CASE REPORT

A case report of old age systemic lupus erythematosus

SIMIN ALMASI¹, NAFISEH EBRAHIMI², KAZEM ZAMANI³

¹Assistant Professor of Rheumatology, Department of rheumatology, Rheumatology Research Center, Firoozgar Hospital, Iran University of Medical Sciences (IUMS), Tehran, Iran.

² Medical student Rheumatology Research Center, Firoozgar Hospital, Iran University of Medical Sciences (IUMS), Tehran, Iran.

³Assistant Internal of Medicine Rheumatology Research Center, Firoozgar Hospital, Iran University of Medical Sciences (IUMS), Tehran, Iran.

ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that predominantly affects young women. Old age lupus occurs after the age 50 years and the disease occurs in 10-20% of patients over 50 years. This case was a 78-year-old woman who admitted to our hospital with polyarthritis, dyspnea, chest pain, typic malar rash, thrombocytopenia, lymphopenia:, positive antinuclear antibody (ANA):,and high titer anti-dsDNA from 3 weeks before admission. However older patients are more likely to have an insidious presentation and anti-ds DNA was very high and the other hand malar rash, polyarthritis and thrombocytopenia was reported very rare in old age in literatures bur our patients had acute presentation and her disease pattern similar pattern of disease in young patients.

Key words: old age, malar rash, lupus

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease predominantly affecting young people and showing a well-known female predilection. Lupus occurs in 10-20% of older patient. Old age lupus is the type of SLE whose manifestations begin after the age of 50 years.¹ However, in recent studies age of 65 years or greater has been recommended.²

Studies suggest that the clinical manifestation and serological features of old age lupus differ from lupus in younger patients. The female: male sex ratio declines from a 10:1 ratio in the younger patient to a 4:1 ratio in the older patient.³ This reduction has been related to the absence of sex hormones.⁴ Caucasian are more affected old age lupus in studies in multi-ethnic groups.⁵ Unlike to patients with regular lupus, old age lupus at the beginning of its course is characterized by the presence of nonspecific symptoms such as weight loss, arthralgia, weakness, fatigue, and cognitive disorders.⁶

Serositis, sicca symptoms, raynaud phenomenon, lung disease, fever and neuropsychiatric manifestation are more common in patients with old age lupus, while malar rash, discoid lupus and glomerulonephri-

Corresponding author:

Dr. Simin Almasi

E-mail: simin_almasi@yahoo.com

tis are less common in old age lupus compared with younger lupus patients. Unlike to younger patients in old age lupus the onset is gradual.⁷ Majority of old age lupus patients have a positive anti-nuclear antibody test (ANA), but anti-double-stranded DNA and hypocomplementaemia is lower in elderly-onset lupus patients. Rheumatoid factor (RF), anti-Ro/ (SS) A and anti-La/SSB are more positive in old age lupus patients.⁸

Aging is associated with decreased immune function. The thymus becomes smaller in old age however some thymic function intact.⁹ Often oligoclonal expansion develops, particularly of CD8+ T cells.10 It is also associated with reduction of B-cell lymphopoiesis.11 There is a shift from Th1 to Th2 cytokine with inceasing age and in old age lupus there are increased some autoantibody production such as RF, anti-Ro and anti-La antibodies.¹²

In old age lupus pro-inflammatory cytokines IL-6 and tumor necrosis factor-a may increased and this can explain the development of autoimmune disease in elderly individuals. On the other hand, a reduction of IL-2 (with the reduction in the activation of Tregulatory cells) has been shown with aging.¹³ The diagnosis of old age lupus may be delayed for many months: insidious onset, low prevalence and similarity to other more common disorders make the diagnosis of lupus challenging in this population.

This may give rise to a delay in diagnosis due to lack of typical clinical manifestations. Here we report a case of late onset-SLE associated with acute and typical lupus manifestation similar young people lupus.

