

## Case Report

# Integrative Gastroenterology and Hepatology

## Abdominal Pain-An Unusual Case of Anti-Phospholipid Syndrome

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### Abstract

Antiphospholipid Syndrome (APS), an autoimmune disease associated with hypercoagulability, commonly presents as arterial and/or venous thrombosis, recurrent spontaneous abortions, and moderate thrombocytopenia. It can manifest as a standalone syndrome or a manifestation of a primary systemic disease. The incidence and prevalence of APS without an acquired cause is not well known, although some estimates show around 5 new cases per 100,000 persons per year. Antiphospholipid antibodies (APLA) include anti-cardiolipin, Lupus anticoagulant (LA), and anti-beta-2 glycoproteins which are responsible for the underlying pathophysiology. APS is known to be associated with SLE, connective tissue disorders, various autoimmune diseases, malignancies, HIV, and drugs. Anti-cardiolipin antibody causing thrombosis represents a spectrum of APS which is usually associated with an acquired condition and rarely presents as a primary syndrome. We present the case of an African American female, aged 30, with an atypical presentation of a thrombotic episode and the presence of anti-cardiolipin antibodies without any associated secondary cause. Our case stands out because of the primary nature of APS and the atypical presentation with abdominal signs, both of which are rare and constitute only 1.5% of cases of APS. Sharp clinical suspicion with prompt diagnosis can potentially prevent progression to a catastrophic event.

**Keywords:** Antiphospholipid syndrome, Antiphospholipid antibodies, Anti-cardiolipin antibody, Venous thrombosis

### Introduction

Antiphospholipid syndrome (APS) is an autoimmune disease that is associated with hypercoagulability, commonly presents as arterial and/or venous thrombosis, recurrent spontaneous abortions, and moderate thrombocytopenia [1]. APS can present as a primary condition or secondary to an underlying systemic disease process. Antiphospholipid antibodies (APLA) are responsible for the underlying pathophysiology. The APLA include anti-cardiolipin, lupus anticoagulant (LA), and anti-beta-2 glycoprotein-1. APS with anti-cardiolipin antibody variant can present in multiple ways, including

deep venous thrombosis, arterial thrombosis, premature coronary artery disease, cerebrovascular accident, retinal disease, and it is five times more common than LA syndrome [2]. We present a case of a 30-year-old African American female with an atypical presentation of a thrombotic episode and the presence of anticardiolipin antibodies without any secondary cause.

### Case Report

A 30-year-old African American female presented with a 3-week history of dull, achy epigastric pain and vomiting. Initially, she presented to the emergency

department (ED), was diagnosed with gastritis and discharged home. She presented back to the ED one week later with worsening abdominal pain. She denied fever, chills, abdominal bloating, and diarrhea, blood in stool, photosensitivity, oral ulcers, rash, or joint pains. Her past medical history is significant for a pulmonary embolism 9 months prior to the presentation, which was attributed to Oral Contraceptive (OCP) use and smoking. She was treated with an oral anticoagulant for 6 months. She had since stopped OCPs and smoking. It is worth noting that she had two elective abortions in the past without any early or late spontaneous miscarriages.

Pertinent examination findings include tenderness upon deep palpation of the epigastric, umbilical, and left upper quadrant regions without any rigidity, rebound tenderness, or guarding. Lab work showed the following: WBC 7.8 K/mcl, Hb 13.1 g/dl, Platelets 364 K/mcl, PT 11.7 seconds, INR 1, and a PTT of 29 sec. Liver function test; lipase, BUN, and creatinine were within normal ranges. Urine analysis showed no proteinuria. HIV screening and RPR test were negative. ANA was negative. An acute hepatitis panel was negative. CT angiogram of the abdomen revealed a small localized non-occluding venous thrombus within the superior mesenteric vein with patent portal and splenic veins. (Figure 1a, b). Nonspecific inflammation in the central abdomen, possibly mesenteric diverticulitis, was also seen without any abscess, bowel obstruction, infarction, splenomegaly, or liver abnormality. Color Doppler of the abdomen showed non-occluding thrombus of the left portal vein with normal right portal vein and hepatic veins (Figure 2). The workup for mesenteric vein thrombosis also included ruling out conditions such as myeloproliferative disorders and paroxysmal nocturnal haemoglobinuria. No IgM spikes were noted on serum electrophoresis and the possibility of lymphoplasmacytic disorder (i.e. Waldenstrom's) was ruled out. She was placed on heparin infusion and was admitted to the medical floor. Hypercoagulable workup, including Protein C, Protein S, and Antithrombin-III results, was unremarkable. Anti-cardiolipin antibody IgM was elevated at 48 MPL (Reference Values 0-12 MPL: Negative, 13-19 MPL: Indeterminate, 20-80 MPL: Low to Moderately Positive, 81 MPL or above: High Positive). LA and anti-beta-2 glycoprotein-1 antibody were absent from the plasma. Factor V Leiden and prothrombin gene mutation were also ruled out. The patient was ultimately switched to a Novel Oral Anticoagulant (NOAC) for long-term anticoagulation. She improved clinically and was discharged home on day four of hospitalization.

