A Male Case with Subacute Cutaneous Lupus Erythematosus Developed Systemic Involvement and Bullous Skin Lesions

SİSTEMİK TUTULUM VE BÜLLÖZ DERİ LEZYONLARI İLE SEYREDEN BİR ERKEK SUBAKUT KUTANÖZ LUPUS ERİTEMATOZUS OLGUSU

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Summary

Subacute cutaneous lupus erythematosus (SCLE) is a distinct subset of systemic lupus erythematosus (SLE) clinically characterized by typical psoriasiform and/or annular lesions and by a mild or absent systemic involvement. Lesions are usually photosensitive and occur in predominantly sun-exposed areas. Major target organ involvement such as nephritis, central nervous system disease and systemic vasculitis develop in only 10 to 15 percent of patients with SCLE. SCLE affects mainly middle-aged women as like SLE. The development of a generalized bullous eruption is uncommon in patients with SLE and SCLE. Bullous lesions usually heal without scarring following treatment.

We report a male patient with SCLE developed systemic involvement and bullous lesions and conclude because of uncommon features of the case in this article.

Key Words: Subacute cutaneous lupus erythematosus, Bullous skin lesions, Male lupus

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Özet

Subakut kutanöz lupus eritematozus (SKLE) tipik olarak papüllo-squamöz ve/veya anüler deri lezyonları ile seyreden sistemik tutulumun olmadığı veya hafif olduğu bir sistemik lupus eritematozus alt grubudur. Lezyonlar genellikle fotosensitiftir ve genellikle güneş gören bölgelerde görülür. SKLE'lu hastaların sadece % 10-15'inde nefrit, santral sinir sistemi tutulumu ve sistemik vaskülit gibi major hedef organ tutulumu gelişir. Sistemik lupus eritematozusda olduğu gibi hastaların büyük çoğunluğunu kadınlar oluşturur. Büllöz deri lezyonları, sistemik lupus eritematozus veya subakut kutanöz lupus eritematozus seyri esnasında nadiren ortaya çıkar ve genellikle tedaviye iyi yanıt verir.

Bu makalede sistemik organ tutulumu ve büllöz deri lezyonları gelişen SCLE'lu bir erkek hasta sunuldu ve az görülen özellikleri nedeniyle tartışıldı.

Anahtar Kelimeler: Subakut kutanöz lupus eritematozus, Büllöz deri lezyonları, Erkek lupus

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Systemic lupus erythematosus (SLE) is a clinically heterogeneous autoimmune disease of unknown etiology. Skin involvement has been reported in 72% to 85% of patients with SLE and often is the first manifestation of the disease (1). Subacute cutaneous lupus erythematosus (SCLE) is recognized as a variant of SLE and clinically characterized by typical papulosquamous and/or annular skin lesions and by a mild or absent systemic involvement. Only 10% of all patients with SLE have SCLE and only 10% of patients with SCLE have major target organ involvement from SLE disease activity. Approximately half of patients with SCLE meet the American College of Rheumatology's revised criteria for the classification of SLE. However, manifestations of severe SLE such as nephritis, central nervous system disease and systemic vasculitis develop in only 10 to 15 percent of patients with SCLE (2,3).

SCLE is different from SLE by cutaneous changes, significantly less frequent kidney involvement, serositis and arthritis, and rare presence of double-stranded DNA (dsDNA), U_1RNP and Sm antibodies characteristic of SLE. Additionally, aggressive therapy in these patients is usually not required and the prognosis is better than in patients

with SLE. Therefore SCLE should be recognized as a separate subset. However, cases of overlapping SLE and SCLE may be seen (2).

Skin lesions of SCLE are diffuse and symmetric. Lesions are characteristically photosensitive and occur in predominantly sun-exposed areas (i.e., upper back, shoulders, extensor aspects of arms, V area of the neck, and less commonly the face). In except of the skin lesions, SCLE most frequently is accompanied by arthralgia, fever and weakness (3).

