

# SYSTEMIC LUPUS ERYTHEMATOSUS – NOT TO BE FORGOTTEN IN THE ELDERLY

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## ABSTRACT

*A 70-year-old woman presented with a skin rash, chronic headaches, congestive cardiac failure, and a moderate pericardial effusion. These were attributed to systemic lupus erythematosus (SLE). She responded to a moderate dose of corticosteroids. SLE though rare in the elderly is a disorder that should not be forgotten.*

*Keywords: elderly, systemic lupus erythematosus, headache*

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## INTRODUCTION

Systemic lupus erythematosus (SLE) predominantly affects young women in their twenties. However, in one study<sup>(1)</sup>, up to 16% of patients with SLE had their disease onset after the age of fifty. Diagnosis in this age group is difficult, and it is likely that SLE goes unrecognised in a number of older patients with non-specific complaints.

SLE in the elderly has similar presentation as the younger onset patients, but is often missed because some signs and symptoms are common to the elderly (for example, arthritis, alopecia, myalgia, rash). Doctors are also reluctant to diagnose because this condition is associated with a younger age of onset.

## CASE REPORT

A 70-year-old female was admitted with a 3-day history of exertional breathlessness. This was her first episode of breathlessness. She had a skin rash which came on insidiously over the past one month. She also complained of a 5-year history of migrainous headache which was not investigated. She was a heavy smoker for the past 55 years and was diagnosed to have hypertension one year ago. Her hypertension was being treated by a general practitioner with Indapamide (Natrilix) 2.5 mg and Enalapril maleate (Renitec) 5 mg daily.

On examination, she was a cachectic lady in congestive cardiac failure. She had hyperkeratotic erythematous plaques all over her body, which on first impression, appeared to be partially treated psoriasis.

Chest X-ray showed cardiomegaly and congested lung fields. ECG, however, reviewed small voltages. There was no evidence of left ventricular hypertrophy or significant ischaemic heart disease. Blood investigations showed anaemia (Hb 8.2 g/l), leucopenia (total white count was 2200), mild renal impairment and a normal thyroid function. Erythrocyte sedimentation rate (ESR) was 45 mm in the first hour.

Further investigations of anaemia were negative (serum folate, vitamin B<sub>12</sub>, iron, ferritin, stools for occult blood and gastroscopy). Direct Coomb's test was negative and reticulocyte

count was 1.2%, CT scan of the brain was normal.

While in the ward, her headache persisted, became more severe and constant, and was not relieved with analgesics. Her heart failure was slow to resolve, and an echocardiogram showed moderate pericardial effusion, mild mitral regurgitation, moderate aortic, tricuspid and pulmonary regurgitation and an ejection fraction of 80%. She was not in cardiac tamponade.

She developed swinging fever of up to 39°C on the second week of admission. No source of sepsis was found. She had myalgia and generalised weakness to the extent that she needed help in her activities of daily living (ADL). Subsequently, autoimmune markers were found to be positive. Anti nuclear antibodies (ANA) was positive at > 1/640 and of speckled pattern, anti-ds DNA was positive at 8.8 mg/l (normal up to 5.4 mg/l), and complement levels were low (C3 was 0.39 g/l [normal range 0.47-1.23g/l], C4 was 0.11g/l [normal range 0.15-0.55g/l]). Anti Ro, La and Sm antibodies were negative. Muscle enzymes were also normal.

She was treated with oral prednisolone 20mg every morning. Within one week, she improved dramatically – her fever subsided, she became stronger and functionally independent, her headache resolved and the skin lesions improved. By the second week of treatment, her skin had returned to normal. On retrospect, these plaques were likely to be cutaneous lupus, though no skin biopsies were done to confirm that.

Three weeks after steroids were started, all her blood parameters had returned to normal except for the anaemia. Her total white cell count rose to 7,300 (with normal differential cell counts), ESR fell to 23 mm in the first hour, and anti-ds DNA became negative. Complement levels had also returned to normal. She was asymptomatic. Six weeks after steroid treatment, the cardiac shadow on repeat chest X-ray returned to normal size while a repeat echocardiogram showed resolution of the pericardial effusion.

## DISCUSSION

This patient presented, for the first time, with an acute onset of heart failure which was slow to resolve. Although she had anaemia, her doctors felt that this was insufficient to be the sole cause of her resistant heart failure. Causes of heart failure in lupus patients are usually multifactorial, and occurred in about 8% of patients<sup>(2)</sup>. In this patient, she also had a moderate pericardial effusion. This was the pathology that led to a systematic work up for the causes of pericardial effusion. Subsequent results pointed to SLE. She had clinical features (serositis, myalgia, skin rashes, fever and headaches) with serological confirmation (leucopenia, raised anti-native DNA antibody and anti-nuclear antibody), and so satisfied the 1982 revised Criteria of the American Rheumatism Association for

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the classification of SLE<sup>(3)</sup>.

