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A RARE PRESENTATION OF ANTIPHOSPHOLIPID SYNDROME: RECURRENT UPPER GASTROINTESTINAL BLEEDING SECONDARY TO PORTAL VEIN THROMBOSIS

Rheumatology

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ABSTRACT

Background: Antiphospholipid syndrome (APS) is an autoimmune prothrombotic disorder characterized by recurrent venous or arterial thrombosis and the presence of antiphospholipid antibodies. While APS commonly manifests as deep venous thrombosis or cerebrovascular events, thrombosis of the portal venous system is rare, especially in the absence of cirrhosis. Portal vein thrombosis (PVT) can lead to non-cirrhotic portal hypertension, resulting in variceal bleeding, ascites, and hypersplenism. Early recognition of APS as an underlying cause of PVT is crucial for appropriate management and prevention of recurrent thrombotic events. **Case Presentation:** We report the case of a 25-year-old female with recurrent upper gastrointestinal bleeding since adolescence, presenting with melena, hematemesis, and abdominal distension. She had undergone multiple endoscopic variceal ligation sessions across earlier admissions. On evaluation, she was hemodynamically unstable with anemia and thrombocytopenia. Imaging revealed a dilated portal vein with extensive periportal and perisplenic collaterals, splenomegaly, ascites, and portal cavernoma formation. Contrast-enhanced CT confirmed thrombosis of the branches of the portal vein. Thrombotic work-up demonstrated elevated anticardiolipin IgG (60.93 U/mL), consistent with APS, while ANA was negative. Upper GI endoscopy showed Grade II/III esophageal varices, for which EVL was performed. The patient was managed with non-selective β-blockers, rivaroxaban, transfusions, and was advised devascularization surgery due to recurrent variceal bleeding. **Conclusion:** This case highlights a rare presentation of APS manifesting as non-cirrhotic portal vein thrombosis with recurrent upper gastrointestinal bleeding. In young patients without cirrhosis who present with variceal hemorrhage, APS should be considered an important differential diagnosis. Early thrombophilia evaluation and timely initiation of anticoagulation are essential to improve outcomes and prevent further thrombotic complications.

KEYWORDS

Antiphospholipid Syndrome, Portal Vein Thrombosis, Non-Cirrhotic Portal Hypertension, Variceal Bleeding, Anticardiolipin Antibody.

INTRODUCTION

Antiphospholipid syndrome (APS) is an autoimmune prothrombotic disorder characterized by recurrent venous or arterial thrombosis and/or pregnancy morbidity in the presence of persistently positive antiphospholipid antibodies, most commonly anticardiolipin antibodies, lupus anticoagulant, or anti-β2-glycoprotein I antibodies (1). It may occur as a primary disorder or in association with systemic lupus erythematosus or other autoimmune conditions (1,2). The hypercoagulable state induced by these autoantibodies predisposes patients to thrombosis in both typical vascular beds as well as unusual sites, including hepatic and mesenteric veins (2).

Portal vein thrombosis (PVT) is a clinically significant vascular disorder most frequently associated with cirrhosis, malignancy, abdominal infections, and myeloproliferative neoplasms. However, non-cirrhotic PVT is increasingly recognized as a separate clinical entity in which inherited or acquired thrombophilias—including APS—play a major etiologic role (3,4). APS-related PVT is rare and often underdiagnosed, especially in the absence of cirrhosis, leading to potential delays in management.

Recurrent upper gastrointestinal bleeding as the principal manifestation of APS-induced PVT is exceedingly uncommon. Only isolated reports have documented this presentation, emphasizing that APS should be considered in young patients presenting with variceal bleeding and portal hypertension without evidence of liver disease (5). In such settings, early identification of the underlying prothrombotic state is essential, as long-term anticoagulation is the cornerstone of management.

The case described here details a young woman with recurrent variceal bleeding since adolescence, ultimately found to have portal vein thrombosis with cavernoma formation and positive anticardiolipin IgG antibodies consistent with APS. This report highlights the importance of thorough evaluation for thrombophilias in non-cirrhotic portal hypertension and adds to the limited literature on APS-associated portal vein thrombosis. thrombophilias in non-cirrhotic portal hypertension and adds

Case Presentation

A 25-year-old female presented with complaints of melena, hematemesis, and progressive abdominal distension for eight days. She had a significant history of six prior hospital admissions for recurrent upper gastrointestinal bleeding since the age of 15 years , requiring multiple packed red blood cell transfusions and three sessions of endoscopic variceal ligation.

