International Journal of Biomedicine 15(3) (2025) 598-600
http://dx.doi.org/10.21103/Article15(3)_CR3

CASE REPORT

INTERNATIONAL JOURNAL OF BIOMEDICINE

Antiphospholipid Syndrome Associated with Systemic Lupus Erythematosus: A Case Report

Ina Toska[1*], Ervin Rapushi[2], Rexhep Shkurti[1], Anila Mitre[3]

[1]*Department of Biotechnology, Faculty of Natural Sciences, University of Tirana, Tirana, Albania*
[2]*University of Medicine, Tirana, Albania*
[3]*Department of Biology, Faculty of Natural Sciences, University of Tirana, Tirana, Albania*

Abstract

Antiphospholipid syndrome (APS) is an autoimmune disorder characterized by venous or arterial thrombosis and/or pregnancy outcomes in the presence of persistent antiphospholipid antibodies like lupus anticoagulant (LAC), anti-cardiolipin (aCL), and anti-β2glucoprotein. Antiphospholipid syndrome may occur as a primary disorder or in association with other autoimmune diseases, especially with systemic lupus erythematosus (SLE). This case reports a 28-year-old woman diagnosed with SLE and APS after her first thrombotic event, a deep vein thrombosis in the right leg. All SLE patients must be screened for antiphospholipid antibodies, even when thrombotic events have not occurred, to determine the antiphospholipid antibody profile, which is important for future thrombotic risk events. (**International Journal of Biomedicine. 2025;15(3):598-600.**)

Keywords: antiphospholipid syndrome • antiphospholipid antibodies • systemic lupus erythematosus

For citation: Toska I, Rapushi E, Shkurti R, Mitre A. Antiphospholipid Syndrome Associated with Systemic Lupus Erythematosus: A Case Report. International Journal of Biomedicine. 2025;15(3):598-600. doi:10.21103/Article15(3)_CR3

Abbreviations

aCL, anti-cardiolipin; **APS**, antiphospholipid syndrome; **ANA**, antinuclear antibody; **CT**, computed tomography; **DVT,** deep vein thrombosis; **ENA**, extractable nuclear antigen; **IVC**, inferior vein cava; **INR**, international normalized ratio; **LAC**, lupus anticoagulant; **PE**, pulmonary embolism; **SLE**, systemic lupus erythematosus.

Introduction

Antiphospholipid syndrome (APS) is an autoimmune disorder characterized by venous or arterial thrombosis and/or pregnancy outcomes in the presence of persistent antiphospholipid antibodies like lupus anticoagulant (LAC), anti-cardiolipin (aCL), and anti-β2glucoprotein.[1] Classification criteria for APS have changed over the years. Sapporo criteria were first published in 1999 and updated in 2006 (Sydney criteria). Diagnosis for APS, based on revised Sapporo criteria, requires at least one clinical and one laboratory manifestation.[2] Clinical criteria comprise arterial/vascular thrombosis or pregnancy morbidity, while laboratory ones include the presence of medium or high titers of IgG and/or IgM anti-cardiolipin (aCL) antibodies, IgG/IgM anti-beta 2 glycoprotein I antibodies (anti-β2GPI), and/or LAC positive on two or more occasions at least 12 weeks apart.[3]

Antiphospholipid syndrome may occur as a primary disorder, but when it is associated with other autoimmune diseases, most commonly with systemic lupus erythematosus (SLE), it is defined as a secondary APS.[4,5]

Systemic lupus erythematosus is an autoimmune disease that affects connective tissues, leading to chronic inflammatory illness of skin, joints, kidneys, lymph nodes, and the lining layers of the blood vessels with increased risk of arterial and venous thrombotic events.[6] When SLE was associated with antiphospholipid antibodies, an increased number of thrombotic events was observed. The presence of antiphospholipid antibodies has been described in around 50% of SLE patients, while around 20% of APS patients have SLE.[7]

***Corresponding author:** Ina Toska, E-mail: ina.toska@yahoo.com*

This case reports a 28-year-old woman diagnosed with SLE and APS after her first thrombotic event, a deep vein thrombosis (DVT) in the right leg.