Four weeks after being discharged, she had her initial follow up with the outpatient clinic. At the time, she was Doi: <http://dx.doi.org/ng> well and had no further complaints. Another follow up at 12 weeks revealed anticardiolipin IgM elevated to 57 MPL. Given the initial clinical presentation and the elevated IgM levels at two different occasions 12 weeks apart without any secondary cause, the diagnosis of APS was made as per revised SAPPORO classification criteria [3].



**Figure 1a, b:** CT angiogram of the abdomen showing a small localized non-occluding venous thrombus within the superior mesenteric vein with patent portal and splenic vein (green arrows).



**Figure 2:** Color Doppler of the abdomen showing non-occluding thrombus of the left portal vein with normal right portal vein and hepatic veins (green arrows).

## Discussion

APS can present as either a primary or secondary condition. APLA include anticardiolipin, LA, and anti-beta-2 glycoprotein. Anticardiolipin antibody causing thrombosis represents a spectrum of APS, which is usually an acquired condition and rarely presents as a primary syndrome. APS can be associated with SLE, connective tissue disorders, various autoimmune diseases, malignancies, HIV and drugs.

This case report emphasizes the primary nature of

the syndrome. The incidence and prevalence of APS without an acquired cause is not well known although some estimates indicate around 5 new cases per 100,000 persons per year and a prevalence of 40-50 cases per 100,000 populations per year respectively [4]. Our case stands out because of the primary nature of APS and the atypical presentation with abdominal signs both of which are rare, constituting only 1.5 % cases of APS [5].

To ascertain APS diagnosis, at least one clinical and one-laboratory criteria should be present according to the revised SAPPORO guidelines, requiring the presence of vascular thrombosis or pregnancy morbidity, with positive LA, anticardiolipin, or anti-beta-2 glycoprotein-1-antibody. For a positive diagnosis, lab test must be verified after at least 12 weeks.<sup>3</sup> Although these guidelines were developed for use in patient selection for research purposes, many clinicians use them to inform their decision of an APS diagnosis [3,6]. The less-than-perfect sensitivity and specificity of the criteria curtails their use for diagnostic purposes, although they are useful in diagnosing some cases, preventing over diagnosis, and helping clinicians report key disease features [7,8]. Hence, a better approach to diagnosing APS is patients who meet the revised SAPPORO criteria for definite APS with no surrogate diagnosis to explain the signs and symptoms. Primary APS can progress to Systemic Lupus Erythematosus or other autoimmune conditions rarely. Therefore, regular follow-up is warranted in patients with APS [9].

Treatment of APS with thrombosis is anticoagulation. During the first episode of venous thrombosis, a finite duration of treatment is recommended. The initial treatment can differ in a patient presenting with abdominal venous thrombosis with hemodynamic instability, bowel ischemia, or acute abdomen. In such cases, exploratory laparotomy with or without resection is the treatment of choice. Some data suggests aggressive therapy, such as plasma exchange, should be started in patients with catastrophic APS [10].

## Conclusion

APS without an associated systemic condition is a rare occurrence, as APS typically presents as a manifestation of some systemic disease. It remains to be seen whether APS without a secondary disorder has any association to a specific group, race, sex, and age. Our case report highlights the importance of adding APS to the differential of unexplained abdominal pain. High level of clinical suspicion leading to an early diagnosis can potentially

prevent the progression to a catastrophic event.

## Acknowledgement

### Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

### Informed consent

Verbal informed consent was obtained from the patient(s) for their anonymized information.

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### Conflict of interest

The authors report no conflict of interest.

### Authorship statement

All authors have contributed equally to the manuscript.

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