On admission, she was hemodynamically unstable, with blood pressure 88/70 mmHg, pulse 112 beats/minute, and oxygen saturation of 97% on room air. Laboratory evaluation revealed a hemoglobin level of 6.8 g/dL, leukocyte count of 2,850/dL, and platelet count of 100,000/dL. PT was 15.1 seconds (INR 1.2). ANA by immuno-fluorescence was negative. Thrombophilia work-up showed elevated anticardiolipin IgG (60.93 U/mL), compatible with antiphospholipid syndrome.

Ultrasound of the abdomen showed a dilated portal vein with multiple periportal and perisplenic collaterals, splenomegaly, and ascites. Portal vein Doppler demonstrated a dilated, tortuous splenic vein with portal cavernoma formation. Ultrasound elastography revealed a median liver stiffness of 7.1 kPa, supporting a non-cirrhotic etiology. Contrast-enhanced CT confirmed thrombosis of the branches of the portal vein, along with a dilated splenoportal axis, collateral formation, and mild ascites.

Upper gastrointestinal endoscopy revealed Grade II/III esophageal varices, gastric varices, and portal hypertensive gastropathy. EVL was performed. The patient was treated with propranolol, rivaroxaban, and transfused with three units of PRBCs. She was advised to undergo devascularization surgery due to recurrent variceal bleeding and extensive collateralization.

DISCUSSION

Antiphospholipid syndrome (APS) is a systemic autoimmune thrombophilia characterized by the presence of antiphospholipid antibodies and recurrent vascular thrombosis. Although venous thrombosis of the lower extremities is the most common manifestation, APS is also associated with thrombosis in atypical venous territories, including the splanchnic circulation (1,2). The presence of elevated anticardiolipin IgG in this patient strongly supports APS as the underlying etiology.

Portal vein thrombosis (PVT) is most frequently seen in association with cirrhosis, abdominal malignancy, or inflammatory conditions. In non-cirrhotic patients, thrombophilias—including APS—play a significant role in the development of PVT (3,4). The absence of cirrhosis in this patient, supported by elastography findings, emphasizes that APS-induced PVT should be considered in young individuals presenting with portal hypertension.

Chronic PVT leads to cavernoma formation, periportal and perisplenic collaterals, splenomegaly, and ascites—all of which were present in this case. These findings are consistent with extrahepatic portal vein obstruction (EHPVO) (4). The long-standing nature of the disease is highlighted by recurrent variceal bleeding since adolescence and multiple prior EVL sessions.

APS-related PVT is rare, and underdiagnosis is well documented. International guidelines recommend thrombophilia screening in all cases of non-cirrhotic PVT to prevent recurrent thrombosis and guide long-term management (5,6). Lifelong anticoagulation remains the standard of care in APS patients with thrombosis.

This case demonstrates the importance of maintaining a high index of suspicion for APS in young patients with unexplained portal hypertension and reinforces the need for a multidisciplinary approach involving hepatology, gastroenterology, and hematology.

CONCLUSION

APS should be considered an important but often overlooked cause of non-cirrhotic portal vein thrombosis, particularly in young patients presenting with unexplained variceal bleeding. This case demonstrates that APS may initially manifest with complications of portal hypertension, including recurrent upper gastrointestinal hemorrhage, cavernoma formation, and ascites. Early thrombophilia evaluation and timely initiation of anticoagulation are crucial to reduce thrombotic recurrence and improve long-term outcomes.

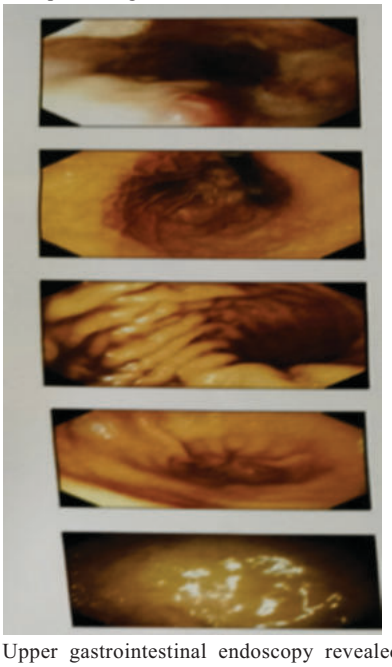

Figure 1: Upper gastrointestinal endoscopy revealed multiple columns of esophageal varices of grade 2/3, gastric varices and portal gastropathy.

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