Case Presentation

A 28-year-old female patient experienced her first thrombotic event at the age of 14: DVT in the right leg, in the absence of varicose veins or other coagulation disorders, and did not report using oral contraceptives. She was hospitalized at Shkodra hospital and was stabilized after treatment with nadroparin calcium. After leaving the hospital, she was treated with acenocoumarol with dosing adjusted according to the INR level.

One year later (January 2012), this patient underwent several examinations due to her poor health condition. CT was also performed, resulting in normal findings. Six months later, this young lady was hospitalized at the University Hospital Center "Mother Tereza," (Tirana, Albania) for an accurate diagnosis. She reported experiencing several symptoms like fatigue, physical weakness, pain in the hands and feet, stains in the skin of the abdomen, hair loss, numbness of hands and feet, and even mouth ulcers. Upon general examination, the patient was alert and oriented. The skin of the face had a butterfly-shaped rash over the cheeks and nose. Respiratory effort appeared slightly labored.

An abdominal ultrasound was performed and resulted in normal findings, while bone and joint pathology was revealed by hand x-ray. A series of laboratory examinations, including cell blood count, biochemical balance, coagulation, and immunological profiles, were performed (Table 1).

Comparison of laboratory results from January to June revealed transient thrombocytopenia, reduced hemoglobin levels, and decreased complement C3 and C4 levels. Antinuclear antibodies (ANA), extractable nuclear antigen (ENA) screening, and anti-cardiolipin IgM/IgG were all positive.

Based on clinical and laboratory evaluation, the diagnosis was SLE and APS with anti-cardiolipin IgM/IgG positive and thrombotic events.

The medical treatment started immediately and included acenocoumarol, according to the level of INR, plaquenil (400 mg/d), medrol (8 mg/d), and vitamin D (2000 IU/d). The clinical situation appeared to be stabilized upon the receipt of the medical protocol.

In 2014, this patient interrupted the treatment with acenocoumarol, and a clinical complication occurred. She was admitted to the hospital with difficulty breathing, and according to laboratory examinations (Table 1) and CT, she was diagnosed with bilateral pulmonary thromboembolism. The treatment protocol remained the same, on the condition that therapy with acenocoumarol, not be interrupted.

In recent years, this patient has reported increased leg pain and varicose veins. The last CT showed blockage of the inferior vein cava (IVC), suggesting a post-thrombotic event occurring there and suggesting the necessity of surgical intervention. The latest laboratory examination data are also presented in Table 1.

Table 1.

Summary of laboratory examinations.

Laboratory tests	01.2012	06.2012	2014	2025	Reference values
RBC	4.73	4.36	5.1	5.55	4-5×10^6/µL
WBC	4.9	4.85	12	5.71	5-10×10^3/uL
Hb	12.3	10.7	8.8	15.4	12-16 g/dL
PLT	17	292	498	263	140-400×10^3/uL
ALT	92.8	23.6	21	22.4	< 35 U/L
AST	76.8	23.9	42	27.4	<31 U/L
TPU 24H		271.5	Negative		< 150 mg/24h
Ca	9.96		8.8		9.2-11 md/dL
CRP		5.1	14	1.05	<5 mg/L
CK		35	107		<170 U/L
LDH	301	205	289		< 480 U/L
RF	2.7	3.9	21	7.1	<14 IU/mL
FIB	439.1		397		200-400 mg/dL
APTT	79.7		47.3	36.9	25-35 sec
D-DI	1.8		1.66		< 0.5 ug/mL
C3		73	134	100	90-180 mg/dL
C4		7	11.7	14	10-40 mg/dL
LAC			107.1	94.5	31.4-43.4 sec
ANA	Positive	Positive	Positive	1:160	<1:80 Negative
DsDNA			Positive		Negative
ENA SCREEN		Positive	Positive		< 20 Negative
aCL IgM	Positive		Positive	Negative	<12 U/mL
aCL IgG	Positive			Positive	>12 U/mL
anti-ß2GPI IgG				Positive	>20 RU/ml

