Systemic Lupus Erythematosus: the rare MANifestations

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Introduction

Systemic lupus erythematosus: chronic autoimmune condition that can affect virtually any system in the body

Presentation varies from mild joint and skin involvement to life threatening renal , neurologic, or hematologic involvement

Failure to recognize SLE cause of life-threatening manifestations may delay treatment and increase mortality.

Presentation:

HPI: 47 year-old is transferred from outside facility with subacute onset hemoptysis, acute hypoxia, and acute renal failure.

Past medical history:
GERD (on omeprazole)
Anxiety (on citalopram)

Family History: no known family history of autoimmune disease

Social History: occasional marijuana use, no tobacco, no alcohol

PE :
Skin: Dry and warm. No rashes, bruises
Lungs: course breath sounds bilaterally with no wheezing, rhonchi, or rales
Cardiovascular: RRR. No murmurs/gallops/rubs
Abdomen: + BS, soft, non-tender, non-distended
Neurologic: Intubated and sedated, responsive to pain

Lab data:
CBC: WBC 20.3, Hgb 6.9, Hct 21%
CMP: Cr 5.26, BUN 49, LFTs normal on admission

Imaging:
CT Chest, abdomen - Mild mediastinal adenopathy. Tiny bilateral pleural effusion, Diffuse patchy groundglass opacities/consolidation in both lungs. Suggestive of acute inflammatory/infectious process. Changes suggestive of fatty liver with minimal fibrosis

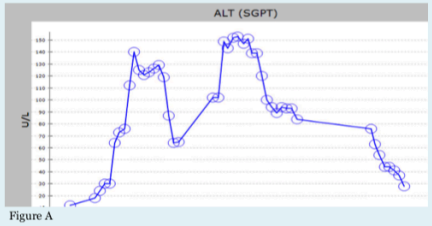

Figure A

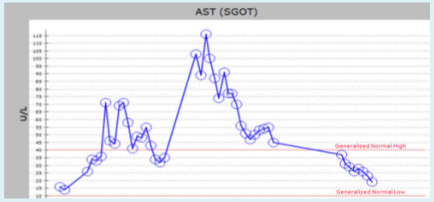

Figure B

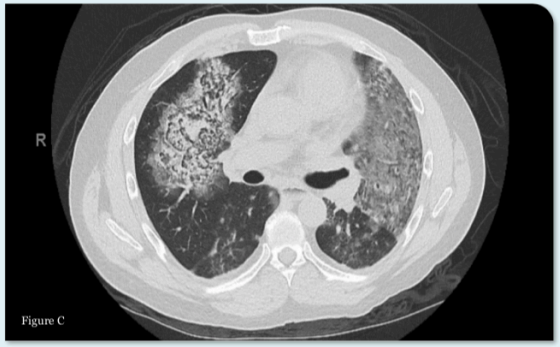

Figure C

Figure A: graph demonstrating peak and fall of ALT after discontinuation of cyclophosphamide
Figure B: graph demonstrating peak and fall of AST after discontinuation of cyclophosphamide
Figure C: Chest CT on admission demonstrating diffuse alveolar hemorrhage

Clinical course:

- Admitted to the ICU for intubation, respiratory support, and initiation of dialysis
- Renal biopsy showed crescentic glomerulonephritis with positive p-ANCA, ANA, and low C3 complement
- Eventually extubated to tracheostomy with no complication
- Cyclophospamide and corticosteroids initiated for likely lupus vasculitis
- LFTs began to rise approximately 2 weeks into treatment
- Initial read from liver showed concern for cyclophosphamide but return read showed early nodular hyperplasia secondary to lupus
- Transitioned to mycophenolate and conservative management for hepatic involvement
- LFTs eventually down trended to within normal range over several weeks

Discussion

This is a case of a middle-aged male presenting with diffuse alveolar hemorrhage and later complicated by early nodular regenerative hyperplasia of the liver due to systemic lupus erythematosus – two rare manifestations of a rare disease.

Epidemiology

- Prevalence of 164-406/100,000 among women
- Typical onset between ages 16-55
- Men have higher frequencies of renal disease, skin manifestations, cytopenias, serositis, neurologic involvement, thrombosis, cardiovascular disease, hypertension, and vasculitis than women
- Men have higher one year mortality than women

Manifestations: may involve nearly every organ system

- Constitutional (fever, fatigue, weight loss) – 90-100% of patients
- Arthritis or arthralgias – 90% of patients
- Mucocutaneous involvement – 80% of patients
- Cardiac manifestations – 25% of patients
- Renal manifestations – 50% of patients

Pulmonary hemorrhage is a rare complication affecting <5% of patients with SLE.

Early nodular hyperplasia is a rarely reported complication of SLE

References:

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L-J Lu, et al. **Review: Male systemic lupus erythematosus: a review of sex disparities in this disease.** Lupus. 2010 Feb;19(2):119-29.