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Late Onset Systemic Lupus Erythematosus in an Afro-Caribbean Patient: A Diagnostic Challenge

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Abstract

Systemic Lupus Erythematosus may be missed in the elderly population as the clinical features may resemble more common conditions. Treatment remains a challenge in this group given the co-existing chronic illnesses and drug-related toxicities. Herein, we report a case of a 72-year-old Afro-Caribbean woman diagnosed with systemic lupus erythematosus after having pulmonary and renal manifestations.

Keywords: ***Systemic lupus erythematosus; Elderly; Dyspnea; Acute kidney injury***

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Introduction

Systemic Lupus Erythematosus (SLE) is a chronic multisystem autoimmune disease that is associated with antibody production and the deposition of complement-fixing immune complexes [1,2]. The incidence of SLE decreases rapidly after age 50-years and is very uncommon in patients older than 70-years of age [2]. The age distribution of acute SLE diagnosis poses a clinical dilemma for physicians encountering acutely ill elderly patients presenting with features suggestive of acute lupus.

New onset acute SLE can often be missed in the elderly population due to the co-existence of chronic illnesses that may resemble SLE [3]. Emerging studies have shown that Caucasians are more likely to manifest with elderly onset Lupus which emphasizes the need for further studies of populations of African and Caribbean ancestry to increase our understanding of environmental, genetic, and health care issues surrounding this diagnosis and mitigate the current disparities [3,4].

We present a case of an elderly Afro-Caribbean woman diagnosed with SLE after insidious onset of shortness of breath, fatigue and weight loss with rapid progression to hypoxic respiratory failure and renal failure. The patient's unusual presentation delayed the diagnosis which highlights the need for prompt recognition and treatment to prevent end-organ damage. This may ultimately determine a patient's outcome and long-term prognosis.

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Case Presentation

The patient was a 72-year-old Afro-Caribbean female with diabetes mellitus, hypertension and atrial fibrillation who presented with dyspnea at rest, dry cough and fatigue. There was no associated orthopnea, chest pain, pedal edema, fever or chills. She had no history of rash, joint pain, stiffness or altered mentation. Of note, she was well until three weeks prior to presentation when she experienced a dry cough and dyspnea on exertion and was given a five-day course of azithromycin for acute bronchitis. However, the symptoms gradually worsened and in addition, she experienced generalized weakness, loss of appetite and unintentional weight loss of 5lbs. She was well controlled on oral medications; aspirin 81mg po daily, losartan 100mg po daily, metformin 500mg po bid and xarel to 15mg po daily. Family history revealed SLE in her sister diagnosed in her 40's.

On presentation her vitals: blood pressure of 162/73 mmHg, pulse 69 beats per minute, temperature 98.2°F, respiratory rate 16 breaths per min and pulse oximetry of 99% on room air. Physical examination revealed temporal wasting and mucosal pallor. There were no oral lesions, lymphadenopathy or skin lesions. Cardiac examination revealed an irregular heart rhythm without any murmurs or gallop. Chest was clear to auscultation; abdominal examination was negative for organomegaly or masses and there was no evidence of joint swelling or leg edema.

Test	Actual result	Reference value/range
Hemoglobin	9.5	12.0-15 g/dl
Hematocrit	26%	38-46 %
MCV	84	80-96 fL
White cell count	4.1	4-11 x 10^9/L
Lymphocyte count	0.8	0.9-29 k/ul
Platelet count	112	130-400 x10^9/L
Prothrombin time (PT)	11.2	10.5-13.1 s
Partial thromboplastin time (PTT)	32.7	28.9-38.9 s
Direct Coombs test	Positive	Negative
Sodium	127	136-145 mEq/l
Potassium	3.5	3.5-5.1 mEq/l
Chloride	96	98-107 mEq/l
Bicarbonate	15	21-31 mEq/l
Blood urea nitrogen	98	5-25 mg/dl
Creatinine	7.21	0.7-1.3 mg/dl

Table 1: Showing initial laboratory results.

Laboratory studies were notable for anemia with hemoglobin of 9.5 g/dl, lymphopenia 0.8 k/ul, thrombocytopenia with platelet count of 112 × 109/L, positive Direct Coombs test and significant renal impairment with metabolic acidosis: blood urea nitrogen 98 mg/dl, creatinine 7.2 1 and bicarbonate of 15 mEq/l (Table 1). Fractional excretion of sodium was 4.8%. Of note, serum creatinine was 0.9 mg/dl three weeks prior. Urinalysis showed moderate proteinuria with the absence of casts and spot urine protein to creatinine ratio was 1.69 g/day. Renal ultrasound showed evidence of renal parenchymal disease without

hydronephrosis. Transthoracic echocardiogram was significant for trivial mitral and aortic regurgitation and left ventricular ejection fraction (LVEF) of 55%.

