A case of idiopathic portal vein thrombosis?


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CLINICAL PRESENTATION
A 21-year-old Caucasian male presented with a short history of epigastric pain and weight loss. Physical examination revealed splenomegaly only. Renal biochemistry and full blood count were normal. There were minor elevations in liver enzymes. A contrast-enhanced abdominal CT scan identified splenomegaly with infarcts, and portal and splenic vein thrombosis without cavernoma formation (figs 1 and 2). A thrombophilia screen comprising anticardiolipin immunoglobulin G (IgG) and lupus anticoagulant, factor V Leiden and prothrombin gene mutation analysis, protein C and S quantification and screening for paroxysmal nocturnal haemoglobinuria was normal. Bone marrow architecture was normal, with a non-significant increase in megakaryocyte count. The BCR-ABL oncogene was absent.

The patient was anticoagulated and monitored as an outpatient. The liver biochemistry normalised and full blood counts remained persistently normal. Figures 3 and 4 are from a repeat CT scan performed 5 years after the initial presentation.

QUESTIONS
What progressive changes have occurred in the 5 years between figs 1 and 2, and 3 and 4? Which additional test may determine the cause of this otherwise idiopathic portal vein thrombosis, and how may its findings alter the management?
See page 136 for answers

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ANSWER

From the question on page 111

As indicated by the arrows, figs 5 and 6 demonstrate the formation of a portal venous cavernoma and collateral circulation. There is persistent splenomegaly with resolution of the initial splenic infarcts.

Hypersplenism and increased plasma volume following a portal vein thrombosis (PVT) can limit the ability of conventional criteria to diagnose an underlying myeloproliferative disorder. Janus-activating kinase 2 (JAK2) mediates erythropoietin receptor signal transmission in haematopoietic cells. The V617F mutation of JAK2, first described in 2005, affords haematopoietic cells an uncontrolled proliferative and survival advantage. The mutation is diagnostic of a myeloproliferative disorder, and is present in ~95% of patients with polycythaemia vera and 50% of essential thrombocythaemia and idiopathic myelofibrosis. Twenty-one per cent of patients with an otherwise idiopathic PVT have the V617F mutation.

This patient had the V617F mutation, and hence was diagnosed with a non-overt myeloproliferative disorder. These patients require close follow-up due to the high development rate of an overt myeloproliferative disorder. As warfarin may fail to fully prevent further thromboses following a PVT, patients with the mutation may potentially benefit from combined aspirin and warfarin therapy. It is known that patients with varices and underlying cirrhosis have a fourfold increased risk of portal hypertensive bleeding following use of >300 mg of aspirin per day. The authors are not aware of any published data on the bleeding risk of low-dose aspirin in the setting of non-cirrhotic portal hypertension.

Acute variceal haemorrhage occurred 4 months after starting combined low-dose aspirin/warfarin treatment. This was despite the prophylactic use of propranolol and a proton pump inhibitor. The episode was successfully managed with band ligation and aspirin was discontinued. To date, there has been no recurrence of bleeding or further thrombosis.

REFERENCES


Figure 5 Follow-up axial CT image, performed 3 years after presentation. The image demonstrates portal venous cavernomatous transformation within the liver, and collateral circulation development at the hepatic and splenic hila (indicated by arrows).

Figure 6 Follow-up coronal reconstruction CT image, performed 3 years after presentation. A spleno-renal shunt has developed (indicated by an arrow) from a distended left renal vein.