Bullous skin lesions are rarely seen in SLE. Histopathologic examination showed subepidermal blistering and immunoglobulins (IgG, IgA, IgM) and complement components (C1q, C3) deposited linearly at the basement membrane zone. Bullous lesions usually heal without scarring following treatment with dapsone or corticosteroids (4,5).

We report a male patient who rapidly developed severe multiorgan involvement and bullous lesions after admitted with typical SCLE lesions and conclude because of uncommon features of the patient in this article.

Case Report

A 27-year old man patient applied with complaints night sweating, lose weight, arthralgia and papullosquamous skin lesions in his hand, face and back to our hospital. He described that lesions have been appeared and disappeared approximately since 2 years in his history. Blood pressure was 110/70 mmHg, pulse rate was 65/ min and body temperature was 36,2°C. Physical examination revealed eritematous and squamous plaques in his forehead and malar area and aphtous lesions on mucous membrane in his oral cavity. Additionally, we found symmetric, hyperpigmented papullosqumous lesions showing local atrophy in his forearm, shoulder and back. The other physical examination findings were evaluated as normal.

According to these findings, we thought that the patient was SCLE and hospitalized him. There were anemia, high erythrocyte sedimentation rate (ESR), high titer of anti-nuclear antibody (ANA) and anti-dsDNA antibody in his laboratory examination (Table 1). Anti-Ro antibody test could not performed because of economical problems. Histopathological findings of biopsy taken from lesions were harmonious with SCLE. We diagnosed the patient as SCLE according to these findings and started therapy with chloroquine (500 mg/day) and topical glucocorticoids.

However the skin lesions were regretting initially, high fever and diffuse flare up in papullosquamous lesions in association with new developing widespread bullous skin lesions were developed on the third day of hospitalization of the patient (Figure 1,2). Moreover dyspnea and oligoanuria were occurred within few days, and then hallucinations, ataxia and dysarthria developed

Table 1. Laboratory examination in our patient with SCLE

	On admission	On the 3. day	On the 10. day	On the 4. week
Hemoglobin (g/dl)	9,7	9,9	8,4	11
Hematocrit (%)	28,9	29,3	25,3	33
Leukocyte (mm^3)	4600	2400	12900	5300
Platelet (mm^3)	196000	91000	153000	196000
BUN (mg/dl)	20	42	47	9
Kreatinin(mg/dl)	0,8	3,4	4,8	0,8
ALT (IU/ml)	49	13	24	44
AST (IU/ml)	14	39	24	26
LDH (IU/ml)	652	487	247	320
HBs Ag (mIU/ml)	Negative			
Anti-HBs (mIU/ml)	Negative			
Anti-HCV (mIU/ml)	Negative			
Anti-HIV (mIU/ml)	Negative			
ESR (mm/h)	70	54	41	31
CRP (mg/dl)	73	-	-	*12*
ANA (IU/ml)	>2048 specled, linear			
Anti-dsDNA (IU/ml)	930			

Figure 1. Bullous lesions in the forehead of the patient.

Figure 2. Bullous lesions in the neck and anterior chest wall of the patient.

suggesting central nervous system (CNS) involvement. Blood pressure was 100/ 70 mmHg, pulse rate was 112/min and body temperature was 39.5°C. Physical examination revealed bilateral diffuse crepitant rales at the end of the inspiration and pleural friction rub at the basement of the right lung, minimal hepatomegaly, and widespread skin lesions defined above. Leukocytopenia, thrombocytopenia and deterioration of renal function tests, which were normal, formerly were seen in laboratory examination. Viral serology for hepatitis B and C, and HIV were negative. There was diffuse bilateral retikulo-nodular infiltration and pleural thickening at the right side in chest radiography and high-resolution computed tomography of lung showed diffuse interstitial lung disease. Abdominal ultrasonography revealed slight hepatomegaly and bilateral parenchymal renal disease.

