

Case Report

Primary antiphospholipid syndrome with celiac and splenic artery thrombosis

Arun Kumar Alagesan*, Kannan R., Vikranth V., Raghav J.

Department of General Medicine, Saveetha Medical College and Hospital, Tamil Nadu, India

Received: 17 February 2023

Accepted: 04 March 2023

*Correspondence:

Dr. Arun Kumar Alagesan,

E-mail: arunronin9@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Antiphospholipid syndrome (APS) is an autoantibody mediated acquired thrombophilia characterized by recurrent arterial or venous thrombosis and or pregnancy morbidity. We describe a case of 32-year-old female who had a medical history of APLA presented with complaints of abdominal pain for 20 days. On evaluation, CT abdomen contrast revealed celiac and splenic artery thrombosis. She was successfully treated with IV unfractionated heparin, warfarin and antiplatelets.

Keywords: APS, Celiac artery thrombosis, Splenic infarction

INTRODUCTION

Antiphospholipid antibodies are autoantibodies that are directed against phospholipid binding proteins. Antiphospholipid syndrome (APLS) is a multisystemic autoimmune disorder. The hallmark of APLS comprises the presence of persistent antiphospholipid antibodies (APLA) in the setting of arterial and venous thrombosis and or pregnancy loss. The deep veins of the lower extremities and the cerebral arterial circulation are the most commonly affected venous and arterial sites, respectively.¹ Thrombosis can also occur in more unusual locations such as the hepatic veins, visceral veins or cerebral venous circulation. Obstetrical morbidity in APS includes the unexplained death of one or more morphologically normal fetuses at or beyond the 10th week of gestation, the premature birth of one or more morphologically normal neonates before the 34th week of gestation because of either eclampsia or severe preeclampsia, and/or three or more unexplained, consecutive spontaneous abortions before the 10th week of gestation.²

The updated Sydney classification scheme also requires specific laboratory criteria: a lupus anticoagulant detected according to guidelines published by the international society on thrombosis and hemostasis (ISTH), anticardiolipin (aCL) antibodies (IgG or IgM) exceeding 40 IgG or IgM antiphospholipid units, or anti-β2GPI antibodies (IgG or IgM) at levels exceeding the 99th percentile, measured by enzyme-linked immunosorbent assay (ELISA).^{3,4}

Beta-2-glycoprotein I is a plasma glycoprotein comprised of 5 'sushi' domains. The first domain contains the binding site for most anti-β2GPI APLA while the fifth domain binds anionic phospholipid.⁵ Inhibition of natural anticoagulant activity, particularly that of the protein C system, was the first identified prothrombotic mechanism of APLA.⁶ APLA impair the activation of protein C, as well as the ability of activated protein C to inactivate factors V and VIII.⁷ These activities are mediated by antibodies to β2GPI and/or prothrombin, and phosphatidylethanolamine plays a critical role.⁸ APLA, particularly anti-β2GPI, activate vascular cells, including endothelial cells, monocytes, neutrophils, and platelets.

Endothelial cell activation transforms the normally anticoagulant endothelial surface to a procoagulant phenotype.⁹ Activated endothelial cells demonstrate increased expression of adhesion molecules (E-selectin, VCAM-1, ICAM-1), von Willebrand factor, tissue factor (TF), proinflammatory cytokines, decreased levels of endothelial cell-derived nitric oxide and release microparticles with pro-inflammatory and procoagulant properties.¹⁰ Complement activation by APLA leads to generation of the anaphylotoxin C5a, which recruits monocytes and neutrophils, activates endothelial cells and induces expression of tissue factor.¹¹

We report a case of a 32-year-old female diagnosed to have celiac artery and splenic artery thrombosis which developed as a complication of APS. Prompt and timely management with IV anticoagulants, oral warfarin and antiplatelets encouraged successful treatment.

CASE REPORT

A 32-year-old female presented with complaints of left side abdominal pain, nausea, vomiting and fever for past 20 days. She had a medical history of abortion and unexplained death of two morphologically normal fetuses beyond 10th week of gestation during her second, third and fourth pregnancy and diagnosed to have APS 1 year back before she presented to us. Following diagnosis, she was not on oral anticoagulants and antiplatelets. On examination, patient was conscious and oriented with the GCS of 15/15. Her pulse rate was 92 beats per minute, blood pressure was 110/70 mmHg, respiratory rate-20 cycles per minute, SpO₂-98% in room air. General examination was unremarkable. On systemic examination, tenderness was present over left hypochondrium.

Her blood reports revealed Hemoglobin- 12.6 gm/dl, Total leukocyte count-14270 cells/microlitre, platelet-4.37 lakh cells/microliter, CRP-25 mg/L, D-dimer-992 ng/ml, PT-11.5 seconds, APTT-29.1 seconds, INR-1, HbA1c-5.5. Renal function test, liver function test, serum electrolytes and urine routine were within normal limits. Her HIV I, II, HBsAg/Anti HCV antibody were negative. Lupus anticoagulant testing by dRRVT (diluted Russel's viper venom test) remained strongly positive, anticardiolipin antibodies IgM >40 antiphospholipid units and IgG >40 antiphospholipid units and antithrombin II glycoprotein I antibody titres of IgM and IgG were greater than 99th percentile. ANA profile, serum homocysteine, protein C and protein S levels were found to be in normal limits. Her blood and urine culture revealed no growth. ECG revealed normal sinus rhythm. CT abdomen contrast revealed partial non enhancing filling defect noted involving celiac axis with non-opacification of complete course of splenic artery due to thrombus (Figure 1) and diffuse hypodense linear and wedge shaped non enhancing areas over the spleen suggesting splenic infarction (Figure 2).

