CASE REPORT OF A SYSTEMIC LUPUS ERYTHEMATOSUS PATIENT WITH CARDIAC, PULMONARY, RENAL, HEMATOLOGIC INVOLVEMENT AND ANTIPHOSPHOLIPID SYNDROME

(Kardiyak, Pulmoner, Renal, Hematolojik Tutulumu ve Antifosfolipid Sendromu Olan Sistemik Lupus Eritematosuslu Bir Olgu Sunumu)

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Summary

Systemic Lupus Erythematosus (SLE), is a multisystemic disease with an unknown etiology. It is especially frequent in females. SLE has a vasculitic feature and may effect many systems of the body. Clinical findings as well as laboratory tests are important for diagnosis. Auto-antibodies are present in the course of systemic lupus [(2)]. Antiphospholipid Syndrome (APS), is characterized by recurrent thromboembolisms and existence of auto-antibodies. APS resembles to SLE with some clinical properties and may be together with SLE as well. Main goal of the treatment is immune supression. Corticosteroids and immuno supressive drugs such as azothiopyrine, cyclophosphamide and metotrexate are used for this purpose.

Multisystemic involvement is found in progressive SLE cases. There were cardiac, pulmonary, renal, hematologic involvements and anti phospholipid syndrome in our case when the diagnosis was made. This case is reported to emphasize the traits of SLE and APS.

Key words: Systemic lupus erythematosus, antiphospholipid syndrome

Özet

Sistemik Lupus Eritematosus (SLE), etiyolojisi tam olarak ortaya konulamamış multisistemik bir hastalıktır. Özellikle kadınlarda sık görülür. Hastalık vaskülitik özellik gösterir ve vücuttaki birçok sistemi etkileyebilir. Tanıda klinik bulgular yanında laboratuar özellikleri de önemlidir. Dokulara karşı otoantikor gelişimi görülür. Antifosfolipid Sendromu (APS), tekrarlayan tromboemboliler ve otoantikor varlığı ile karakterizedir. Bazı özellikleri SLE'ye benzemektedir ve birlikte görülebilir. Tedavide amaç immün supresyondur. Bunun için sıklıkla kortikosteroidler ve azotioprin, siklofosfamid ve metotreksat gibi immünsupresif ilaçlar kullanılır.

Multisistemik tutulum, ilerlemiş SLE vakalarında görülür. Olgumuzda tanı konulduğunda kardiak, pulmoner, renal, hematolojik tutulum ve antifosfolipid sendromu mevcuttu. Bu sunum SLE ve AFS'nin özelliklerinin vurgulanması amacıyla yapıldı.

Anahtar kelimeler: *Sistemik lupus eritematosus, antifosfolipid sendromu*

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INTRODUCTION

Systemic lupus erythematosus (SLE), is an inflammatory connective tissue disorder that occurs principally in females. The pathogenesis of SLE consists of antibodies (anti-nuclear antibody, anti-dsDNA, anti-histone antibody) and immune complexes, resulting in damage to many tissues and organs [(1)]. Some types of SLE are idiopathic, drug induced and neonatal. American College of Rheumathology's (ACR) criteria are used to draw the diagnose for SLE (table 1). At least 4 criteria of 11 must be present for diagnosis. Overlap syndrome with other systemic disorders can also be seen, such as rheumatoid arthritis, Sjögren's disease, antiphospholipid syndrome [(2)]. Antiphospholipid syndrome (APS), is characterized by recurrent thromboembolisms and existence of auto-antibodies. Antiphospholipid syndrome resembles to systemic lupus with some clinical properties and laboratory findings (table 2-3). Clinical features are thrombocytopenia and recurrent spontaneous abortion with recurrent arterio-venos thromboembolisms. Important part of the therapy is heparin and warfarin administration.

Table 1. ACR criteria for SLE

CRITERION	FEATURE
1-malar rush	erythema on malar eminences
2-discoid rush	erythematous lesions with keratotic scalling
3-oral ulcer	oral and nasopharyngeal ulcers
4-photosensitivity	dermal sensitivity to sun light
5-arthritis	non-erosive oligo arthritis
6-serositis	pleuritis, pericarditis
7-renal	glomerulonephritis, proteinuria
8-hematologic	leucopenia, anemia, thrombocytopenia
9-neurologic	seizure, stroke, phsycosis
10-immunologic	anti ds DNA, anti cardiolipin antibodies
11-antinuclear ab.	existence of antinuclear antibody

Table 2. Clinical Criteria for APS

CLINICAL CRITERIA FOR APS
1-Recurrent arterio-venous thromboembolism
2-Thrombocytopenia
3-Recurrent spontaneous abortion

