

Recurrent Rheumatic Fever with Multiple Organ Dysfunction Syndrome in a 19-year-old Man: Case Report and Literature Review

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BACKGROUND

- Acute rheumatic fever (RF) is a known nonsuppurative complication of tonsillopharyngeal infection with group A Streptococcus (GAS).
- Signs and symptoms manifest within two to three weeks following pharyngitis with a constellation of symptoms including arthritis, carditis, Sydenham chorea, erythema marginatum and subcutaneous nodules.
- Recurrent RF can occur in incompletely treated acute RF with an incidence rate between 15 and 34%.
- Rheumatic heart disease is the most common sequelae in affected patients.
- Here we present a rare case of multiple organ dysfunction syndrome in a young male patient with recurrent acute rheumatic fever.

CASE DISCRIPTION

- A 19-year-old man with a PMH of acute RF at age 7 years presented with 4-5 days of fevers, chills, malaise, sore throat, dyspnea, orthopnea, and polyarthralgia of ankles, knees, wrists, and shoulders. This was accompanied by trouble walking which was so severe that he had to crawl around the house to get from bed to bath.
- On physical exam, his vitals included temp 38.7, HR 105, RR 37, with findings notable for jaundice, scleral icterus, friction rub, 3/6 systolic murmur at the apex, 2/6 diastolic murmur, decreased breath sounds, multiple tender joints.
- Diagnostic investigations were notable for multiple lab derangements which worsened over the course of his hospitalization with the peaks/nadirs as noted here: ESR 125, CRP 179.6, ASO 2865, AST 1876, ALT 611, total bilirubin 6.1, direct bilirubin 1.1, WBC 15.9, Hgb 6.6, creatinine 4.61, BUN 78, LDH 1348, INR 3.8, and BNP 1620. Urinalysis revealed hematuria, proteinuria, and glucosuria, with urine sediments of muddy brown casts consistent with ATN. Autoimmune and microbiological studies were all negative.
- Echocardiogram revealed preserved EF, mild mitral stenosis, moderate mitral regurgitation, moderate to severe tricuspid regurgitation, mild aortic regurgitation, moderate pulmonary hypertension and a moderate circumferential pericardial effusion without tamponade.
- ECG revealed prolong PR interval. CXR showed cardiomegaly. Abdominal US showed fatty infiltrates but no biliary dysfunction.
- He developed acute hypoxic respiratory failure and hypotension requiring mechanical ventilation and vasopressors in the ICU.
- Recommended treatments included high dose aspirin, diuretics, colchicine for pericardial effusion/pericarditis, and IM benzathine penicillin G which was also to be continued prophylactically.

CLINICAL & DIAGNOSTIC IMAGES

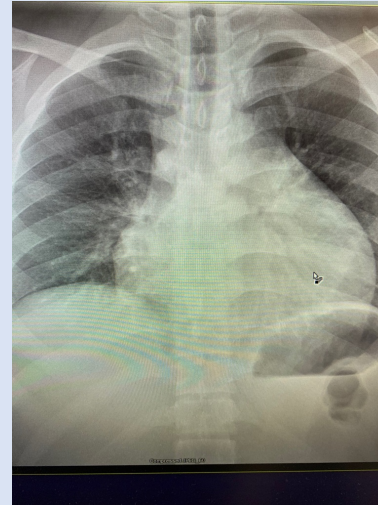


Figure 1: Cardiomegaly on chest x-ray

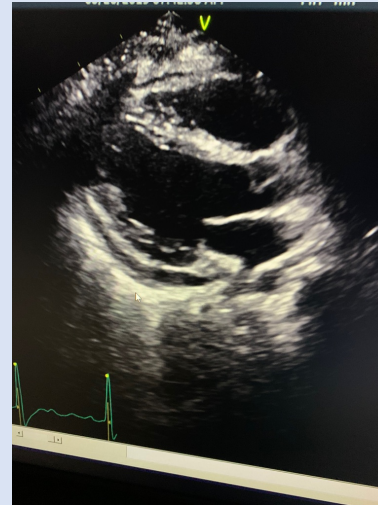


Figure 2: Echo complete showing a small-moderate circumferential pericardial effusion

DISCUSSION

- Early detection of GAS infection with rapid streptococcus antigen test and appropriate treatment has made acute rheumatic fever relatively uncommon.
- The multisystem organ failure reported here represents a rare manifestation of recurrent acute RF in a previously sensitized patient.
- This patient experienced progressive decline in musculoskeletal, cardiac, pulmonary, renal, hepatobiliary, hematologic function, all of which ultimately led to a state of multisystem shock.
- The decline in his respiratory status is presumed secondary to rheumatic pneumonitis since no infectious etiology was found and his cardiac function was not severe enough to cause pulmonary edema. His renal and hepatobiliary manifestations are presumed multifactorial stemming from cardiogenic shock in the setting of his disease complications.
- Overall, this rare manifestation of recurrent acute RF likely represents an exaggerated autoimmune response to the inciting pathogen by way of molecular mimicry.
- This phenomenon would benefit from additional investigations into its pathophysiology and clinical manifestations.

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