She was started on injection enoxaparin 60mg subcutaneous BD, oral warfarin 5 mg at 6 pm daily, tablet

ecospirin 75 mg OD. During the course of stay in hospital she improved symptomatically and subcutaneous enoxaparin 60 mg BD was given over a period of 4 days following which IV heparin 5000U 6th hourly was given over a period of 4 days along with overlap therapy of oral warfarin 5mg daily with serial monitoring of INR. On 10th hospital day patient was symptom free and was discharged with oral warfarin at dose of 5 mg per day and tablet ecospirin 75 mg OD. After one week with her target INR maintaining at 3.0, warfarin dose was reduced to 3 mg per day and she was doing significantly better on proper evaluation. She was followed up at the outpatient department once a month with oral warfarin being continued and target INR maintaining between 2 to 3.

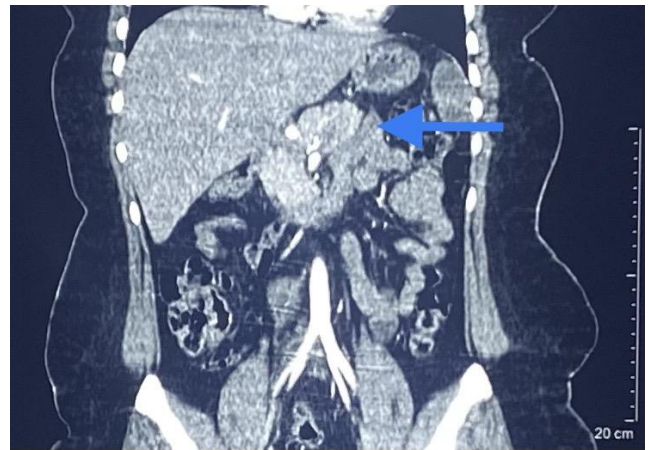


Figure 1: CT abdomen contrast revealing splenic artery thrombosis (blue arrow).

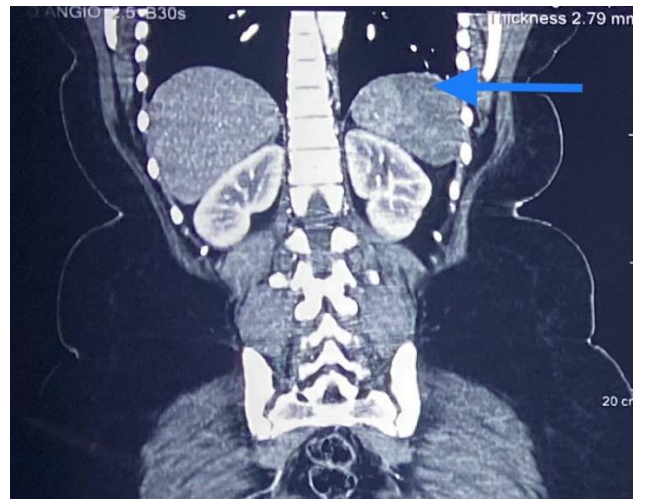


Figure 2: CT abdomen contrast revealing splenic infarction (blue arrow).

DISCUSSION

The celiac trunk is classically divided into three major branches: left gastric, common hepatic, and splenic artery. It supplies blood to the foregut, namely the distal esophagus, stomach, second part of the duodenum, liver,

pancreas, gallbladder, and spleen. The clinical presentations and the outcomes of celiac trunk thrombosis are variable. The spleen is more susceptible to ischemia and infarction, and dual blood supply and superior collateral flow ensure that the liver is resilient to the effects of thrombosis. Gastric ischemia is uncommon as the stomach has a rich blood supply from branches of the celiac trunk and the SMA collaterals.¹² Splenic infarction is common with celiac trunk thrombosis because the splenic artery is the main blood supply of the spleen.

Alharbi published a similar case report where a 52-year-old female presented with acute abdomen diagnosed to have celiac artery thrombosis with splenic infarction and was successfully treated with low molecular weight heparin and low dose aspirin.¹³

Chai et al published a case report where a 56 year old female presented with a 15 month history of diffuse abdominal pain and on evaluation found to have celiac and mesenteric arterial thromboses leading to splenic infarction and was successfully treated with heparin, warfarin and low dose aspirin.¹⁴

Genc published a case report where a 53-year-old male patient presented with complaints of left upper abdominal pain for 4 days and diagnosed to have splenic artery thrombosis without any predisposing factors.¹⁵

Clinicians should also be aware of the abdominal manifestations of APS which include Budd Chiari syndrome, hepatic veno occlusive disease, occlusion of small hepatic veins, nodular regenerative hyperplasia, hepatic infarction, cirrhosis, portal hypertension, autoimmune hepatitis, biliary cirrhosis, acute intestinal infarction, intestinal angina, intestinal bleeding, splenic infarction, autospplenectomy or functional asplenia and acute pancreatitis.¹⁶

An uncomplicated spleen infarction can be safely managed with medical treatment, but early surgery is needed when complications of the infarct, including abscess and rupture, have occurred. Our case was diagnosed early, and the anticoagulant treatment began at an early stage, so the outcome was successful.

