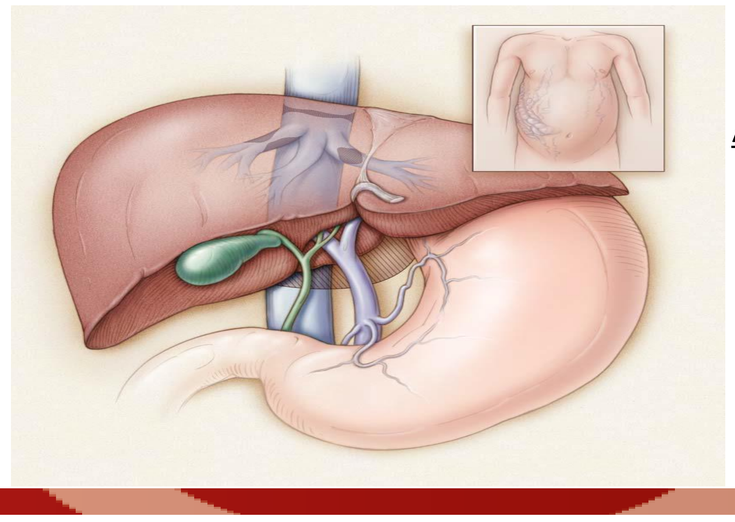
Recurrent Budd-Chiari syndrome (BCS) in a patient with polycythemia vera (PV), protein S deficiency and hyperhomocysteinemia (HH) and abnormal resistance to activated protein (APCR) in the absence of factor V Leiden mutation (FVL) following liver transplant



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Introduction:

Budd-Chiari syndrome (BCS) described in 1840 is a rare condition recognized in imaging studies and characterized by thrombotic obstruction of the hepatic venous outflow. The main cause of this syndrome are myeloproliferative diseases in particular polycythemia vera (PV) identified in 10-40% of cases. The latter may cause multiple splanchnic thrombosis, including portal vein thrombosis, particularly important for its clinical outcomes (ascites, collateral vessels genesis, etc.). Clinically it may manifest with the spectrum of symptoms; patients may remain asymptomatic for many years or may present with symptoms of fulminant hepatic failure. Imaging techniques pay a crucial role in the diagnosis of this syndrome.

Aims:

Presentation of the BCS clinical course in a patient with PV and additional thrombotic risk factors following liver transplant (LT).

Methods:

Factor V Leiden mutation was detected using RFLP/PCR method (EURx,Poland); APCR and protein S deficiency using Siemens reagent.

Case report :

A 35 year old patient with BCS and PV, progressive fulminant hepatic impairment as result of decompensated liver cirrhosis was subjected to orthotopic cadaveric liver transplant using piggy-back technique. Percutaneous endoscopic jejunostomy (PEJ) was used for nutritional support. Crossmatching was negative as no antibodies against lymphocytes were found in the recipient's serum. The transplanted organ took up function immediately after surgery. On the second day an inferior vena cava (VCI) obstruction was reported just below the right atrium. CT angiography revealed thrombotic obstruction of the hepatic venous outflow, hepatosplenomegaly, ascites that required paracentesis.

Gastroscopic findings included oesophageal varices grade 1, reduction in daily urine output, increased blood urea, HH (19.8 mmol/L; n: 5.0-12.0 mmol/l), hypoproteinaemia, hypoalbuminemia, protein S deficiency - 53% (n: 67.5-139%), APCR 0.48 (n: 0.7-1.2); absence of FVL (G1691A) mutation. Balloon technique was applied for VCI obstruction which resulted in resolution of ascites. Successful therapy followed with Enoxaparinum natricum twice a day, hydroxycarbamidum and immunosuppressive corticosteroids, Mycophenolas mofetil and Tacrolimusum.

Conclusion:

Protein S deficiency with HH and abnormal APCR in the absence of Leiden mutation may be considered risk factors for recurrence of hepatic vein thrombosis in a patient with PV following liver transplant.

References:

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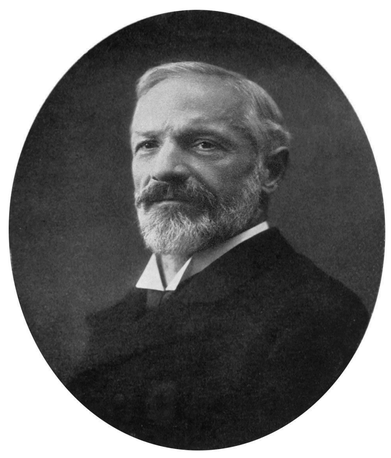

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