Recurrent pericarditis as the presenting symptom for diagnosis of systemic lupus erythematosus

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ABSTRACT

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Case Report: A case of an African-American female with pericarditis at the time of diagnosis. The patient was a 28-year-old female who presented to the emergency department complaining of chest pain and shortness of breath. She reported experiencing similar symptoms on two separate occasions prior to this admission. After appropriate workup an ANA panel was ordered on suspicion which revealed pertinent findings such as an ANA titer of 1:1280, anti-DNA(ds) 10 IU/mL, RNP antibodies >8 AI, Smith antibodies 2.2 AI.

Conclusion: Diagnosing SLE can be considered challenging, since there are varying degrees of clinical manifestations from patient to patient. The lifetime prevalence of some cardiac manifestation in SLE is estimated to be 50%, and should be high on the differential of any presentation with chest pain or shortness of breath. It is crucial that physicians consider SLE as a diagnosis when new onset pericarditis occurs in African-American females.
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Keywords: Autoimmune, Criteria, Diagnosis, Pericarditis, Systemic lupus erythematosus (SLE)

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic inflammatory autoimmune disease that affects multiple organs. Common findings at presentation are fatigue, fever, weight loss, arthritis/arthralgias, skin manifestations, and renal pathologies. Herein, we focus on pericarditis as a less frequent although significant sign leading to the diagnosis of SLE. This disease targets an estimated one and a half million Americans, and over five million worldwide. Sixty-three percent of these patients report being incorrectly diagnosed, with more than half the patients visiting four or more doctors before correct diagnoses [1]. Pericardial involvement many times precedes clinical manifestations of SLE. In SLE specifically, pericardial involvement concurrently...
presenting with pericardial effusion is the most common type of echocardiographic abnormality found in >50% of adult patients, suggesting a significant reason why a patient presenting with the latter should be screened for SLE. In addition, combined autopsy series revealed pericardial involvement in 62% of patients with SLE [2]. It is critical to consider pericarditis as an important presenting symptom of SLE, since early recognition and management will reduce morbidity and mortality.

CASE REPORT

In this report, we describe a case of an African-American female with pericarditis at the time of diagnosis. A 28-year-old female presented to the emergency department complaining of chest pain and shortness of breath. Upon admission to our hospital she described her chest pain as 6–7 out of 10 with radiation to the scapula. The pain was aggravated by movement and inspiration and alleviated by leaning forward. She denied fever, nausea, vomiting, rash or joint pain. Family history was significant for a mother with SLE. On initial examination, the patient had temperature 98.3°F, blood pressure 128/78 mmHg, respiratory rate 20 breaths per minute, and heart rate 95 beats per minute. Normal S1 and S2 heart sounds were heard on auscultation with a regular rate and rhythm. No murmurs, rubs, or gallops were heard. Decreased breath sounds were noted bilaterally at lung bases.

The patient reported experiencing similar symptoms on two separate occasions. The first episode occurred one month prior to the current admission, where she was treated with NSAIDS. Presuming a classic case of pericarditis, the patient was discharged from the hospital. A second episode with identical chest pain occurred three weeks prior to admission, which she sought help from a different hospital than the former. At that time, an echocardiogram was performed revealing a pericardial effusion. NSAIDs were given and a rheumatology follow-up was advised.

Pertinent laboratory findings which lead to the diagnosis in this admission included a comprehensive ANA panel; ANA screen positive with anti-nuclear antibody titer of 1:1280 (<1:80 negative), anti-DNA(ds) 10 IU/mL (≤4 IU/mL negative), RNP antibodies >8 AI (<1.0 AI negative), Smith antibodies 2.2 AI (<1.0 AI negative), anti scleroderma-70 0.3 AI (<1.0 AI negative), Sjögren’s anti-SSA >8.0 AI (<1.0 AI negative), Sjögren’s anti-SSB 3.3 AI (<1.0 AI negative), positive antichromatin >8.0 AI (<1.0 AI negative), anti-Jo1 <0.2 AI (<1.0 AI negative), anticientromere B <0.2 AI (<1.0 AI negative), sedimentation rate-Westergren 79 mm/hr (women under 50 years old: < 20 mm/hr), complement C3 127 mg/dL (75–175 mg/dL), complement C4 13 mg/dL (14–40 mg/dL), elevated CRP cardio 22.052 (≤10 mg/L in non-acute phase range). A transthoracic echocardiogram was performed which showed a pericardial effusion.

DISCUSSION

Pericarditis is the most common cardiac manifestation seen in SLE patients. Most cases can be managed with NSAIDS. However some cases may lead to cardiac tamponade. The pathogenesis of SLE is not entirely understood, however, antibodies and immune complexes (IC) are factors contributing to the clinical manifestations seen in these patients. They cause tissue damage via vasculopathy, which is ultimately responsible for many of the clinical manifestations of SLE. Immune complexes cause the activation of compliment and cause further damage to the associated organ affected. Immunofluorescence and electron microscopy have identified immune complexes at the dermal-epidermal junction of the pericardium. This deposition may precipitate pericarditis as a presentation in SLE patients. Immune complexes pathogenicity depends on the characteristics of the antibodies. Different factors include the size, charge, affinity, specificity, and the ability to activate inflammatory mediators and/or compliment. The rate at which the Fc receptors clear the immune complexes on macrophages also plays a role in pathogenicity [3].