Table 3. Laboratory Criteria for APS

LABORATORY CRITERIA FOR APS
1-Lupus anticoagulants
2-Anti cardiolipin anticores

CASE

The case is a 27 years old female patient. Main complaints of the patient are fatigue, myalgia, arthralgia, oral ulcers, photosensitivity, allopesia which persists for 8 months. 7 years ago, the patient had 4 abortions with an unknown origin. In physical examination, systemic tension was 150/100 mm/Hg, radial pulse 84/min, body heat 37,9 C. There were not any problems in cardiac and abdominal examination. Basal pulmonary cracks were evident in pulmonary examination. Direct pulmonary graphy revealed basal infiltrations, and occlusion of the left costo-phrenic sinus. She had arthralgia on ankles and knees.There wasn't any evidence of deformation on joints with X-ray examination. Electrocardiographic (ECG) evaluation showed left bundle branch block. Results of hematologic and biochemical evaluation were as following; white cells: 8200 in mm^3, hematocrit 22,8%, hemoglobin 7,7g/dl, platelets 132.000 in mm^3, glucose: 91 mg/dl, urea: 58 mg/dl, creatinine: 1,7 mg/dl, lactat dehidrogenase (LDH): 2600 u/lt, total protein: 6,3 mg/dl, proteinuria 300 mg/lt. Anti nuclear antibody (ANA), anti ds DNA and anti cardiolipin antibody were positive. Direct coombs was positive. High LDH levels and anemia indicated hemolitic anemia. There was not any apparent pathology in other biochemical parameters. Gruber Widal test, Wright test, Anti HBs Ab, HBs Ag, Anti HCV titrations were all negative. Heamoculture and uroculture remained sterile. Urinary ultrasound graphy (USG) were performed because of the proteinuria. Urinary USG revealed grade II renal paranchymal disease. Renal biopsy was carried out and pathological examination was made. Diffuse proliferative glomerulonephritis was diagnosed in biopsy material. Thorax tomography was carried out in order to enlighten pulmonary examination findings. In thorax tomography there was basal pleural effusion, pulmonary paranchymal interstitial infiltrations and pleural thickness. Due to ECG findings and hypertension, transthoracic echocardiographic examination (TEE) was performed. According to TEE there was left ventricular hypertrophy, segmental hypochinesia of left ventricular wall and mild aortic valve insufficiency. Myocardial perfussion scintigraphy was performed and inferolateral and apical myocardial infarction were reported. There was not any obstruction of coronary arteries as evaluated with coronary angiography. Positivity of ANA and anti dsDNA, photosensitivity, oral ulcers, renal, cardiac, pulmonary and hematologic evidences have pointed out SLE, therefore the diagnosis was drawn. Pulse steroid treatment (500 mg/day) was administered for the first three days at the beginning of the therapy. The treatment continued with 1mg/kg/day prednisolon. The patient's complaints relieved with prednisolon treatment.

DISCUSSION

SLE is a multisystemic vasculitic disease and can effect many organs. Auto antibodies are responsible for the multisystemic involvement. Skin lesions and dermal sensitivity are the other clinical features of SLE as it was evident in this case. Systemic lupus must be kept in mind in female patients who presents to hospital with arthralgia, fever and skin lesions. In our case, there was involvement of different organs in addition to these traits. The reason of hemolytic anemia is the presence of auto antibodies against blood cells [(3)]. High LDH levels and positive Coombs test indicates hemolysis. Anemia causes fatigue, exercise intolerence. Thrombocytopenia causes purpuras and mucosal hemorrhagies. In our case, there was anemia and thrombocytopenia due to chronic hemolysis.

Serositis, one of the criteria of systemic lupus erythematosus may be present in patients with pleuritis, pericarditis, pleural and pericardial effusion. Lung injury may cause long term chronic lung damage and respiratory complications in advanced systemic lupus and the other connective tissue

disorders [4]. Main reason of serositis is immun mediated inflamation on serous membranes caused by auto antibodies. Pleuritis restricts inspiration and causes dyspnea. In our case serositis was demonstrated by radiological investigation. According to thorax tomography, basal pleural effusion, pulmonary paranchymal interstitial infiltrations and pleural thickness were evident.

Lupus nephritis is an important prognostic indicator and it can lead to nephrotic syndrome and renal failure in developed patients by autoantibodies [5]. Lupus nephritis develops in 50% of systemic lupus patients. Nephritis is divided into 5 pathologic classication groups by World Health Organisation (Table 4). One of the laboratory findings of lupus nephritis is proteinuria. In our patient, there was 300 mg/lt proteinuria and diffuse proliferative glomerulonephritis which was shown in biopsy material. Immune supresive therapy inhibits progression of nephritis.

Table 4. Pathologic Classification of Lupus Nephritis

Grade - 1	Normal biopsy material
Grade - 2	Mesangial glomerulonephritis
Grade - 3	Focal proliferative glomerulonephritis
Grade - 4	Diffuse proliferative glomerulonephritis
Grade - 5	Membranous glomerulonephritis

Ischemic heart disease can develop in SLE in which vasculitis is the main cause. In this case, infarction area in myocardium was shown in myocardial scintigraphy. Coronary angiography was normal. But, pericarditis was not demonstrated in echocardiography. There are laboratory and clinical relationships between lupus and antiphospholipid syndrome (APS). Main pathologic cause of APS is autoantibodies that damage tissues and lead to organ failure [6].

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