbirths prior to the diagnosis of her primary hematologic disease.

Key Words : Essential Thrombocytosis, Budd-Chiari Syndrome, Pregnancy.

INTRODUCTION

Essential thrombocytosis is a myeloproliferative disorder characterised with increased megakaryocyte production and abnormal platelet functions. It is commonly seen in the middle age and sex ratio is equally distributed. Platelet count may rise 15 fold from the baseline level (1,2). Thrombotic and vascular complications due to increased platelet counts is the main cause of clinical manifestations. Thrombosis and hemorrhage are common findings. Thrombotic complications which may involve venous or arterial system are frequently seen in older patients (3). Epistaxis and gastrointestinal system bleedings are frequently seen hemorrhagic complications (4). Severe hemorrhage may occur following major trauma. Petechias are uncommon (1,2,5).

Bud-Chiari syndrome is a clinical condition which is characterised by inferior vena cava or hepatic venous occlusion, refractory ascite and hepatomegaly and usually seen secondary to various disorders (6). This condition has been reported to be responsible for 23 per cent of cases of hepatic outflow obstruction in a series from Los Angeles (7). It usually occurs in hypercoagulopathic conditions such as myeloproliferative diseases. Altough it mimics cirrhosis because of elevated transaminase levels, icter and portal hypertension, histopathologic examination reveals centrilobuler congestion. It is essential to treat the underlying disease. Shunt procedures may be required for intractible condition to the medical treatment (8).

CASE REPORT

A 30 year - old female patient, suffering from abdominal distension, was hospitalised. She had two stillbirths prior to the diagnosis of essential thrombocytosis in the second trimester of her last pregnancy. She was scheduled on weekly thrombocytopheresis to keep platelet count at normal levels. This procedure had been continued until the 8th month of her pregnancy when she underwent cesarean section leading to a healthy delivery in another hospital. One month after the delivery she was transferred to our hospital due to increased platelet count, ascites and jaundice. (there was a 3 fold increase in bilirubin levels and an 8 fold in transaminase levels from normal values). Viral markers of hepatitis (HBsAg, AntiHBs Ab, AntiHBc Ab, HBe Ag, AntiHBe Ab, AntiHVC Ab) were negative. Main hepatic veins and their branches were thrombosed in doppler ultrasonographic examination. Grade I varices were seen by esophagoscopy. Protein-C, S and antithrombin-III levels were found in normal range. LAPA (Leukocyte alkaline phosphatase activity) was elevated.

Hydroxiurea 3 gr/daily was administered to the patient and adjusted according to thrombocyte count. Furosemide and spironolactone were given. Hypoalbuminemia was restored by albumin replacements. Heparin (fraxiparine) was administered as anticoagulant. All branches of hepatic vein recanalised after anticoagulation (Fig.1). In spite of radiologic improvement there was no regression in portal hypertension and ascites. The left hepatic lobectomy and mesoatrial shunt was performed. Budd-Chiari syndrome was reported after histopathological examination.