Assistant Professor of Rheumatology, Firoozgar hospital, Iran University of Medical Sciences, Tehran, Iran

RBC	550×10⁴ ų/L
Hb	13.2gr/dl
WBC	4500/ UI(22.6% Lymph, 70%PMN, 7%Mixed)
PLT	8×10 ⁴ ų/L

Table 1, Laboratory Data on Admission

FANA	1/160(up to 1/40)(homogenous pattern)
Anti ds-DNA	749.7(up to 100)
C3	95(80-100mg/dl)
C4	26.2(13-75mg/dl)
ESR	15
RF	negative
CRP	+3
Anti-CCP	0.1(negative)
Troponin 1	negative
Cr	1mg/dl

Table 2, Laboratory Data on Admission

CASE REPORT

A 78-year-old woman was admitted to our hospital with polyarthritis in upper and lower limbs, dyspnea (functional class 3), retrosternal chest pain, pitting edema in lower limbs, malar rash from 3 weeks ago. Upon admission he was afebrile vital signs on presentation included blood pressure 120/70 mmHg, pulse rate 85 per minute with regular rhythm, body temperature 37.0 C°, and respiration 26 per minute. In joint examination symptoms of arthritis included swelling, erythema and severe tenderness was in all MCP (metacarpophalangeal) joints, knees, elbow and ankle joints. In lung examination crackle were heard at the base of both lung and 2+ pitting edema in lower extremities and jugular vein was prominent. The abdomen was soft and flat, and the spleen was not palpable, other abdominal physical exams were unremarkable. Neurological status was normal. Laboratory evaluation (Table 1), (Table 2) revealed thrombocytopenia and positive FANA and high titer anti-dsDNA.

Urine analysis is normal without cast or protein and also Hbs-Ag, HCV-Ab, HIV-Ab all were negative. Bone marrow aspiration and biopsy was unremarkable and malignant cells were not found. Abdominal and pelvic sonography was normal. In echocardiography moderate aortic insufficiency, EF(ejection fraction) 30% and, PAP(pulmonary arterial pressure)33 mmHg and Doppler ultrasonography of the lower limbs unremarkable and in lung CT-Scan fibrotic band, diffuse bronchiectasis, honeycomb in left lung was seen. The patient had no history of taking medication that causes drug lupus. Ischemic changes were seen in patients' ECG (electrocardiography) in 1, avL, V1-V6 leads. Based on clinical manifestation, history and laboratory test our diagnosis was lupus and on the basis 1997 American College of Rheumatology (ACR) revised criteria for classification of SLE, in

view of the presence of lymphopenia, positive ANA, high titer anti-ds DNA, poly arthritis, malar rash and thrombocytopenia. She was admitted in CCU and treated with prednisolon and hydroxychloroquine, her heart failure was treated and after two weeks the patient was discharged in good condition.

DISCUSSION

SLE is an autoimmune disease that predominantly affects women under 50 years of age. In a national survey of 1614 SLE patients in Japan, the onset in persons age 50 and above was rare at 6.4%.2 The disease has been reported after 65 years, much less and at the age of 90, only one case was reported recently.14 Old age lupus patients are relatively rare and patients usually have mild course of the disease, overall survival rate at 10 years is about 92%. The lag time from disease onset to diagnosis is significantly longer in the elderly group compared with the younger groups. Skin manifestation, photosensitivity and arthritis rarely occur in elderly SLE patients but our patient had typic malar rash and polyarthritis. Death occurred more frequently in old age lupus, in this patients cause of death is usually unrelated to SLE. Analysis of pooled data from the literature confirmed that old age lupus is characterized by a higher occurrence of serositis and pulmonary involvement. Generally, the onset is insidious, and the interval between onset and diagnosis is longer than in younger patients 3 but in our patient symptoms of lupus had acute onset. Prior studies have suggested that late onset-SLE is associated with a benign clinical course and better prognosis than SLE in younger patients.³ But some authors have considered that the degree of organ damage is greater in old age lupus3; therefore, early diagnosis and treatment is necessary for these patients. In this patient's serositis, interstitial lung disease (ILD) and Sjögren's syndrome are more prevalent. Arthritis, arthralgia and cytopenia are initial manifestation of old age lupus and in our case it was the same, although it has been reported that anti-dsDNA and anti-Sm are present less frequently in late onset-SLE.¹⁴ But in our case anti-dsDNA was very high, our patient had malar rash and polyarthritis and thrombocytopenia that this manifestation was reported very rare in literatures. Diagnosis of SLE in elderly individuals is difficult because of the lack of typical physical and laboratory features that are usually present in younger patients. In conclusion SLE occurs rarely in patients of advanced age since our patient was very old and Review in literature shown that the incidence of the disease is extremely rare in this age and we were reported this patient because disease is very rare in this age. Therefore, careful attention needs to be paid to latent symptoms and laboratory findings.

