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Case Report

Rare Presentation of Adult-onset Still Disease with Myocarditis

Ann Sedeeq*, Akram Al-Salman, Hasan Sedeeq, Melak Idrees

Department of Internal Medicine, Hurley Medical Center, Michigan, USA

***Corresponding author:** Ann Sedeeq, Department of Internal Medicine, Hurley Medical Center, Michigan, USA

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Abstract

Adult-onset still disease is an inflammatory disorder that causes multiple system involvement. Patients usually present with fever, rash, arthritis, or arthralgia. Additionally, others might manifest with myalgia, pharyngitis, lymphadenopathy, and splenomegaly. One of the severe manifestations of adult onset still disease is cardiopulmonary disease including Pericarditis, pleural effusions, and transient pulmonary infiltrates. Rare presentation of adult-onset still's disease is myocarditis with dreaded outcomes including arrhythmias, heart failure, and cardiac tamponade. We are presenting a case report of a patient with no previous diagnosis of adult-onset still's disease who presented with chest pain fever and troponin elevations and was diagnosed with myocarditis secondary to adult-onset still's disease which is a rare presentation.

Keywords: Adult-onset still disease; Myocarditis; Chest pain

Abbreviations: EKG: Electrocardiogram; PCR: Polymerase Chain Reaction; COVID-19: Corona Virus disease 2019; ANA: Antinuclear Antibodies; HIV: Human Immunodeficiency Virus; CT: Computed Tomography; IV Intravenous; RF: Rheumatoid Factor; AOSD: Adult-Onset Still Disease

Case Presentation

A 30-year-old female with past medical history of protein C deficiency and pulmonary embolism who presented with chest pain. The patient developed substernal chest pain, which was intermittent, non-radiating, and lasted for 5-10 minutes starting the day before her presentation. According to the patient, chest pain worsened with breathing, lying flat and relieved by leaning forward.

Two sets of EKGs reported sinus tachycardia and nonspecific T-wave abnormalities. The initial lab results were significant for troponin was elevated at 1.30 and repeat troponin was 1.07. Additionally, white blood cells of 27 and D-dimer of more than 10. Other significant labs include C-reactive protein of 77.66 and Erythrocyte Sedimentation Rate (ESR) of 100, alkaline phosphatase 192-alanine aminotransferase of 69, normal aspartate aminotransferase, Thyroid function test was within normal limits and urine test negative for illicit drugs including cocaine. COVID-19 by PCR was negative. The patient also mentioned that she started to develop a rash that started 10 days before her presentation. The rash started on her arms then distributed slowly to her trunk and lower extremity, rash was itchy sometimes. She also developed multiple joint pain, swelling and tenderness of bilateral knee and ankle joints. She also had bilateral wrist and elbow joint pain. Joint pain and swelling started a few days before her presentation. In addition, she has associated low-grade fever, nausea and shortness of breath that increases when lying flat.

On arrival at the emergency department, Blood pressure was 102/65, pulse rate 145, respiratory rate 20, and SpO2 95% on room air. Due to history of Autoimmune disease in family (rheumatoid arthritis in mother); autoimmune panel sent, including ANA, which was positive, complements were with normal limits. Lyme antibodies, syphilis, hepatitis panel and HIV Antigen/antibody were normal. Urinalysis showed cloudy urine with 2+ protein with normal white and red cells.

Due to high suspicion for Pulmonary Embolism, CT chest with contrast was done which came back later negative for pulmonary embolism however was showing bibasilar lung atelectasis and Cardiomegaly.

Hepain was started while in the emergency department due to concerns for non-ST elevation MI. Cardiology team was consulted due to concerns for chest pain and elevated troponins. The Transthoracic Echocardiogram was done showing ejection fraction is 55% - 60% with moderate concentric left ventricular

hypertrophy and left ventricular diastolic dysfunction consistent with grade II pseudo normalization. No thrombus, mass or pericardial effusion seen. Patient underwent cardiac catheterization due to ongoing concern for chest pain, which eventually revealed normal epicardial coronaries. During her hospital stay, the patient continued to spike fevers up to 39.3 over several days. Multiple Blood cultures drawn were negative for any growth but due to persistent fever, the patient was started on broad-spectrum antibiotics with vancomycin and cefepime, which was later, broadened to Piperacillin/Tazobactam due to ongoing fever. However, she continued to spike fever despite broad-spectrum antibiotics. Given the high suspicion for adult onset still syndrome with very high ferritin 25000, IV antibiotics were stopped and steroid treatment with methylprednisolone started. Over the next few days, the rash started improving as well as joint pain improving, and the patient did not have further temperature spikes, of notice patient's chest, pain has subsided as well. Patient was discharged with a dose of prednisone 60 mg daily and will follow up with rheumatology outpatient for treatment optimization.

Discussion

Adult onset still disease is an inflammatory disorder that causes multiple system involvement, including joints, muscles, liver, spleen, cardiopulmonary, and gastrointestinal syndromes. Myocarditis as a presentation of adult onset still disease was found to be rare in previous literature review. Diagnosis of myocarditis in adult onset still disease is highly suggestive if we have symptoms including fever, chest pain, dyspnea, and tachycardia, labs/imaging finding including increased cardiac biomarkers, electrocardiogram (ECG), transthoracic echocardiography (ECHO), and/or cardiac magnetic resonance imaging (MRI) and good response with steroids and/or immunosuppressive medications. A case based review [1] done in 2020 which included patient with these finding are high suspicious for Adult onset still disease and most receive appropriate treatment including steroids and/or immunosuppressive medications to prevent dreaded cardiac outcomes including arrhythmias, heart failure, cardiac tamponade and cardiogenic shock.

Labs including ESR, C-reactive protein, very high ferritin levels, leukocytosis, elevated serum alanine and aspartate aminotransferases, elevated lactate dehydrogenase [2-4]. As for the immunological studies, Antinuclear Antibodies (ANA) and Rheumatoid Factor (RF) might be elevated in small percentages. Interestingly, IL-18 was found to be elevated in AOSD, the elevation appears to be more specific for AOSD than for other systemic rheumatic diseases [5].

A retrospective multicenter study of 28 patients All AOSD patients identified as fulfilling Yamagushi's [6] or Fautrel's [7] criteria were included in the study. 96 AOSD patients in this study: 28 (29%) had documented cardiac involvement (AOSD + C group) and 68 (71%) had no cardiac involvement (control group). Cardiac complications were observed at diagnosis in 89% of cases. These were pericarditis (n=17), tamponade (n=5), myocarditis (n=5) and non-infectious endocarditis (n=1). Corticosteroids were effective with or without methotrexate in 71% of patients but not in severe involvement as myocarditis or tamponade [8].

One case series showed that in these 24 myocarditis complicated AOSD cases, myocarditis occurred early and was present at Adult onset still disease in 54% of the cases. Steroids alone were effective in 50% of patients with myocarditis. Intravenous immunoglobulins, methotrexate, and tumor necrosis factor-α-blockers were also prescribed and often found effective. Only one death with cardiogenic shock was reported [9].

Conclusion

High index of suspicion must be suspected of adult-onset still disease with cardiac complications like myocarditis especially in young patients presenting with chest pain, fever and myalgia/arthralgia and rash with cardiac biomarkers elevation after ruling out ischemic events. Prompt treatment with steroid and/or immunosuppressive regimen should be initiated to prevent cardiac complications including arrhythmia, cardiac tamponade, and cardiogenic shock.

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