RBC, red blood cells; WBC, white blood cells; Hb, hemoglobin; PLT, platelets; ALT, alanine transaminase; AST, aspartate transaminase; TPU 24H: proteinuria; Ca, calcium; CRP, C-reactive protein; CK, creatin kinase; LDH, lactate dehydrogenase; RF, rheumatoid factor; FIB, fibrinogen; APTT, activated partial thromboplastin time; D-DI, d-Dimer; C3, C3 complement; C4, C4 complement; LAC, lupus anticoagulant; ANA, antinuclear antibodies; DsDNA, anti-double stranded DNA antibodies; ENA Screen, extractable antigen test; aCL IgM, anti-cardiolipin IgM; aCL IgG, anti-cardiolipin IgG; anti -ß2GPI, antiß2-glycoprotein.

Discussion

Antiphospholipid syndrome and systemic lupus erythematosus are two diseases related to each other due to the similarities that they have in terms of thrombotic events and clinical manifestations, including the presence of antiphospholipid antibodies. When SLE is associated with APS, it significantly increases the risk of thrombotic events in comparison with the general population. In SLE patients, approximately 50% are aPL positive, compared to those with SLE alone.[8]

According to recent data and meta-analysis of venous thromboembolism in SLE, patients with lupus erythematosus have a 4.38-fold higher relative risk of venous thromboembolism than the general population. When SLE coexists with APS, the absolute venous thromboembolism

risk increases to 63%. For DVT and PE, the risk rises from 1% to ~26% and 22%, respectively, when SLE is associated with APS.[9]

All laboratory findings in this patient, including positive ANA, ENA screen, anti-cardiolipin IgM/IgG, and proteinuria, confirmed the diagnosis of SLE with APS according to their respective classification protocols. SLE patients should be screened for antiphospholipid antibodies to determine their aPL profile. It is reported that in new SLE patients, double or triple aPL-positivity increases the risk for future vascular events.[10]

This case is an example of the coexistence of SLE and APS, which highlights the increased probability of thrombotic events. A young girl experienced DVT in the right leg at the age of 14, followed by bilateral pulmonary thromboembolism and, recently, an obstruction of the IVC, suggesting a post-thrombotic event occurring there. Although it is a very rare complication, particularly when the anticoagulation therapy is insufficient without immunosuppression, some cases of IVC have been reported in the literature. It is important to emphasize that our patient was not treated with immunosuppressants, a fact that could be a possible cause of the current situation.

Currently, in a review of several publications regarding immunosuppression in APS-positive patients with recurrent thrombotic events, anticoagulant therapy, hydroxychloroquine, and rituximab are recommended to reduce the titer of circulating antiphospholipid antibodies.

One of the cases reports a 14-year-old patient with active SLE, Evans syndrome, and secondary APS with acute abdominal pain, resulting in mesenteric vasculitis and thrombosis of the IVC.[11] Initial medical therapy with fraxiparin, followed by acenocumarol according to INR levels, showed that it was effective until its discontinuation, which led to PE, emphasizing the importance of continuing therapy in patients with SLE/APS with prior thrombotic events. Immunosuppressive therapy is also recommended, especially when thromboses persist despite anticoagulation therapy.

Thus, patients with SLE and APS have an increased risk for thrombotic events. All SLE patients must be screened for antiphospholipid antibodies, even when thrombotic events have not occurred, to determine the antiphospholipid antibody profile, which is important for future thrombotic risk events.

Acknowledgments

The authors are grateful to the patient for kindly allowing us to publish her case.

