Case Report
Extra Hepatic Portal Vein Thrombosis in a Child Associated with Lupus Anticoagulant

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Summary
The presence of lupus anticoagulant has been implicated in venous as well as arterial thrombosis. We report here a 10-year-old boy who presented to us with hematemesis, malena and splenomegaly. An ultrasound showed a recanalized portal vein with collaterals suggestive of portal vein thrombosis. He had grade IV esophageal varices. The liver function tests were normal. Investigations for prothrombotic factors showed that tests for PNH and for APC resistance were negative. Levels of anti-thrombin III and protein C were normal. There was a prolonged activated partial thromboplastin time with a normal prothrombin time. Presence of lupus anticoagulant was confirmed with dilute Russell viper venom time and platelet neutralization test. Repeat tests after 10 weeks showed persistence of the lupus anticoagulant. ELISA test for anti-phospholipid antibody was negative. The association of lupus anticoagulant with portal vein thrombosis in the pediatric age group is very rare.

Introduction
Extra-hepatic obstruction due to portal vein thrombosis (PVT) is a common cause of portal hypertension in India. In children the most common causes are believed to be infection and umbilical sepsis; however, congenital vascular abnormality has also been implicated. In adults, besides infection, pancreatitis, trauma, severe enteritis, duodenal ulcer, malignancies, and biliary tract diseases are considered to be etiological factors. More recently, however, prothrombotic factors have been increasingly recognized as a cause of the disease. Lupus anticoagulant, an acquired defect, has been documented as a prothrombotic factor in both arterial and venous thrombosis. Only a few case reports of lupus anticoagulant associated with PVT have been reported. We report here a 10-year-old boy with PVT in whom lupus anticoagulant was identified.

Case Report
A 10-year-old boy presented with a history of four episodes of hematemesis over a period of 2 years preceded by fever each time. Eighteen months previously, he had jaundice for 5 days and splenomegaly was noticed. There was no ascites, pedal edema, or hepatic encephalopathy following any bleed. On clinical examination, he was well nourished, anicteric, and the spleen was palpable 5 cm below the left costal margin. There was no hepatomegaly or lymphadenopathy. On ultrasound, the splenic vein was 7 mm at the splenic hilum and 8 mm in the pancreatic bed. The portal vein had an irregular lumen with thick walls and was 13 mm in diameter and surrounded by collaterals. Retroperitoneal collaterals were also present. The gallbladder, pancreas, and liver were normal. Endoscopy revealed grade IV esophageal varices. A diagnosis of extra-hepatic obstruction with PVT was made.

The liver function tests were normal: serum bilirubin 0.4 mg/dl, serum aspartate amino-transferase 40 IU/ml, serum alanine amino-transferase 23 IU/ml, serum alkaline phosphatase 355 KAU/dl, and serum albumin 5.0 g/dl. Hepatitis B surface antigen was negative. He had a normal hemogram with Hb 125 g/l, hematocrit 37%, WBC 5.9 × 10^9/l, and platelet count 260 × 10^9/l.

Hematological tests for prothrombotic factors showed normal levels of anti-thrombin III and protein C, and the absence of activated protein C resistance. Ham’s test and sucrose water test for paroxysmal nocturnal hemoglobinuria were negative. Coagulation tests showed a normal plasma prothrombin time (patient 14/control 14 s) but the activated partial thromboplastin time (APTT) was prolonged (patient 53/control 53 s) on repeated occasions. Lupus anticoagulant was suspected and...
was confirmed with dilute Russell viper venom time and platelet neutralization test.\textsuperscript{12} The test for anti-phospholipid antibody (using a ELISA kit from Diagnostica Stago, France) was negative. A repeat test for lupus anticoagulants done 10 weeks later remained positive. Investigation for ds DNA was negative.

**Discussion**

Anti-phospholipid syndrome refers to a heterogeneous family of antibodies detected either by clotting tests (lupus anticoagulant) or by specific immunoassay of anti-cardiolipin and anti-beta2 glycoprotein 1.\textsuperscript{1,13} Lupus anticoagulants are antibodies against prothrombin bound to anionic phospholipid and are generally identified on the basis of their capacity to prolong the phospholipid-dependent coagulation tests.

Lupus anticoagulant is strongly associated with arterial and venous thrombosis and thrombo-embolism.\textsuperscript{8,9} However, reports of an association of lupus anticoagulants with splanchic vein thrombosis, such as hepatic vein thrombosis (HVT) and PVT, are very few. Three adult patients with PVT\textsuperscript{10,11} have been reported, of which two patients (a 20-year-old male and a 30-year-old female) were from India.\textsuperscript{11} Three adult patients with HVT have also been reported in association with lupus anticoagulants.\textsuperscript{15} However, in the pediatric age group, PVT with lupus anticoagulant is very rare and only a single case has been documented.\textsuperscript{16}

Investigations to find the cause of thrombosis in splanchic vein thrombosis, including PVT presenting as extra-hepatic vein obstruction, should include investigations for prothrombotic factors.

**References**