Thinking Twice: Recurrence of Acute Rheumatic Fever in an Unprophylaxed Patient with History of Carditis

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ACUTE RHEUMATIC FEVER

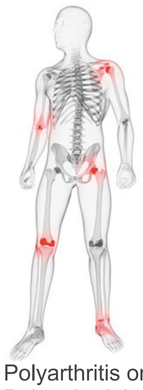
- Acute rheumatic fever (ARF) is an inflammatory disease involving cardiac and joint tissue that develops due to inadequately treated group A streptococcal (GAS) infection
- In cases of carditis, secondary prophylaxis requires monthly injections of penicillin G for a duration of ten years
- Recurrence of ARF is associated with history of carditis and lack of adherence to secondary prophylaxis

PENICILLIN FOR SECONDARY PROPHYLAXIS

- Patients with well-documented histories of rheumatic fever require continuous antibiotic prophylaxis for secondary prevention of recurrent ARF
- The agent of choice is penicillin G benzathine 1.2 million U IM every four weeks (AHA evidence rating 1A)
  - Per guidelines, this therapy is particularly beneficial in cases where adherence to oral therapy is likely to be poor
  - Less supported alternative therapies include penicillin V potassium (1B), sulfadizine(1B), macrolides (1C), or azalides (1C)
- The duration of secondary prophylaxis varies based on clinical presentation and age of onset (1C)
  - Rheumatic fever with carditis and residual valvular disease requires a duration of ten years or until age 40 (whichever is longer)
  - Rheumatic fever with carditis but no residual valvular disease requires a duration of ten years or until age 21 (whichever is longer)
  - Rheumatic fever without carditis requires a duration of five years or until age 21 (whichever is longer)

A CASE OF RECURRENCE

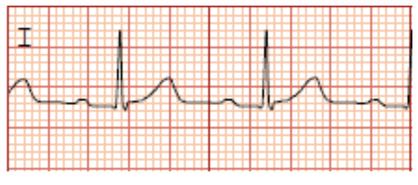
- A 47-year-old man Micronesian with history of acute rheumatic fever (ARF) complicated by carditis with inconsistent medical follow-up and nonadherence to secondary prophylaxis presented to the Emergency Department with acute-on-chronic, diffuse polyarthralgias
- History of **throat culture positive** for group A *Streptococcus*
- **ASO Ab titer** elevated to 1066 IU/ml
- **DNAse B Ab** titer elevated to 1560 IU/ml
- Found to meet:
  - **Two major Jones Criteria**
    - Polyarthritis/polyarthralgia
    - Sydenham chorea
  - **All minor Jones Criteria**
    - Polyarthralgias
    - Hyperpyrexia
    - Elevated ESR/CRP
    - Prolonged PR interval on ECG



Polyarthritis or Polyarthralgias

Sydenham chorea

Fever ≥38.5° C
ESR ≥60 mm
CRP ≥3.0 mg/dl



PR interval prolongation

- Given his persistent symptoms in the absence of alternative diagnoses, his well-documented history of ARF, and his history of nonadherence to secondary prophylactic therapy, the patient was treated based on standard-of-care with a single dose of penicillin G 1.2 million units IM
- He was discharged with a plan for monthly injections for the recommended duration of ten years of secondary prophylaxis; the patient was lost to follow-up shortly after discharge

REVISED JONES CRITERIA FOR PRESUMPTIVE DIAGNOSIS OF RECURRENT ARF

Presumptive diagnosis of recurrent ARF requires:

1. History of ARF or rheumatic heart disease
2. Evidence of preceditreptococcal infection, AND
3. Either
   a) Two major,
   b) One major and two minor, or
   c) Three minor manifestations

- Level C, Class IIb evidence (consensus of expert opinion based on clinical experience or case studies; usefulness is not well established)

CASE HIGHLIGHTS

- This case highlights:
  - The importance of secondary prophylaxis in prevention of recurrent ARF
  - The importance of recognizing and managing the once common syndrome of recurrent ARF
  - The need for further clinical research to aid in evidence-based decision-making in cases with high clinical suspicion for recurrent ARF
  - The roles of therapeutic adherence and access-to-care in achieving successful outcomes in the management of chronic illnesses

References:
1. Gerber MA, Baltimore RS, Eaton CB, et al. Prevention of rheumatic fever and diagnosis and treatment of acute Streptococcal pharyngitis: a scientific statement from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on Cardiovascular Disease in the Young, the Interdisciplinary Council on Functional Genomics and Translational Biology, and the Interdisciplinary Council on Quality of Care and Outcomes Research: endorsed by the American Academy of Pediatrics. Circulation. 2009;119(11):1547
2. Gewitz MH, Baltimore RS, Tani LY, et al. Revision of the Jones Criteria for the Diagnosis of Acute Rheumatic Fever in the Era of Doppler Echocardiography: A Scientific Statement From the American Heart Association. *Circulation* 2015;Apr 23.
3. https://www.masterfile.com/