Because the patient's general status was getting worse rapidly, he was taken to intensive care unit for close monitorisation of the vital functions. Pulse glucocorticoid (1 gr daily for 3 days) and oral cyclophosphamide (at doses of 2.5 mg /kg /day) together with choloroquine were started immediately for treatment of the severe systemic involvement. Besides, we applied four plasmapheresis him because of rapid progress of the disease. Afterwards, therapy was continued with oral glucocorticoid (at dose of 1 mg/kg/day), cyclophosphamide and chloroquine.

The patient's status improved gradually by all these therapy and normal diuresis reachieved. Lung, CNS and skin findings in the patient were resolved within 3 weeks, and renal functional tests were returned to normal within 4 weeks. We discharged the patient in condition with coming regular control and maintaining his therapy.

Discussion

SLE is often called a woman disease because it occurs 10-15 times more frequently among adult females than among adult males. However, lupus can occur in either sex, and at any age. The symptoms of SLE are identical in men and women at the time of initial presentation of the disease, however there is more severe systemic involvement in men than in women at the later course of disease. Men account for 4 %-22 % of most large series of patients with SLE (6-8). Molina et al evaluated 1316 SLE patients and reported that males were 8.2 % of total of lupus patients. They found statistically higher prevalence of renal disease and vascular thrombosis and the presence of anti-dsDNA antibodies in the male than the female (6).

Subacute cutaneous lesions are seen 7-10 percent in patients with SLE. Skin involvement is most frequent in men than women patients with SLE (1,2). SCLE is a distinct subset of SLE clinically characterized by psoriasiform and/or annular lesions, by a mild or absent systemic involvement and by the presence of anti-Ro (SS-A) antibodies. Anti-Ro antibodies were found in 63 %-71 % of

the cases, but anti-dsDNA antibodies were found only in 3%-5% of cases (2-4).

It has been suggested that the papullosquamous type of SCLE, leukopenia, high titer of ANA (> 1: 320) and anti-dsDNA antibodies are risk factors for the development of SLE in a patient presenting with SCLE lesions. However, risk of systemic involvement is only 10%-15% in patients presenting with SCLE lesions (9). Cohen et al reported that the men with papullosquamous SCLE may be at higher risk for severe extracutaneous disease (10).

The male patient reported in here described subacute cutaneous lesions from time to time approximately for 2 years and was diagnosed as SCLE by history, physical and laboratory examination. ANA and dsDNA antibodies were positive at high titer at the admission time. Anti-Ro antibodies could not performed because of economical problems. In the light of explained knowledge above, it is clear that the patient has high risk for systemic involvement. Although, we evaluated him as an interesting case having rapid multiorgan involvement following high fever. Laboratory tests were negative for infections that may be a cause of this. Duffy et al reported that patients with lupus hospitalized have high risk for infections and disease was usually occurred with activation of lupus (11). Therefore, there may be a trigger factor (may be a virus) for activation of lupus in our patient, but we did not found any cause. Whatever, we evaluated the patient as a rare case because of special features such as to be man, to have bullous lesions and to have rapid severe multiorgan involvement in spite of diagnosed as SCLE.

Of the various cutaneous manifestations in patients with SLE, the development of a generalized bullous eruption is uncommon. The widespread vesiculobullous eruptions are usually appeared on the trunk and neck, and are characterized by dermal-epidermal separation with neutrophil-predominant inflammation in the upper dermis. There are a few case reports about bullous SLE rather than large series in literature (4,5,12,13). Between 15 and 20 percent of patients with SCLE lesions may develop acute cutaneous lupus lesions at some point. These skin lesions tend to be more transient than SCLE lesions and more commonly affects the malar areas of the face. An extremely acute form of acute cutaneous lesions is rarely seen that can simulate toxic epidermal necrolysis (2,3). We thought that the cause of bullous lesions in our patient might be a severe form of acute cutaneous lesions.

We report and conclude a male SCLE patient who have uncommon features such as bullous lesions and severe systemic involvement in this article. We emphasized that systemic organ involvement and bullous lesions may be seen sometimes and so it needs close follow up like SLE in patients with SCLE.

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