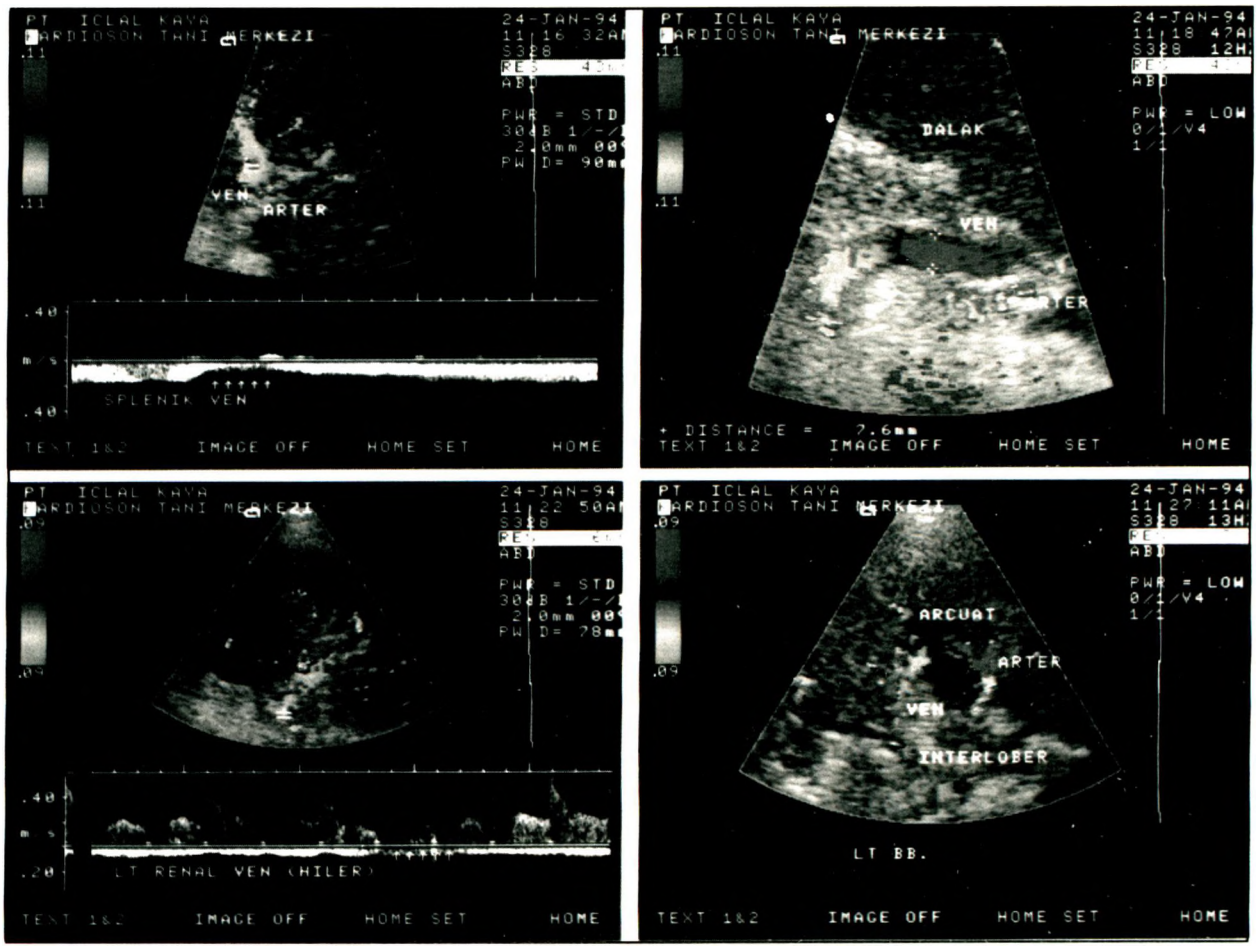

Fig 1. Doppler ultrasonographic appearance of branches of hepatic vein after anticoagulation

DISCUSSION

Essential thrombocytosis has thromboembolic and hemorrhagic complications due to elevated thrombocyte count. Thrombosis may occur practically in any vessel. It occasionally occurs in hepatic veins (1,3,6). Pregnancy is an additional predisposing factor. There were two stillbirths in this patient.

Budd-Chiari syndrome usually occurs in the hypercoagulopathic conditions such as myeloproliferative disorders and PNH or protein C/S deficiency. There is intractible ascites, jaundice and centrilobuler congestion in histopathologic examination (10,11). There is neither virologic marker positivity in this patient nor anticoagulant protein deficiency. After diagnosing Budd-Chiari syndrome, the anticoagulant therapy was administered to the patient. The main hepatic vein and their branches were recanalised after three weeks therapy but there was no change on the patient's clinical condition and the signs of portal hypertension. It was considered that this situation may be due to either hepatic degeneration that occurred before recanalisation or the peripheral hepatic veins were occluded .It was diagnosed as the irreversible liver paranchymal disease by the liver needle biopsy.

The risk of the abortion is very high in essential thrombocytosis so the platelet levels should be kept in normal range in order to minimize the risk of the abortion in pregnant women with essential thrombocytosis.

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