An Atypical Presentation of Systemic Lupus Erythematosus

Lawrence Wayne Liu1,2 and Duc To2
1Loma Linda University School of Medicine, Loma Linda, CA, US
2Riverside University Health System, Moreno Valley, CA, US

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Abstract

Background: Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disorder that affects almost every organ system in the body. Its pathogenesis, although unclear, seems to be related to higher levels of estrogens resulting in the increased formation and deposition of immune complexes leading to widespread systemic inflammation [1]. It has a predilection for non-Caucasian women of child-bearing age and can cause serious complications in many organ systems.

Case presentation: In this case, we report a highly atypical presentation of SLE previously healthy 46 year old female. She presented with atypical chest pain and elevated troponin levels concerning for Acute Coronary Syndrome (ACS). During the course of clinical workup, SLE was diagnosed and Lupus Nephritis was found on Kidney Biopsy. She was started on treatment for her SLE and Lupus Nephritis with improvement of her symptoms.

Conclusions: This study highlights an interesting variant presentation of SLE and the unique diagnostic and treatment challenges that it posed. Further research into the pathogenesis, prevention, and diagnosis of SLE is warranted and this study aims to promote research into this and other autoimmune conditions.

Keywords: Rheumatology; Lupus; Autoimmune; Cardiology; Chest pain; Nephrology; Nephritis

Introduction

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disorder that affects almost every organ system in the body. Its pathogenesis, although unclear, seems to be related to higher levels of estrogens resulting in the increased formation and deposition of immune complexes leading to widespread systemic inflammation [1]. It has a predilection for Asians, Hispanics, and African Americans over Caucasians. And it favors females over males of child-bearing ages in a 12:1 ratio. SLE is common diagnosed from 15 to 44 years of age. It is strongly associated with high levels of Antinuclear Antibodies (ANA) with variants of SLE demonstrating additional serologies: e.g. Anti-Double stranded DNA Antibodies (anti-dsDNA) and Anti-Histone Antibodies. SLE most commonly presents with fatigue, fever, myalgia, weight loss, arthritis/arthralgias, cutaneous (malar rash, discoid rash, photosensitivity) and mucosal (painless oral and nasal ulcers) lesions [2]. Exacerbations of Lupus, coined “Lupus Flare,” are typically triggered by environmental factors (e.g. Epstein-Barr virus, UV light, infections, chemicals, or other environmental stressors).

Case Report

A previously healthy 46 year old Hispanic female presented to the ED with one day of a new, 8/10 sub-ternal chest pain with radiation to the back. It was worse with movement and breathing and better with rest. No improvement after nitroglycerin, gabapentin, and ibuprofen. Three weeks prior to admission, she began to have arthralgias and edema of her legs, ankles (left greater than right) and wrists beginning after a bug bite on the dorsal aspect of the left foot. Associated symptoms: subjective fevers (resolved by Tylenol and Ibuprofen), chills, fatigue, weakness, headaches, and nausea. She denied dyspnea, dyspnea on exertion, weight change, orthopnea, PND.

On exam, she had TTP to the mid-sternal area and bilateral lower extremities, mild edema of the LLE with active and passive ROM limited by pain. A small 2x2 cm area of erythema without TTP, exudates, hematoma, or fluctuance was noted at her left medial malleolus (where the bug bite occurred). Negative heart and lung exam: HEART Score 2. ASCVD 10 year risk of 2.3%.

The patient was afebrile with stable vital signs. Labs were significant for elevated troponin of 0.127, Hgb 10.9. EKG and Chest X-Ray were unremarkable. On Hospital Day (HD) 4, cardiac enzymes were discontinued because of low suspicion of cardiac etiology for chest pain given negative EKG and down-trending Troponin levels (0.127>0.67>0.81>0.46>0.24>0.12). Left Ankle X-ray showed mild soft tissue swelling (consistent with arthritis). She had nocutural fevers (102-103F) but cultures and microbial serology studies were negative. A Rheumatologic workup showed positive anti-dsDNA, SSA, ANA, C3/C4, ESR, CRP, Haptoglobin, LDH. UPC showed significant proteinuria. Other studies were negative.

We diagnosed her with SLE given her polyarthritis, proteinuria, positive ANA, Positive anti-dsDNA. She was started on Solumedrol and Plaquenil with improvement. IR-guided renal biopsy was conducted for evaluation of possible Lupus Nephritis. She was then discharged with Prednisone and Plaquenil with instructions to follow up with outpatient Nephrology and Rheumatology within a week. Renal Biopsy Results from HD 15 showed Lupus Nephritis Stage 3A (Focal Proliferative Lupus Nephritis) and treatment with CellCept was started.
Conclusion

This is a rare incidence of SLE presenting with Atypical Chest Pain. It is interesting that this patient's Lupus Flare demonstrated slightly positive Troponin levels. Most cases of SLE present with fatigue, fever, myalgia, weight loss, poly-arthritis/arthritis, exanthems, and/or enanthems, and positive Lupus serologies (e.g., ANA, anti-dsDNA, anti-Smith, anti-Histone, etc.). Another interesting aspect of this case is that most cases of SLE involve a history of cutaneous or mucosal lesions (photosensitivity, discoid lesions, malar rash, nasal or oral ulcers) whereas this patient had no history of such lesions. The patient was also diagnosed at an older age than is common for SLE. Overall, this study highlights an interesting variant presentation of SLE and the unique diagnostic and treatment challenges that it posed. Further research into the pathogenesis, prevention, and diagnosis of SLE is warranted and this study aims to promote research into this and other autoimmune conditions.

References