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CASE REPORTS

ANTIPHOSPHOLIPID SYNDROME (APS) WITH CHRONIC THROMBOSIS OF PORTOVENOUS SYSTEM AND EXTRA HEPATIC PORTAL HYPERTENSION - ROLE OF ANTICOAGULATION

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ABSTRACT
Antiphospholipid syndrome (APS) presenting with chronic thrombosis is rare in children. We present a 2-year-old child with APS who had chronic thrombosis of portovenous system with extra hepatic portal hypertension (EHPHT) and had multiple episodes of melena and hematemesis contraindicating use of anticoagulation.

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KEYWORDS
Antiphospholipid syndrome, APS, chronic thrombosis, extra hepatic portal hypertension.

Introduction

Antiphospholipid syndrome (APS) is a disorder characterized by recurrent venous or arterial thrombosis with characteristic laboratory abnormalities, such as persistently elevated levels of antibodies directed against membrane anionic phospholipids [i.e, anticardiolipin antibody (ACLA)] or their associated plasma proteins, predominantly beta-2 glycoprotein I (apolipoprotein H), or evidence of a circulating anticoagulant.[1] APS can occur in patients without evidence of any definable associated disease or in association with systemic lupus erythematosus (SLE) or another rheumatic or autoimmune disorder.[1,2] We present a 2-year-old boy with chronic thrombosis and positive ACLA leading to extra hepatic portal hypertension (EHPHT). Due to repeated episodes of malena and hematemesis, we were unable to give him anti-coagulation.

Case Report

A 2-year-old boy presented with previous 6 admissions for melena, hematemesis since 7 months of age. At 1 year of age, ultrasonography (USG) abdomen with colour doppler of portal system showed small size liver with coarse echotexture, splenomegaly, and non-visualisation of main portal vein (PV). At 15 months doppler showed replacement of main splenic vein (SV) by multiple collaterals around the pancreas, with collaterals around intrahepatic portal vein and superior mesenteric vein (SMV) was not visualised. At 17 months of age, CT portovenogram showed non-visualisation of PV, SV suggestive of total thrombotic occlusion with periportal collaterals. He had received multiple blood transfusions till date and had undergone endoscopic sclerotherapy twice for oesophageal varices. On presentation to us, in view of chronic thrombosis, thrombotic workup was done showed positive anticardiolipin antibody (ACLA) IgG (22 GPL units, Normal <15 GPL units) and IgM (10 GPL units, Normal <10 GPL units). Factor V leiden mutation was not detected and Protein C and Protein S levels were normal. Child was treated with propranolol and advised devascularisation surgery. He could not be started on any anticoagulation due to frequent gastrointestinal bleeds. Shunt surgery was also not advised due to risk of thrombosis in the shunt due to APS.

Discussion

APS is an acquired prothrombotic state where thrombosis is related to the presence of antiphospholipid antibodies. Venous thrombosis is most seen in deep or superficial veins of the legs and arterial thrombosis most commonly presents as cerebral infarct.[1,2] Cutaneous involvement may present as: livedo reticularis, splinter haemorrhages, leg ulcer, blue toe syndrome.[1] Central nervous system (CNS) affection may lead to multi-infarct dementia, chorea, transverse myelopathy, pseudotumor cerebri, cerebral venous thrombosis. Coronary artery disease, valve vegetations, intracardiac thrombus may be found due to cardiovascular involvement.[2,3] Rare cases of perinatal thrombosis in infants born to mothers with APS or mothers with antiphospholipid antibodies have been reported.[4] However portal venous thrombosis due to APS has rarely reported. Liver involvement is the most frequent abdominal manifestation associated with APS. Various hepatic manifestations have been reported including Budd-Chiari syndrome, hepatic-veno-occlusive disease and occlusion of small hepatic veins, nodular regenerative hyperplasia, hepatic infarction, cirrhosis, portal hypertension, autoimmune hepatitis, and biliary cirrhosis.[5]

Laboratory tests that APS is the presence of antiphospholipid antibodies (APL) antibodies i.e anti cardiolipin antibodies, anti–beta-2 glycoprotein I antibodies and lupus anticoagulants. Other

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antiphospholipid antibodies are anti-phosphatidylinositol, anti-phosphatidylserine, anti-phosphatidylcholine, antinuclear antibody, Anti-DNA.[6] Our patient had positive ALCA.

No prophylactic treatment is required for asymptomatic children in whom APL antibodies are incidentally found.[6,7] Recent data suggest a higher risk of thrombosis in children with elevated aPL and underlying systemic autoimmune disease, such as SLE or lupus-like diseases. Low-dose aspirin (at the antiplatelet dosage of 3-5 mg/kg/d) may be appropriate thrombo-prophylaxis in these patients. For acute thrombotic event, patients are anticoagulated with heparin followed by oral anticoagulants.[8] In our patient, due to recurrent episodes of hematemesis, anticoagulation or aspirin could not be given.

Compliance with Ethical Standards

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Conflict of Interest: None

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