In this patient, drug induced lupus was deemed unlikely. This was because of the insidious onset, multi-systemic involvement including skin lesions, raised titre of anti ds DNA and low complement levels, all of which are unusual in drug induced lupus. In addition, the patient was not taking any known at-risk drug. Both Indapamide and Enalapril have not been reported to cause lupus although there have been reports that Captopril, another ACE inhibitor, does<sup>(4,5)</sup>.

Although we did not know the onset of SLE in this patient, it was possible that her condition presented at the time of her headaches 5 years ago. Markus et al, in their study of migraine and headache in SLE<sup>(6)</sup>, found that a history of migraine was more common in SLE patients than controls ( $p < 0.05$ ), and within this group, headache was associated with disease activity and responded to specific SLE treatment.

In a review of literature comparing onset of SLE in older and younger patients, the age criteria differ between different investigators. The ages used were 45<sup>(7)</sup>, 50<sup>(8)</sup>, 55<sup>(9)</sup> and 60<sup>(10)</sup> years old.

Ballou et al<sup>(9)</sup> found that the most striking difference was in the racial distribution; 59% of the young onset patients (onset less than 55 at clinical diagnosis) were black, compared with only 20% of the older onset patients ( $p < 0.001$ ). There was also a significantly higher incidence of serositis in older whites and in blacks regardless of age, and more frequent hypocomplementemia in younger patients within both racial groups.

The interval between the time of onset and diagnosis was five years in the older group compared with three years in the younger group<sup>(7)</sup>.

Disease manifestations typical of older-onset SLE patients are known to vary widely among published studies. Arthritis, as a first symptom, was less common in the older onset group<sup>(1)</sup>. In contrast, this group showed an increased incidence of myositis. Serositis, interstitial pulmonary disease, Sjogren's syndrome<sup>(1)</sup>, discoid lupus, photosensitivity<sup>(11)</sup> and hepatic involvement<sup>(12)</sup> were most strongly and consistently associated with older-onset SLE. Pericarditis was detected clinically in 20% to 30% of the older-onset SLE and 50% to 60% was found on autopsy<sup>(2)</sup>. Alopecia, Raynaud's phenomenon, fever, lymphadenopathy<sup>(1)</sup> cutaneous vasculitis, oral ulcers, peripheral neuropathy, neuropsychiatric illness<sup>(11)</sup> and nephropathy<sup>(13)</sup> were present less frequently in this group. Other studies, however, showed conflicting results.

Gossat et al<sup>(7)</sup> found a high incidence of neuropsychiatric disturbances and low incidence of serositis, while non specific complaints of fever, weight loss, and malaise were often the only presenting clinical features. Locally, Feng PH and Boey ML<sup>(14)</sup> reported a predominance of pleuro-pericardial symptoms and non specific complaints of myalgia, arthralgia and unexplained weight loss among elderly Chinese. Regression analysis suggested linear change in disease expression with age rather than distinct age-related subgroups<sup>(8)</sup>.

High titres of anti-ds DNA tended to occur less often and the incidence of anti-Ro antibodies was lower in the older onset group<sup>(1)</sup>. Serologic abnormalities were milder in older patients. Hypocomplementemia, anti-DNA antibodies, and C1g precipitins occurred less frequently while rheumatoid factor<sup>(8)</sup> and anti-La antibodies<sup>(10)</sup> occurred more often in older onset patients with SLE. Other series found an increased incidence of anti-Ro and anti-La antibodies in keeping with the increased prevalence of Sjogren's syndrome in older onset SLE<sup>(15,16)</sup>.

Biochemically, older onset lupus resemble drug-induced lupus in that anti single stranded DNA (anti ss DNA) antibodies are more common than anti-ds DNA antibodies<sup>(8,17)</sup> and serum complements are rarely low.

Laboratory measurements of antibody titre may constitute a problem in interpretation, unless the titres are high. Antibody titres must be viewed in the context of clinical symptoms and signs, which in the elderly patient, may be non specific as elaborated above. In a cross-sectional study<sup>(18)</sup> of 100 healthy elderly individuals, 5% was found to have significant titre of anti-nuclear antibodies. Other series showed significant titres of anti-nuclear antibodies in 20% of those older than 90 years of age<sup>(17)</sup>.

Factors associated with disease activity, such as elevated ESR, neutropenia, and thrombocytopenia were less frequently encountered than in younger patients<sup>(7)</sup>. The concomitant appearance of both IgG and IgM anti-histone antibodies was observed only in SLE patients and seem to be a marker for lupus and its variants<sup>(19)</sup>.

Treatment is similar to that in younger patients. However, they usually require lower doses of steroids or azathioprine<sup>(11)</sup>. About 40% of the patients require steroids at doses of 25mg or more a day, 25% require low dose only, and 35% do not need steroids<sup>(11)</sup>.

The course of disease appeared milder<sup>(15)</sup>. Five-year survival in the younger onset group was similar to that of the older group<sup>(9)</sup>. Survival had been reported to be 92% for 5 years and 83% for 9 years<sup>(15)</sup>. Therefore, this condition should be borne in mind to reduce the morbidity of the patient.

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