The patient was treated with intravenous normal saline for acute kidney injury with metabolic acidosis presumed from dehydration and medication side effect. Despite adequate fluid resuscitation the serum creatinine remained elevated. On day four of admission she developed fever of 100.4°F, chills, worsening dyspnea, one episode of small-volume hemoptysis and was found to have diffuse course crackles in both lung fields. Her hemoglobin decreased from 8.2 to 6.9 g/dl. Chest radiograph showed increased alveolar markings and a heterogenous opacification in the right upper zone (Figure 1). Computed tomography chest showed multifocal alveolar disease, most severe in the right upper zone (Figure 2A and 2B). Due to acute hypoxic respiratory failure (fall in oxygen saturation to 84% on room air) BIPAP was initiated with improvement in oxygen saturation. The constellation of symptoms suggested acute pulmonary edema versus acute pulmonary vasculitis. Acute pulmonary edema was subsequently ruled out when there was no improvement following high dose furosemide over a 2 day period. Connective tissue disease was considered at this point due to acute concomitant pulmonary and renal manifestations.

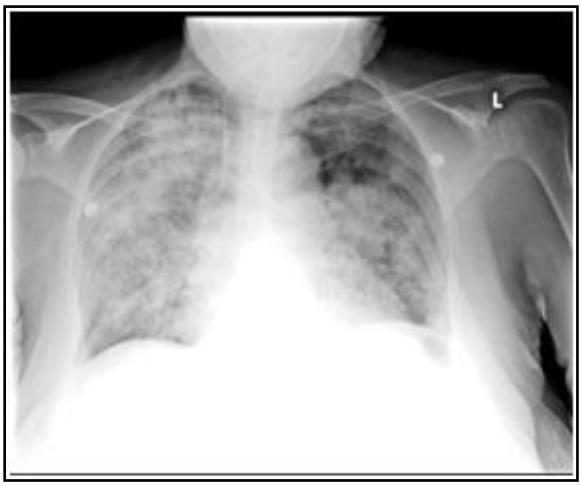
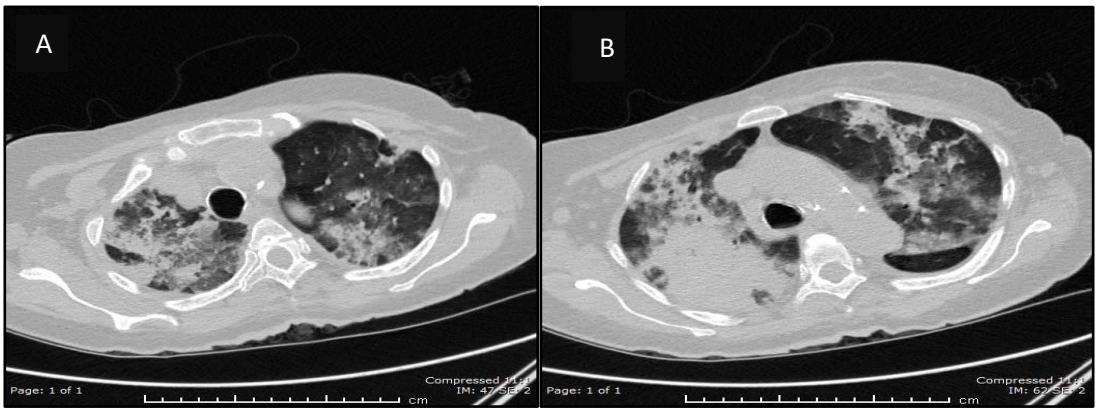


Figure 1: Chest radiography which showed patchy infiltrates in both lung fields with near confluence seen in right upper zone.



Figures 2A and 2B: CT chest findings showing patchy bilateral multifocal disease, most severe in the right upper lobe.

Rheumatologic work up of acute pulmonary - renal syndromes revealed: Positive ANA 1:640, positive Anti-Smith, Anti dsDNA and Anti-myeloperioxidase antibodies (Table 2). Kidney biopsy showed features in keeping with hypertensive

nephrosclerosis with secondary focal segmental glomerulosclerosis and superimposed acute tubular injury. The sample was not sufficient for immunoflorescence study but periodic acid schiff (PAS) (Figure 3) and electron microscopy showed few sub endothelial deposits that may be due to SLE.

Test	Actual result	Reference range/value
Anti dsDNA	60	0.00-10 U/mL
Anti-Smith	11	0.00-7.00 U/mL
Anti-SSB	Negative	Negative
Anti-SSA	Equivocal	Negative
Rheumatoid factor	132	0-20.0iU/ml
Anti-RNP	Negative	Negative
C 3, C4	C4 18.7	16-47.0mg/dl
	C3 65.4	83-200mg/dl
CRP	64.9	0-8.0 mg/L
Anti-GBM	Negative	Negative

Table 2: Showing results of Rheumatologic work-up.

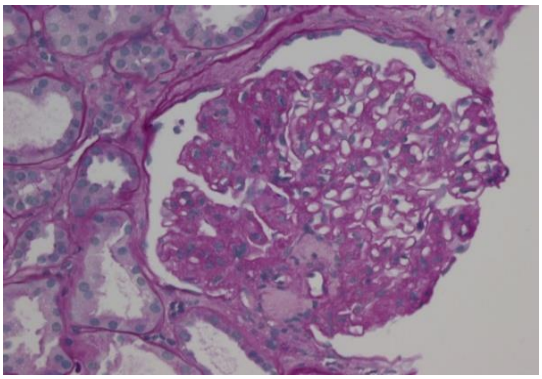


Figure 3: Periodic acid Schiff stained image of a representative glomerulus with few sub endothelial deposits.

