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Case Report

Hepatic venous outflow block in a young patient with Systemic Lupus Erythematosus

Ali Ghavidel*[1]

[1] *Assistant Professor, Liver and Gastrointestinal Diseases Research Center, Imam Reza Hospital, Tabriz University of Medical Sciences, Tabriz, Iran*

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Abstract

Introduction: Hepatic venous outflow block or Budd-Chiari syndrome is a severe liver disease with a 3 years survival rate of 50%. Several conditions have been implicated as a cause of Budd-Chiari syndrome, including myeloproliferative disorders, paroxysmal nocturnal hemoglobinuria, the presence of lupus anti-coagulant, oral contraceptives, pregnancy, and others. In a small number of cases, Budd-Chiari syndrome is associated with the presence of lupus anticoagulant. Anticardiolipin antibodies (ACA) are similar to lupus anti-coagulant antiphospholipid antibodies (APLAs), which have been described in patients with recurrent arterial and venous thrombosis, thrombocytopenia, fetal loss, or miscarriage.

Case Report: A 23-year-old woman is reported with Budd-Chiari syndrome in whom lupus anticoagulant and anticardiolipin antibodies were shown; 9 months after diagnosis of systemic lupus erythematosus (SLE) treatment with steroids admitted with gastrointestinal problems, abdominal pain and ascites and treated oral anticoagulants induced a considerable improvement. This treatment was continued after 1 year, but interruption was followed by redevelopment of ascites. Further treatment with anticoagulants was continued for 5 years with noticeable improvement.

Conclusion: Patients with Budd-Chiari syndrome should be tested for lupus anticoagulants and anticardiolipin antibodies, Budd-Chiari syndrome resulting from this cause may have a good response to treatment with oral anticoagulants; this treatment should be maintained permanently, and pregnancy in such patients may initiate serious difficulties. The condition of the patient at follow-up was good.

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Introduction

Hepatic venous outflow block was first described in 1845 by George Budd to describe the classic triad of abdominal pain, hepatomegaly, and ascites. Its association with systemic lupus erythematosus (SLE) was first reported in 1986 by Averbuch and Levo.[1,2] The reaction between the antibody and antigen could result, on the one hand, in inhibition of clotting factors and thrombocytopenia, and on the other hand, in inhibition of prostacyclin production, release of procoagulant activity, and enhanced thrombosis.[3,4] The pathogenesis of thrombosis is unknown but the proposed mechanisms include: direct endothelial cell injury, antigen-antibody mediated platelet activation, and inhibition of endogenous anticoagulants such as protein C, dysfunction of the coagulant cascade, induced by oral contraceptive use, and some other mechanisms.[5]

The acute syndrome presents with rapidly progressive server upper abdominal pain, jaundice, hepatomegaly, ascites, elevated liver enzymes, caudate lobe hypertrophy, and eventually encephalopathy. The clinical presentation of patients is governed by the

*** Corresponding Author:** Ali Ghavidel, Email: ali.ghavidel3@gmail.com

extent and swiftness of hepatic outflow obstruction juxtaposed to the body's ability to decompress the liver via development of collateral blood flow.[6,7] When hepatic venous outflow block is suspected, measurements are made of liver enzyme levels and other organ markers (creatinine, urea, electrolytes, and lactate dehydrogenase).

Physical examination and laboratory studies are not specific for the disease Doppler ultrasonography must be the initial choice because of its high sensitivity and specificity for determining the obstruction site and venous flow pattern in hepatic veins or inferior vena cava (IVC). A system of venous collaterals may form around the occlusion that may be seen on imaging as a spider's web.[2,8] Hepatic venography is a reference procedure, but recently this invasive technique to measure venous pressure.

A minority of patients can be treated medically with sodium restriction, diuretics to control ascites, anticoagulants such as heparin and warfarin, and general symptomatic management. The majority of patients require further intervention. Milder form of Budd-Chiari syndrome may be treated with surgical shunts to divert blood flow around the obstruction or the liver itself.[9,10]

Liver transplantation is an effective treatment for Budd-Chiari. It is generally reserved for patients with fulminant hepatic failure, failure of shunts, or progression of cirrhosis that reduces the life expectancy to 1 year.[5] Long-term survival after transplantation ranges from 69 to 87%. Up to 10% of patients may have a recurrence of Budd-Chiari syndrome after the transplant.

