Antiphospholipid Antibody Syndrome Presenting as Subacute Abdominal Pain Due to Portomesenteric Thrombosis

Thoguluva Seshadri Chandrasekar¹  Bollu Janakan Gokul¹  Thoguluva Chandrasekar Viveksandeep²  Kalamegam Raja Yogesh¹  Suriyanarayanan Sathiamoorthy¹  Menta Sanjeevaraya Prasad¹

¹Department of Medical Gastroenterology, MedIndia Hospitals, Chennai, Tamil Nadu, India
²SUNY Upstate Medical University, Syracuse, NY, United States

Abstract

Antiphospholipid antibody syndrome (APS), a hypercoagulable state, affects organ by causing venous or arterial thrombosis. We present an unusual case of a 58-year-old male who presented with diffuse abdominal pain and on evaluation diagnosed as having portomesenteric venous thrombosis due to primary APS. Upon successful treatment with enoxaparin followed by anticoagulants for 6 months, recanalization of the portal vein was documented by endoscopic ultrasonography. Early identification and treatment of portomesenteric thrombosis is crucial to prevent bowel ischemia. Lifelong anticoagulation with vitamin K antagonists should be considered in those patients with major thrombosis and established APS.

Introduction

Antiphospholipid antibody syndrome (APS) is a hypercoagulable state caused by antibodies against the phospholipids and proteins in the cell membrane. It can affect any part of the body by causing both venous and arterial thrombosis and in pregnancy it can lead to fetal loss. It can also cause thrombocytopenia, thrombotic microangiopathy, bleeding episodes, valvular heart disease, and cutaneous manifestations like livedo reticularis. We present an unusual case of portal and mesenteric vein thrombosis due to primary APS successfully treated with anticoagulants leading to recanalization of the portal vein (PV) and documented by endoscopic ultrasonography (EUS).

Case Report

A 58-year-old man presented with intermittent severe diffuse abdominal pain that worsened after food intake, for 1 month. The general physical examination was unremarkable. The abdominal examination revealed mild diffuse tenderness with sluggish bowel sounds. The rest of the systemic examination was normal. Except for an elevated erythrocyte sedimentation rate (25 mm in the first hour), his urine analysis, complete hemogram, hepatic and renal function tests, and electrolytes were within normal limits. An abdominal X-ray was non-contributory. A transabdominal ultrasonography and esophagogastroduodenoscopy were also normal. The presence of severe abdominal pain in the absence of overt signs prompted us to investigate for an intra-abdominal vascular event. Hence, a contrast-enhanced computed tomography (CECT) with abdominal angiogram was done. It revealed complete thrombosis of the superior mesenteric vein (SMV) and the proximal main PV involving the confluence without any collateral (†Fig. 1), suggesting a recent onset of thrombosis. Bowels were found to be viable. Otherwise the CECT of the abdomen was unremarkable. A EUS examination with linear array echo endoscope (Olympus Corporation Shinjuku Monolith, 2-3-1 Nishi-Shinjuku, Shinjuku-ku, Tokyo 163-0914, Japan)-confirmed PV thrombosis extending into the SMV (†Fig. 2).

In view of portomesenteric venous thrombosis, he was investigated for any underlying hypercoagulable state. His prothrombin time (PT) was found to be 15 seconds (control—14 seconds) and activated partial thromboplastin...
time was 40.2 seconds (control—30 seconds). Anticardiolipin (aCL) and β2 glycoprotein 1 antibodies (both immunoglobulin M and immunoglobulin G), serum homocysteine, antithrombin-III, protein-C, and protein-S levels were within the normal range. Antinuclear antibodies, rheumatoid factor, and factor V mutation were negative. Lupus anticoagulant (LA) test was positive by kaolin clotting time, dilute PT, and platelet neutralization method. A repeat test after 12 weeks also showed a positive result. He had no cutaneous rash, arthritis, or ophthalmic involvement to suggest any autoimmune disorders. A diagnosis of primary APS was established in this patient, with the clinical and radiological evidence of recent onset of portomesenteric thrombosis along with a positive LA test. Subcutaneous enoxaparin was started along with oral acenocoumarol, a vitamin-K antagonist. The patient became asymptomatic in a week’s time. During the follow-up period of 6 months, the PT and international normalized ratio (INR) levels were monitored regularly to maintain the

![Fig. 1](image1.png) **Fig. 1** Computed tomography abdomen showing complete thrombosis of the superior mesenteric vein (SMV) with extension of thrombosis to the proximal main portal vein with no evidence of collaterals.

