

Systemic Lupus Erythematosus and Secondary Antiphospholipid Syndrome Diagnosed After Pulmonary Thromboembolism in a Male Patient

Pulmoner Tromboemboli Sonrası Sistemik Lupus Eritematozus ve Sekonder Antifosfolipid Sendromu Tanıları Konulan Bir Erkek Olgu

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ABSTRACT Systemic lupus erythematosus (SLE) is an autoimmune, chronic, and multisystemic disease that can cause variable clinical manifestations. Antiphospholipid syndrome (APS) is a systemic autoimmune disease which occurs secondary to antiphospholipid antibodies (aPL) and results in vascular thromboses and/or pregnancy morbidity. Both diseases are more common in women. In this article, a male patient diagnosed with SLE and secondary APS during the etiologic investigations in terms of pulmonary thromboembolism will be discussed.

Keywords: Antiphospholipid Syndrome; pulmonary thromboembolism;
systemic lupus erythematosus

ÖZET Sistemik lupus eritematozus (SLE), etiyolojisi tam belli olmayan, değişken klinik tablolara neden olabilen, otoimmün, kronik ve multisistemik bir hastalıktır. Antifosfolipid sendromu (AFS), tekrarlayan vasküler tromboz ve/veya gebelik kayıplarıyla seyreden ve antifosfolipid antikorlara sekonder olarak ortaya çıkan sistemik, otoimmün kökenli bir hastalıktır. Her iki patoloji de kadınlarda daha sık görülmektedir. Bu yazıda pulmoner tromboemboli açısından yapılan etiyojik araştırmalar sırasında SLE ve sekonder AFS tanısı konulan bir erkek olgu tartışılacaktır.

Anahtar Kelimeler: Antifosfolipid sendromu; pulmoner tromboemboli;
sistemik lupus eritematozus

Systemic lupus erythematosus (SLE) is an autoimmune, chronic, and multisystemic disease that can cause variable clinical manifestations. The aetiology of SLE is not fully understood, but both genetic predisposition and environmental triggers are believed to be involved.¹ In a recent study, the overall age-adjusted incidence and prevalence per 100,000 persons were 5.5 and 72.8, respectively.² The incidence and prevalence of SLE is higher in females compared with males regardless of age or ethnic origin. In the studies, the sex ratio ranged from 2:1 to 15:1.¹

The antiphospholipid syndrome (APS) is a systemic autoimmune disorder characterized by recurrent thrombosis (artery, vein or small vessel) and/or obstetrical morbidity along with persistent antiphospholipid antibodies (aPL), including lupus anticoagulant (LA), anti-β2-glycoprotein I (anti-β2GPI) and/or anticardiolipin antibodies (aCL).^{3,4} The APS can be found in patients having neither clinical nor laboratory evidence of another definable condition (primary APS) or it may be associated with other diseases, mainly SLE (secondary APS).⁵ In the “Euro-Phospholipid” Project, a

female predominance (female:male ratio, 5:1) was confirmed in APS patients, but this was greater in patients with associated SLE (7:1) than in the primary APS (3.5:1).⁵

In this article, I present a male patient with SLE and secondary APS diagnosed after pulmonary thromboembolism. A written informed consent was obtained from the patient.

CASE REPORT

A 34-year-old male patient admitted to another hospital with a complaint of dyspnea and chest pain. The patient was diagnosed as pulmonary thromboembolism and the lower extremity deep venous thrombosis was detected as an embolism source and anticoagulant treatment was initiated to the patient. The patient was sent to our hospital for rheumatologic evaluation when the antinuclear antibody and anticardiolipin antibodies were detected positive.

Approximately four months after the diagnosis of pulmonary thromboembolism, he referred to our rheumatology outpatient clinic. He had no other chronic illness in his medical history and was not using any treatment other than warfarin. There was not any feature in rheumatological question-

ing except mild arthralgia and short-term morning stiffness in hand joints. Except from mild tenderness in the hand joints, there was no feature in his physical examination. The laboratory tests results of the patient are summarized (Table 1).