CONCLUSION

Celiac trunk thrombosis has a variety of clinical manifestations and outcomes. A high degree of suspicion is required for the diagnosis. In background clinicians should also be aware of the other abdominal manifestations of APS. Early blood flow establishment through anticoagulation, surgical, or endovascular intervention is essential for a successful course of treatment.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

1. Cervera R, Piette JC, Font J, Khamashta MA, Shoenfeld Y, Camps MT et al. Antiphospholipid syndrome: clinical and immunologic manifestations and patterns of disease expression in a cohort of 1,000 patients. *Arthritis Rheum.* 2002;46:1019-27.
2. Miyakis S, Lockshin MD, Atsumi T, Branch DW, Brey RL, Cervera R et al. International consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). *J Thromb Haemost.* 2006;4:295-306.
3. Brandt JT, Triplett DA, Alving B, Scharrer I. Criteria for the diagnosis of lupus anticoagulants: an update. On behalf of the Subcommittee on Lupus Anticoagulant/Antiphospholipid Antibody of the Scientific and Standardisation Committee of the ISTH. *Thromb Haemost.* 1995;74:1185-90.
4. Pengo V, Tripodi A, Reber G, Rand JH, Ortel TL, Galli M et al. Update of the guidelines for lupus anticoagulant detection. Subcommittee on Lupus Anticoagulant/Antiphospholipid Antibody of the Scientific and Standardisation Committee of the International Society on Thrombosis and Haemostasis. *J Thromb Haemost.* 2009;7:1737-40.
5. Agar C, van Os GM, Morgelin M, Sprenger RR, Marquart JA, Urbanus RT et al. Beta2- glycoprotein I can exist in 2 conformations: implications for our understanding of the antiphospholipid syndrome. *Blood.* 2010;116:1336-43.
6. Urbanus RT, de Laat B. Antiphospholipid antibodies and the protein C pathway. *Lupus.* 2010;19:394-9.
7. Marciniak E, Romond EH. Impaired catalytic function of activated protein C: a new in vitro manifestation of lupus anticoagulant. *Blood.* 1989;74:2426-32.
8. Smirnov MD, Triplett DT, Comp PC, Esmon NL, Esmon CT. On the role of phosphatidylethanolamine in the inhibition of activated protein C activity by antiphospholipid antibodies. *J Clin Invest.* 1995;95:309-16.
9. Williams FM, Parmar K, Hughes GR, Hunt BJ. Systemic endothelial cell markers in primary antiphospholipid syndrome. *Thromb Haemost.* 2000; 84:742-6.
10. Betapudi V, Lominadze G, Hsi L, Willard B, Wu M, McCrae KR. Anti-beta2GPI antibodies stimulate endothelial cell microparticle release via a non-muscle myosin II motor protein dependent pathway. *Blood.* 2013;122:3808-17.
11. Redecha P, Tilley R, Tencati M, Salmon JE, Kirchhofer D, Mackman N et al. Tissue factor: a link between C5a and neutrophil activation in antiphospholipid antibody induced fetal injury. *Blood.* 2007;110:2423-31.
12. Kelekis NL, Athanassiou E, Loggitsi D, Moisidou R, Tzovaras G, Fezoulidis I. Acute occlusion of the celiac axis and its branches with perforation of gastric fundus and splenic infarction, findings on spiral

- computed tomography: a case report. *Cases J*. 2010;3:82.10.1186/1757-1626-3-82.
13. Alharbi M, Benitez CY, Alharbi AM. Celiac Trunk Thrombosis a Rare Cause of Acute Abdomen. *Case Presentation. Cureus*. 2022;14(9):e29727.
 14. Choi BG, Jeon HS, Lee SO, Yoo WH, Lee ST, Ahn DS. Primary antiphospholipid syndrome presenting with abdominal angina and splenic infarction. *Rheumatol Int*. 2002;22(3):119-21.
 15. Genc V, Cetinkaya OA, Kayilioglu I, Karaca AS, Cipe G, Unal AE. Splenic infarction as a complication of celiac artery thromboembolism: an unusual cause of abdominal pain. *J Korean Surg Soc*. 2011;81(5):360-2.
 16. Uthman I, Khamashta M. The abdominal manifestations of the antiphospholipid syndrome. *Rheumatology (Oxford)*. 2007;46(11):1641-7.

Cite this article as: Alagesan AK, Kannan R, Vikranth V, Raghav J. Primary antiphospholipid syndrome with celiac and splenic artery thrombosis. *Int J Adv Med* 2023;10:314-7.