REFERENCES

1. PU SJ, Luo SF, Wu YJ, Cheng HS, and Ho HH. The clinical features and prognosis of lupus with disease onset at age 65 and older. Lupus with disease onset at age 65 and older. Lupus 2000; 9:96-100.

2. Boddaert J, Huong DL, Amoura Z, Wechsler B, Godeau P, Piette JC: Late-onset systemic lupus erythematosus: a personal series of 47 patients and pooled analysis of 714 cases in the literature. Medicine Baltimore) 83(6), 348 -359 (2004).

3. (Lockshin MD: Biology of the sex and age distribution of systemic lupus erythematosus. Arthritis Rheum. 57(4), 608 -611 (2007).

4. Bertoli AM, Vila LM, Reveille JD, Alarcon GS: Systemic lupus erythaematosus in a multiethnic US cohort (LUMINA) LIII: disease expression and outcome in acute onset lupus. Ann. Rheum. Dis. 67(4), 500 -504 (2008).

5. Ramos-Casals M, Garcia-Carrasco M, Brito MP, Lopez-Soto A, Font J: Autoimmunity and geriatrics: clinical significance of autoimmune manifestations in the elderly. Lupus 12(5), 341 -355 (2003).

6. Bertoli AM, Vila LM, Reveille JD, Alarcon GS:

Systemic lupus erythaematosus in a multiethnic US cohort (LUMINA) LIII: disease expression and outcome in acute onset lupus. Ann. Rheum. Dis. 67(4), 500 -504 (2008).

7. Lazaro D Elderly-onset systemic lupus erythematosus: prevalence, clinical course and treatment. Drugs Aging. 2007;24(9):701-15.

8. Gruver AL, Hudson LL, Sempowski GD: Immunosenescence of ageing. J. Pathol. 211(2), 144 -156 (2007).

9. Posnett DN, Edinger JW, Manavalan JS, Irwin C, Marodon G: Differentiation of human CD8 T cells: implications for in vivo persistence of CD8+. Int. Immunol. 11(2), 229-241 (1999).

10. Johnson SA, Cambier JC: Ageing, autoimmunity and arthritis: senescence of the B cell compartment - implications for humoral immunity. Arthritis Res. Ther. 6(4), 131-139 (2004).

11. Prelog M: Aging of the immune system: a risk factor for autoimmunity? Autoimmun. Rev. 52), 136-139 (2006).

12. Hakim FT, Gress RE: Immunosenescence: deficits in adaptive immunity in the elderly. Tissue Antigens 70(3), 179 -189 (2007).

13. Ward MM, Polisson RP. A meta-analysis of the clinical manifestations of older-onset systemic lupus erythematosus. Arthritis Rheum 1989; 32: 1226-32.

14. Hashimoto H, Hirose S, Kano S, Tsunematsu T, and Kabasawa K. Studies on clinical subsets and severity of systemic lupus erythematosus based on a 1987 questionnaire conducted in Japan--clinical analysis of the outcome and treatments in clinical subsets. Ryumachi 1992; 32; 27-38(in Japanese, Abstract in English).)

15. Cervera R, Khamashta MA, Font J, Sebastiani GD, Gil A, Lavilla P, et al; Morbidity and mortality in systemic lupus erythematosus during a 10-year period: a comparison of early and late manifestations in a cohort of 1,000 patients. Medicine (Baltimore). 2003 Sep; 82(5):299-308.