The patient was diagnosed with SLE according to the SLICC classification criteria because of ANA, anti-dsDNA and antiphospholipid antibodies positivity, hypocomplementemia and thrombocytopenia.⁶ In addition, antiphospholipid syndrome was diagnosed according to the Sapporo classification criteria due to deep vein thrombosis and pulmonary thromboembolism, aCL IgG/IgM positivity (4 months ago, aCL IgG and IgM were positive, too).⁷ Anticoagulant treatment was supplemented with 400 mg/day of hydroxychloroquine and 6 mg/day of methylprednisolone. After 1 month, the patient's joint complaints and morning stiffness had improved, platelet counts, ESR and CRP levels were normal. If the patient continues to be in good clinical condition, our treatment plan is to discontinue methylprednisolone, continue with hydroxychloroquine and warfarin treatment. The treatment was planned according to the EULAR recommendations for the management of SLE.⁸

TABLE 1: Laboratory tests results of the patient.

	Result	Normal range
White Blood Cell Count	10200/ μ L	3570-11000/ μ L
Hemoglobin Level	15.1 g/dL	11.4-16.4 g/dL
Platelet Count	92000/ μ L	150000-372000/ μ L
Creatinine	1.1 mg/dL	0.7-1.3 mg/dL
Urine analysis	Normal	Normal
Erythrocyte Sedimentation Rate	32 mm/h	0-15 mm/h
CRP	10 mg/L	0-8 mg/L
INR (International Normalized Ratio)	2.13	0.8-1.2
Rheumatoid Factor	Negative	0-20 IU/mL (Negative)
Antinuclear Antibody (ANA)*	1+ homogenous	Negative
Anti-ds DNA	Positive	Negative
Extractable Nuclear Antigen	Anti-SSA positive	Negative
Anticardiolipin antibody of IgG	120 GPL/mL	<12 GPL/mL
Anticardiolipin antibody of IgM	28.58 GPL/mL	<12 GPL/mL
C3c	0,828 g/L	0.79-1.52 g/L
C4	0.148 g/L	0.16-0.38 g/L

*ANA was detected by indirect immunofluorescence assay (IFA) method.

DISCUSSION

SLE is a chronic autoimmune disease with multi-system involvement that can cause life-threatening clinical conditions from time to time. SLE affects women more often than men and it is commonly known as the disease of women of child-bearing age.^{1,2} In a study, to assess the differences between SLE clinics in men and women; men were more affected than women in terms of disability, hypertension, thrombosis, renal and haematological involvement. Women were more likely to have malar rash, photosensitivity, oral ulcer, alopecia, Raynaud phenomena and arthralgia. It is also stated that end-organ damage and related deaths such as neuropsychiatric, renal, cardiovascular, peripheral vascular disease and myocardial infarction were more frequent in men. In general, differences between males and females were more numerous and striking in whites, especially with respect to lupus nephritis, abnormal serologies and thrombosis.^{9,10}

In a report presented by Cefle et al; in male patients with SLE, renal and pulmonary involvement, cardiomyopathy, serositis, discoid rash, and antiphospholipid syndrome were more frequent and the course of the disease was more severe.¹¹

It is mentioned that in another study; male patients with SLE present more cardiovascular comorbidities and also more serositis, adenopathies, splenomegaly, renal involvement, convulsion, thrombosis and lupus anticoagulant positivity than women.¹²

The antiphospholipid syndrome is a systemic autoimmune disease which occurs secondary to antiphospholipid antibodies and results in vascular thromboses and/or pregnancy morbidity. Deep vein thrombosis is the most common venous and stroke is the most common arterial involvement. Although the exact rate is unknown, it is stated that; APS is seen more frequently in women. However, it is also emphasized that this may be related to pregnancy losses and the more frequent occurrence of SLE in women.^{3,5} In one study; lupus anti-

coagulant levels were found to be statistically higher in males than females. In the same study, anticardiolipin and anti- β 2-glycoprotein I antibodies levels were found to be higher, although not reaching statistical significance, and thrombotic events were also seen more frequently in male patients.¹⁰ In another study with a high number of patients; deep vein thrombosis and pulmonary embolism were found to be significantly higher in patients with SLE than in the general population.¹³

It is an important condition that presented case did not have any symptoms or signs that need to be investigated in terms of SLE and/or APS until he was diagnosed with pulmonary thromboembolism. Because, these diseases can rarely start with life-threatening clinical conditions without any other symptoms. If we do not consider these diseases among our preliminary diagnoses, in patients with appropriate clinical and laboratory findings, the disease may progress and may cause the prognosis to worsen with multisystemic involvements.

In conclusion; although SLE and primary/secondary APS are more prevalent in women, when the etiology of thrombosis is being investigated, these diseases should also be considered in male cases. Research in this direction is very important and vital.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

This study is entirely author's own work and no other author contribution.

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