![Fig. 2](image2.png) **Fig. 2** Endoscopic ultrasonography showing portal venous thrombosis.
INR between 2.0 and 3.0. A repeat CECT abdominal angiogram after 6 months revealed resolution of the thrombus in the main PV with recanalization and development of few collaterals draining from the SMV into the main PV (►Fig. 3) and a repeat EUS examination confirmed complete resolution of the PV thrombosis and restoration of blood flow (►Fig. 4).

Discussion
APS was first described in the year 1983 by Harris et al.1 Thrombosis of the mesenteric vein was first reported by Warren and Eberhard in 1935.2 Primary APS is not associated with any predisposing factor, whereas secondary APS is associated with autoimmune conditions like systemic lupus erythematosus or rheumatoid arthritis. According to the 2006 international criteria, the diagnosis of APS requires at least one clinical criterion: vascular thrombosis or pregnancy-related morbidity and one laboratory criterion with the presence of one of the three antibodies: LA, aCL, or anti-β2 glycoprotein-1 antibodies, measured on two separate occasions at least 12 weeks apart.3 Among the acquired thrombophilias, APS is known to cause 4.1% of all the venous thromboses.4 The highest risks of thrombosis are associated with increased titers of LA and immunoglobulin G aCL antibodies5 or with a profile that includes positivity for all three antibodies mentioned previously.6

As noticed in this patient, abdominal pain is the most common symptom in patients with portomesenteric thrombosis. Other symptoms such as nausea, vomiting, abdominal distension, change in bowel habits, hematemesis, or melena due to bowel infarction can also be present. The thrombotic events can involve arteries, veins, or smaller vessels of any organ and must be confirmed with radiological or histopathological evidence. The most common venous thrombotic event in APS is deep vein thrombosis, especially of the calf. In a study including 1,000 patients done by Cervera et al, deep vein thrombosis was found in up to 30% of the patients.7 Thrombosis of both SMV and PV together is relatively rare with only sporadic reports in the world literature. Recurrent thrombotic events are also known to occur commonly in APS. Generally, an initial arterial event is followed by another arterial event and in a similar way, a venous thrombosis by another venous event. Studies have shown up to 93% recurrence in cases of arterial thrombosis and 76% recurrence in cases of venous thrombosis.8 The risk of recurrence and death is related to presence of higher titers of aCL antibodies 6 months after the initial venous thrombotic event.9

Though CECT can support the diagnosis of portomesenteric thrombosis and document resolution of the same after treatment, EUS provides finer details such as blood flow in real time in addition to thrombosis and obviates exposure to repeated radiation during follow-up. Also, EUS has additional advantage of avoiding contrast in those with renal failure and contrast hypersensitivity.

The mainstay of treating nonobstetric cases of APS is heparin and warfarin.10 Aspirin is used as an add-on for resistant cases. There is no role for any primary prophylaxis for patients with a positive antiphospholipid antibody titer.
Lifelong therapy with warfarin is recommended for patients with significant thrombosis with established APS. The target INR to be maintained is 2.0 to 3.0 in those with venous thrombosis, 3.0 in those with arterial thrombosis, and 3.0 to 4.0 in those with recurrent thrombotic events.

The case we report is a rare case of portomesenteric thrombosis due to primary APS, treated successfully with low-molecular-weight heparin and warfarin initially and followed by warfarin alone for 6 months, which resulted in the resolution of the thrombus in the PV and recanalization that has been documented by both EUS and CECT imaging, before and after the treatment. Subsequently, the patient is on anticoagulant therapy and is presently asymptomatic.

Early identification and treatment of portomesenteric thrombosis is crucial to prevent bowel ischemia. Lifelong anticoagulation with vitamin K antagonists should be considered in those patients with major thrombosis and established APS. Monitoring of the INR is crucial not only to prevent further progression of thrombosis but also to stop iatrogenic bleeding, because of the narrow therapeutic index of vitamin K antagonists.

This case is reported to highlight the fact that in all cases of portomesenteric thrombosis without underlying cirrhosis or hepatocellular carcinoma, APS needs to be ruled out. EUS provides all the required details of portomesenteric thrombosis and obviates the need for repeat CT abdomen and its attendant risk of radiation and allergic reaction to contrast during follow-up.

Conflict of Interest
None declared

References

Fig. 4 Endoscopic ultrasonography showing resolution of thrombosis with recanalization as demonstrated by Doppler.