Several studies have attempted to predict the survival of patients with Budd-Chiari syndrome. In general, nearly 2/3 of patients with Budd-Chiari are alive at 10 years.[11] Survival is also highly dependent on the underlying myeloproliferative disorders may progress to acute leukemia, independently of with Budd-Chiari syndrome. Branch et al. have observed thrombotic episodes in 20 out of 29 SLE patients. About 15 patients had a history of one or more arterial thrombotic episodes. Nearly, 9 patients had central nervous system involvement, 2 coronary artery thrombosis, 2 visceral infarctions involving the pancreas or the spleen, and 7 had one or more episodes of deep vein thrombosis.[12] Association of Budd-Chiari syndrome with other autoimmune diseases has been reported in many studies.[13]

Case Report

The patient is a 23-year-old woman, married and has one child. She is reported with Budd-Chiari syndrome in which lupus anticoagulant and anticardiolipin antibodies (ACA) were shown. After 8 months of diagnosis of SLE and medical treatment with steroids in this period, she referred to hospital due to digestive problems, abdominal swelling and pain and full tests of blood, urine, endoscopy, ultrasound, X-rays, and computed tomography scans (CT scan) were performed on patients and the results demonstrate the Budd-Chiari Syndrome.

According to the experimental results, anti-nuclear antibody was positive. Anemia, hypoalbuminemia, proteinuria, hyperbilirubinemia, and several other parameters were observed. Imaging results also showed venous thrombosis in the IVC. Patient with very bad abdominal pain and ascites and underlying SLE disease referred to the hospital. After performing clinical examinations and various tests, Budd-Chiari syndrome was diagnosed. For the treatment, oral anticoagulants induced a considerable improvement. This treatment was continued after 1 year; but interruption was followed by redevelopment of ascites. Further treatment with anticoagulants was continued for 5 years with noticeable improvement. Paracentesis was used in order to control ascites fluid and various medications. After 2 weeks of diagnosis, patients got nephrotic syndrome. Patient's response to medicines including cyclophosphamide, prednisolone, warfarin, furosemide, and spironolactone are positive, and the patient's condition is improving.

Discussion

Budd-Chiari syndrome is characterized by

ascites, hepatomegaly, abdominal pain, varicose veins on the lower legs, and collateral vessels at the abdominal wall because of the obstruction of the hepatic vein or the IVC.[1,3,7,8] The syndrome was described, at first, by Budd and Chiari in 1845 and 1899, respectively and has been mainly reported in East Asia, South Africa, and India.[13,14] The pathogenesis of vascular obstruction is still unclear but some cases of membranous obstruction of the IVC have been reported in Korea, and some cases have been known to have a relationship with oral contraceptive agents[15] or type B viral hepatitis.[14] Recently, Budd-Chiari syndrome associated with lupus anticoagulant, has been reported in many countries,[11,12] but Budd-Chiari syndrome, as a manifestation of secondary antiphospholipid (APL) syndrome in SLE, is rare and has been reported in literature.

The patient has secondary Budd-Chiari syndrome due to SLE. In this instance, the differential diagnosis is often difficult between lupus vasculitis, thrombotic thrombocytopenic purpura, or disseminated intravascular coagulation.[15,16] To eliminate immunosuppression with pulse cyclophosphamide, plasmapheresis or gamma globulin infusion is sometimes effective, but there is usually a rapid rebound to pre-treatment levels shortly after discontinuation of the therapy. Anti-hypertensive, cholesterol-lowering agents, and treatment of active nephritis are needed to remove additional risk factors. It is important to prevent occlusive vascular complications in patients who have not experienced thrombosis. Long-term use of aspirin is useful in the prevention of arterial occlusion. For the prophylaxis of venous thrombosis, low-dose subcutaneous heparin (10000-15000 units/day) is useful and patients who show resistance to the above doses require higher doses of subcutaneous heparin (25000 units/day) or intravenous heparin (40000 units/day).[14,16]

In the case of occlusive vascular complications, patients require anticoagulation on a long-term basis, and probably for life, because of the tendency of recurrence. Warfarin is used up to 10 mg/day and the prothrombin time (PT) should be kept a level of 3 to 4 international normalized ratio (INR) rather than 2-2.5. In the case of using heparin, anticoagulation should be controlled by the re-calcification test (Howell test) not by activated partial thromboplastin time (PTT) because of the interference of these two tests. To prevent fetal loss, low-dose aspirin (30-80 mg/day) has proven useful and safe from the beginning of pregnancy.[15] If the patient is receiving warfarin, because of previous thrombotic manifestation, this must be changed to subcutaneous heparin, at least in the first trimester, since warfarin is teratogenic. Thrombocytopenia associated with APLs is usually mild and does not require intervention.[15,16]

Conclusion

Patients with Budd-Chiari syndrome should be tested for lupus anticoagulants and anticardiolipin antibodies. Budd-Chiari syndrome resulting from this cause may have a good response to treatment with oral anticoagulants; this treatment should be maintained permanently, and pregnancy in such patients may initiate serious difficulties. The condition of the patient at follow-up was good.

Conflict of Interests

Authors have no conflict of interest.

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