She was treated with high-dose intravenous methylprednisolone (1 g/day for 3 days) followed oral prednisone 60 mg/day. The respiratory symptoms improved significantly following initiation of steroids such that she was weaned off oxygen therapy. The patient's renal function did not improve and she later required hemodialysis.

Four months after the initial manifestation of lupus, the fatigue, malaise and dyspnea resolved, she however remains on dialysis. The prednisone has been tapered to 30 mg po daily and mycophenolate mofetil added at 500 mg po daily and later up titrated to twice daily.

Discussion

Systemic Lupus Erythematosus is a chronic, multisystem, inflammatory disorder of autoimmune etiology caused by dysregulated immune response in genetically predisposed individuals [1]. It has a complex phenotype with variable manifestations. The overall crude incidence rates are 0.4 for white males, 3.5 for white females, 0.7 for African-American males, and 9.2 for African-American females [5]. One study suggested that African-American females with SLE were

diagnosed at a younger mean age compared with white females 44.6 ± 10.3 years vs. 47.6 ± 10.6 years [6]. Late onset SLE is defined as onset of disease after the age of 50 years and has been reported in close to 20% of patients [2]. It is described most commonly in Caucasians, but more multi-ethnic large-scale studies are needed [3,4].

The low estrogen state of menopause is thought to be the main reason for diminished prevalence of SLE in older women. At the same time, the older patient with acute SLE often has decreased survival due to increased frequency of infections, cardiovascular disorders, malignancies and drug-induced complications [7]. As in our case, these patients often have delayed diagnosis in part due to the consideration of more common diagnoses first and the lack of awareness of disparate incidence in different ethnic groups resulting in increased morbidity and mortality.

Glomerulonephritis is seen less commonly in elderly onset lupus patient compared with younger patients with findings of arthritis, fever, serositis, sicca symptoms, Raynaud's syndrome, lung disease and neuropsychiatric symptoms seen more frequently. Most elderly patients have a lower prevalence of anti-double-stranded DNA and hypocomplementaemia [7]. These findings are in contrast to the patient presented. The diagnosis of connective tissue disease such as SLE should be considered in elderly women of Afro-Caribbean descent presenting with constellation of pulmonary, renal and hematological manifestations (Coombs positive, anemia, thrombocytopenia). Laboratory studies such as Anti-dsDNA (97%) and anti-Smith (98%) are specific for the diagnosis of SLE but are not uncommon for patients to also have Rheumatoid factor and Anti-Neutrophil Cytoplasmic Antibodies (20-30%) [7]. The diagnosis of SLE was made based on the Systemic Lupus International Collaborating Clinics Classification (SLICC) criteria [8] (Table 3).

Clinical Criteria	Immunologic Criteria
Acute cutaneous lupus (maculopapular lupus rash, malar rash, photosensitive lupus rash etc)	**High ANA concentration**
Chronic cutaneous lupus (classical discoid rash, mucosal lupus, lupus panniculitis, etc)	**High anti-dsDNA antibody concentration**
Oral or nasal ulcers	**Presence of anti-Smith**
Nonscarring alopecia	Positive antiphospholipid antibody
Synovitis in >2 joints	Low complement (**C3**,C4, CH50)
Serositis	**Direct Coombs test**
Renal (**urine protein** or RBC casts)	
Neurologic (seizures, psychosis, others)	
Hemolytic anemia	
Leukopenia or **lymphopenia** (without an identifiable cause)	
Thrombocytopenia (without an identifiable cause)	

Table 3: SLICC classification for systemic lupus rrythematosus.
Requirements: ≥4 criteria (at least 1 clinical and 1 laboratory criteria) or biopsy-proven lupus nephritis with positive ANA or Anti-DNA.
Note: Highlighted in red and bold are the features used to make the diagnosis in this patient.

The case presented highlights not only late onset Systemic Lupus Erythematosus but a diagnostic challenge. The respiratory findings initially led to consideration of pulmonary edema. Pure pulmonary edema, however, produces vascularization and not the patchy opacities seen on the x-rays. Instead, this finding along with the improvement on steroids strongly supports the diagnosis of Lupus Pneumonitis. The diagnosis of Lupus Nephritis can be made on light microscopy alone, Immunoflorescence or Electron Microscopy is only supportive [9]. It is more likely that the patient had underlying intrinsic

kidney damage from hypertension and diabetes mellitus and subsequent changes in hemodynamics, increase in cytokines and other inflammatory markers as seen in an acute illness hastened progression to acute renal failure and need for dialysis.

In summary, the first-time presentation of SLE in patients older than 50-years is an underappreciated and overlooked phenomenon. As such, it is often missed and remains a diagnostic conundrum for many physicians given its atypical epidemiology and presentation, despite literature highlighting older age and SLE as an emerging association.

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