There are two main hypotheses concerning the formation of antibodies in SLE patients. One involves polyclonal B cell activation, which occurs when B lymphocytes are stimulated non-specifically to form antibodies. The other is an antigen driven response with antibodies directed against these specific antigens. Antigens can be either autologous or exogenous. Autologous antigens may be altered through sunlight, infection, or other sources of tissue injury to stimulate an immune response. Exogenous antigens may become immunogenic through molecular mimicry [4].

Diagnosing SLE can be considered challenging, since there are varying degrees of clinical manifestations from patient to patient. Clinically, patients may have symptoms ranging from a rash and joint pain to life-threatening central nervous system, hematological, or renal involvement. Differentiating SLE from other autoimmune diseases proves difficult, however, increasing your index of suspicion and following the guidelines below will be clinically useful in the diagnosis of SLE in future patients. Ultimately, diagnosing SLE is based upon a skilled clinician who recognizes the signs and symptoms consistent with SLE and supports them with serological markers [5].

The current recommended guidelines to follow are the systemic lupus international collaborating clinics (SLICC). The diagnosis of SLE by the SLICC requires 4 out of the 17 criteria listed below, which contain at least one criterion representing the clinical and immunologic sections respectively. Diagnosis can also be made if the patient’s biopsy proves Lupus nephritis in the presence of a positive ANA or anti-dsDNA. The following are the clinical criteria: acute cutaneous lupus, chronic cutaneous lupus, non-scarring alopecia, oral or nasal
ulcers, joint disease, serositis, renal involvement, neurological involvement, leukopenia or lymphopenia, and thrombocytopenia. Immunologic criteria are as follows: ANA, anti-dsDNA, anti-Sm, antiphospholipid, low complement, and direct Coombs test positive in the absence of hemolytic anemia [6].

Furthermore, there are varying degrees of probability that the diagnosis is in fact SLE. A definitive diagnosis of SLE consists of the SLICC diagnostic criteria previously stated. Probable SLE is defined as patients having two or three SLICC diagnostic criteria and at least one of the following criteria: optic neuritis, aseptic meningitis, glomerular hematuria, pneumonitis, pulmonary hemorrhage, pulmonary hypertension, interstitial lung disease, myocarditis, verrucous endocarditis, abdominal vasculitis, Raynaud phenomenon, and elevated acute phase reactants. Lastly, possible SLE is considered when only one SLICC criteria is met and at least one of the other features listed above is present [7]. In this case, positive findings included ANA, anti-dsDNA, anti-Sm and pericarditis thus satisfying the SLICC criteria for SLE.

Treatment typically depends on the disease manifestations. When patients become symptomatic with signs of pericarditis, non-steroidal anti-inflammatory drugs (NSAIDs) are typically initiated. Colchicine may be added to NSAID therapy if risk of recurrence is highly likely. In cases where inflammation does not subside with NSAIDs and colchicine, or if the patient is not able to tolerate these medications, then glucocorticoid therapy could be considered an alternative therapy [8].

In acute pericarditis some patients are classified as high-risk, so hospitalization is recommended for close evaluation and treatment. The following criteria suggest such patients; fever (>38°C [100.4°F]) and leukocytosis, evidence suggesting cardiac tamponade, a large pericardial effusion, immunosuppressed state, a history of therapy with vitamin K antagonists, acute trauma, failure to respond within seven days to NSAID therapy, elevated cardiac troponin (myopericarditis). If patients do not have any high-risk criteria, they can be managed as outpatient [9].

Rheumatology Departments of Yaound, Central and General Hospitals, in Cameroon noticed similarity of presenting signs of pericarditis amongst those diagnosed with SLE. Beginning January 2001 a three-year trial occurred in observing SLE patients that presented with signs of pericardial involvement. A thorough analysis was done on these patients consisting of 22 females and 1 male with the age range of 13–65. The hospitals detected that 10 out of 23 patients (43.48%) presented with pericarditis. This study is substantial because it shows that these patients had the disease for over two years before it was shed to light. Findings of significance were pericardial rub in seven patients, and dyspnea in six patients. Echocardiography was performed to justify the diagnosis of SLE in every case. These tests also revealed abnormal repolarization in seven patients and a low voltage QRS complex on three occasions. Physicians on site administered corticosteroids to alleviate symptoms. Serological workup was also obtained and was indicative of SLE in all patients (i.e., anti-nuclear antibody, anti-double stranded antibody). Cardiomegaly was also observed in all patients on X-ray. Thereafter, four patients experienced relapses of pericarditis during ensuing flares of SLE [10].

**CONCLUSION**

The lifetime prevalence of some cardiac manifestation of systemic lupus erythematosus (SLE) is estimated to be 50%, and should be high on the differential of any presentation with chest pain or shortness of breath. It is crucial that physicians consider SLE as a diagnosis when new onset pericarditis occurs in African-American females of reproductive age. If diagnosed early, patients can receive appropriate treatment while reducing the chance of recurrent episodes and iatrogenic treatment.

**Author Contributions**

Gurneet Matharoo – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

William Tyler Whitmire – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Sam Sirotnikov – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Manoj Jagtiani – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Niket Sonpal – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

**Guarantor**

The corresponding author is the guarantor of submission.

**Conflict of Interest**

Authors declare no conflict of interest.

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