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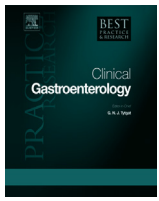

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Liver in haematological disorders

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A B S T R A C T

Prothrombotic haematological disorders, in particular myeloproliferative disorders, are identified in a significant proportion of patients with Budd–Chiari syndrome and portal vein thrombosis (PVT). Multiple prothrombotic disorders may coexist. PVT is diagnosed in one fourth of patients with cirrhosis and is more common with advanced liver disease and hepatocellular carcinoma. PVT in cirrhosis can precipitate decompensation. Intrahepatic microthrombosis may play a role in the pathogenesis of hepatic fibrosis. Sinusoidal obstruction syndrome is usually a complication of myeloablative treatment before haematopoietic stem cell transplantation. Post-transplant lymphoproliferative disorders can complicate liver transplantation and are related to Epstein–Barr virus infection. Hepatitis B reactivation in patients receiving chemotherapy for haematological malignancies is very common without pre-emptive treatment, and can lead to liver failure. Liver involvement is common in primary haematological diseases, such as haemolytic anaemias, lymphomas and leukaemia.

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Practice points

- Prothrombotic disorders can be identified in 80% of patients with Budd–Chiari syndrome and 50% of those with non-cirrhotic portal vein thrombosis (PVT). More than one prothrombotic disorders coexist in a significant proportion.
- Myeloproliferative disorders are strongly associated with splanchnic vein thrombosis (SVT).
- Testing for JAK2 V617F mutation contributes greatly to the diagnosis of occult MPD.
- The prevalence of PVT in patients with advanced liver disease is as high as 25%, and even higher with hepatocellular carcinoma.
- Cirrhosis is not a hypocoagulable state, as both pro- and anticoagulant factors are decreased.
- Cirrhosis has been associated with increased risk of thrombosis.
- PVT is a major cause of acute decompensation in patients with cirrhosis, and may have implication for liver transplantation.
- There is no established treatment for cirrhotic PVT, but anticoagulation and transjugular intrahepatic portosystemic shunts (TIPS) may have a beneficial role.
- Sinusoidal obstruction syndrome is usually a complication of myeloablative treatment before haematopoietic stem cell transplantation.
- The risk of post-transplant lymphoproliferative disorders (PTLD) is related to donor and recipient EBV status.
- There is an association between hepatitis C virus (HCV) infection and B-cell-non-Hodgkin's lymphoma.
- Exacerbation of viral hepatitis in patients receiving chemotherapy is more common with rituximab. Hepatitis B reactivation can be life-threatening and pre-emptive treatment is required.
- Liver involvement in haematological malignancies is not uncommon, but primary hepatic lymphomas are rare.

Research agenda

Further research is required to elucidate the following areas:

- The role of novel JAK2 mutations in the pathogenesis of SVT.
- The role of micro-thrombosis in the pathogenesis of hepatic fibrosis.
- The efficacy and safety of anticoagulation for treatment of PVT in patients with cirrhosis.
- The role of TIPS in patients with partial PVT to prevent complete PV occlusion.
- The efficacy of defibrotide for treatment of SOS.
- The role of EBV DNA monitoring for early PTLD diagnosis.
- The risk of PTLD with different immunosuppression regimens.
- The pathogenesis of HCV-related lymphoma and the role of antiviral treatment.
- The significance of HCV exacerbation in patients receiving chemotherapy and the potential prophylactic role of novel antiviral agents.

Introduction

Recent research for the cause of vascular disorders of the liver, such as Budd–Chiari syndrome (BCS), portal vein thrombosis (PVT) and sinusoidal obstruction syndrome (SOS), with the contribution of novel diagnostic modalities has shed light on the cardinal role of haematological disorders in their pathogenesis. There are primary liver diseases that can be complicated by haematological disorders, such as post-transplant lymphoproliferative disorders (PTLD), HCV-related lymphoma, and