Competing Interests

The authors declare that they have no competing interests.

References

1. Musiał J. New classification criteria for antiphospholipid syndrome —2023. Journal of Transfusion Medicine [Internet]. 2023 [cited 2023 Nov 24];16(3):103–9. Available from: https://journals.viamedica.pl/journal_of_transfusion_medicine/article/view/97795/74984

2. Miyakis S, Lockshin MD, Atsumi T, Branch DW, Brey RL, Cervera R, Derksen RH, DE Groot PG, Koike T, Meroni PL, Reber G, Shoenfeld Y, Tincani A, Vlachoyiannopoulos PG, Krilis SA. International consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). J Thromb Haemost. 2006 Feb;4(2):295-306. doi: 10.1111/j.1538-7836.2006.01753.x. PMID: 16420554.

3. Dabit JY, Valenzuela-Almada MO, Vallejo-Ramos S, Duarte-García A. Epidemiology of Antiphospholipid Syndrome in the General Population. Curr Rheumatol Rep. 2022 Jan 5;23(12):85. doi: 10.1007/s11926-021-01038-2. PMID: 34985614; PMCID: PMC8727975.

4. Bahar Keleşoğlu Dinçer A, Erkan D. The ABCs of antiphospholipid syndrome. Archives of rheumatology [Internet]. 2023 May;38(2):163–73. Available from: https://pubmed.ncbi.nlm.nih.gov/37680521/

5. Branch DW. What's new in obstetric antiphospholipid syndrome. Hematology Am Soc Hematol Educ Program. 2019 Dec 6;2019(1):421-425. doi: 10.1182/hematology.2019000043. PMID: 31808896; PMCID: PMC6913435.

6. Ibrahim AAG, Shadi HWE, Elamin AAY, Draz HE. Retrospective cohort study of thromboembolic events in systemic lupus erythematosus with or without secondary antiphospholipid syndrome and their correlation to lupus activity and dyslipidemia. Egyptian Rheumatology and Rehabilitation. 2023 Feb 16;50(1).

7. Bazzan M, Vaccarino A, Marletto F. Systemic lupus erythematosus and thrombosis. Thromb J. 2015 Apr 23;13:16. doi: 10.1186/s12959-015-0043-3. PMID: 25908929; PMCID: PMC4407320.

8. Nakashima M, Kobayashi M. Preceding Antiphospholipid Syndrome before the Onset of Systemic Lupus Erythematosus Presenting with Iliocaval Deep Vein Thrombosis: A Case Report and Literature Review. Annals of Vascular Diseases [Internet]. 2024 [cited 2025 Jul 13];17(4):409–12. Available from: https://pmc.ncbi.nlm.nih.gov/articles/PMC11669031/

9. Bello N, Meyers KJ, Workman J, Marcano Belisario J, Cervera R. Systematic Literature Review and Meta-analysis of Venous Thromboembolism Events in Systemic Lupus Erythematosus. Rheumatol Ther. 2023 Feb;10(1):7-34. doi: 10.1007/s40744-022-00513-1. Epub 2022 Dec 6. PMID: 36471199; PMCID: PMC9931974.

10. Pericleous C, D'Souza A, McDonnell T, Ripoll VM, Leach O, Isenberg D, Giles I, Rahman A. Antiphospholipid antibody levels in early systemic lupus erythematosus: are they associated with subsequent mortality and vascular events? Rheumatology (Oxford). 2020 Jan 1;59(1):146-152. doi: 10.1093/rheumatology/kez239. PMID: 31257438; PMCID: PMC6909892.

11. Wang G, Li B, Luo Y, Li J, Wen Z. Mesenteric vasculitis with inferior vena cava thrombosis associated with systemic lupus erythematosus. Arch Med Sci. 2023 Feb 17;19(2):544-545. doi: 10.5114/aoms/158532. PMID: 37034506; PMCID: PMC10074299.