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VALVULAR HEART DISEASE

CASE REPORT: CLINICAL CASE

Progression of Rheumatic Heart Disease

A Case of Failed Secondary Prevention

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ABSTRACT

Recurrent acute rheumatic fever in patients who have completed secondary antibiotic prophylaxis is exceedingly rare. This paper presents a case of recurrent acute rheumatic fever with progressive rheumatic heart disease in a middle-aged woman requiring emergent surgical valvular repair and an additional course of secondary antibiotic prophylaxis. (JACC Case Rep. 2024;29:102620) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 57-year-old woman presented with sore throat, palpitations, dyspnea on exertion, orthopnea, high daily fevers, migratory polyarthralgia, and rash for the past 1 month. Her vitals showed a temperature of 37.3 °C, heart rate of 102 beats/min, respiratory rate of 24 breaths/min, blood pressure of 126/71 mm Hg, and 99% oxygen saturation on room air. Physical examination was remarkable for an uncomfortable appearing woman with swelling and tenderness to the right wrist with an overlying rash. Electrocardiogram showed normal sinus rhythm, left ventricular hypertrophy, possible left atrial enlargement, and frequent premature atrial complexes. Complete blood count, complete metabolic panel, thyroid function studies, troponin level, and pro-B type natriuretic peptide were within normal limits. Erythrocyte sedimentation rate was elevated at 106 mm/h (reference <29 mm/h), and C-reactive protein was elevated at 71.0 mg/L (reference <4.9 mg/L). Bedside point-of-care ultrasound revealed grossly normal left ventricular ejection fraction and severe mitral regurgitation with thickened leaflets. Chest radiograph showed pulmonary congestion and bilateral pleural effusions. She was started on intravenous diuretics and admitted to the cardiovascular intensive care unit for management and work-up of severe mitral regurgitation.

LEARNING OBJECTIVES

- To consider recurrent acute rheumatic fever in adults with rheumatic heart disease who have completed secondary antibiotic prophylaxis.
- An additional 10 years of antibiotic prophylaxis may be warranted to prevent further recurrent episodes.

PAST MEDICAL HISTORY

The patient has a history of hypertension, breast cancer with metastasis to the lungs, and rheumatic heart disease (RHD).

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ABBREVIATIONS AND ACRONYMS

ARF = acute rheumatic fever

GAS = group A Streptococcus

RHD = rheumatic heart disease

Her breast cancer was treated with radiation therapy, tamoxifen, and robotic video-assisted thoracic surgery, and is currently in remission on anastrozole with no evidence of disease.

The patient immigrated from South Korea 23 years ago. During early childhood, she was diagnosed with acute rheumatic fever (ARF) that progressed to RHD. At that time, she was treated with intravenous penicillin to eradicate group A Streptococcus (GAS) infection and then subsequently completed 10 years of monthly penicillin G injections until 2006.

DIFFERENTIAL DIAGNOSIS

At the time of admission, the etiology of the patient's constellation of symptoms was unclear. Differential diagnosis included mitral regurgitation due to infective endocarditis, heart failure with preserved ejection fraction due to chronic valvular disease, septic arthritis, and undifferentiated rheumatologic disease. Acute coronary syndrome was thought to be less likely due to normal troponin levels, nonischemic electrocardiogram findings, and normal wall motion on bedside cardiac ultrasound.

INVESTIGATIONS

Formal transthoracic echocardiogram showed normal left ventricular ejection fraction of 61%, severely dilated left atrium, and severe mitral regurgitation with thickened leaflets, and restricted opening suggestive of possible rheumatic etiology (Video 1). Subsequent transesophageal echocardiogram was performed for further characterization that revealed rheumatic appearing aortic and mitral valves, both with moderate to severe regurgitation. Antistreptolysin O titer was markedly elevated at 1,063 IU/mL (reference <200 IU/mL). Antinuclear antibody, rheumatoid factor, and anticyclic citrullinated peptide antibody returned within normal limits. Blood cultures and tuberculosis acid fast culture and polymerase chain reaction were negative.

MANAGEMENT

The patient received 2 million units of intramuscular penicillin G for eradication of GAS colonization. Due to findings of severe aortic and mitral valve pathology, the patient was taken for surgical aortic and mitral valve replacement. The procedure was without complications, and the patient successfully received a St. Jude Masters (Abbott) mechanical aortic and mitral valve. She was discharged on hospital day 11. On discharge, she was started on monthly intramuscular penicillin G for a planned 10-year course and furosemide for heart failure with preserved ejection fraction.

DISCUSSION

This report demonstrates the importance of including recurrent ARF in the list of differential diagnosis when working-up patients with valvular disease paired with a constellation of nonspecific symptoms. ARF is a sequelae of GAS infection characterized by carditis, Sydenham chorea, migratory polyarthritis, erythema marginatum, and subcutaneous nodules.[1,2] RHD is a severe sequela of ARF that occurs in up to 50% of patients who present with carditis during diagnosis of ARF.[3] Secondary prophylaxis is recommended for patients diagnosed with ARF or RHD to prevent recurrent ARF, which commonly occurs in the first couple of years after the initial episode.[4] Prior studies have shown recurrent ARF to increase with noncompliance to secondary prophylaxis, with <90% compliance with secondary prophylaxis adherence being associated with 12.9 times increase in odds of AR recurrence.[5] Currently, there are only observational data suggesting efficacy of secondary antibiotic prophylaxis in preventing recurrent ARF.[6-9]

We report a case of recurrent ARF in a woman nearing her sixth decade of life that completed secondary antibiotic prophylaxis nearly 20 years ago. The patient was diagnosed with recurrent ARF after satisfying 2 major and 1 minor criteria based on the revised Jones criteria. The mean age of recurrent ARF is 23.4 years old with an SD of 9.9 years,[10] making recurrent ARF diagnosis in this patient more than 3 SDs above the mean. Preventing repeat episodes of ARF is imperative in reducing the risk of developing or worsening RHD.[2] Given the rarity of recurrent ARF in late adulthood and in low-income countries, there are limited literature and guidelines for the management and prevention of additional future recurrences. The multidisciplinary team approached the treatment of this complex case by first eradicating GAS infection with high-dose intramuscular penicillin G, and then emergently repairing the valvular disease. An additional 10 years of intramuscular penicillin G treatment was also started because, although both aortic and mitral valve were replaced, future recurrent episodes of ARF can still affect other native valves, perivalvular tissue, and chordae.

This case illustrates the importance of keeping recurrent ARF on the differential diagnosis in older

adult patients with RHD who have completed secondary antibiotic prophylactic therapy. Additionally, this case also suggests the risk of recurrent ARF may extend into middle adulthood and an additional course of secondary prophylaxis should be considered to prevent ARF recurrence and progression of RHD.

FOLLOW-UP

The patient was seen in cardiology outpatient clinic 1 and 4 months after hospital discharge. At the 1-month visit, she reported minimal leg swelling, 1 pillow orthopnea, and occasional episodes of paroxysmal nocturnal dyspnea. Diuretic dose was up-titrated and she was started on metoprolol tartrate. At the 4-month visit, the patient was asymptomatic and repeat interval transthoracic echocardiogram showed left ventricular ejection fraction of 65% with normal functioning aortic and mitral bileaflet prosthesis with trace physiological regurgitation (Video 2). The patient continues to be compliant with prescribed medications and secondary antibiotic prophylaxis.

CONCLUSIONS

Recurrent ARF is rare but possible in older adults with previously diagnosed RHD despite full adherence to secondary antibiotic prophylaxis. Recognition of this phenomena by clinicians can expedite timely diagnosis and treatment. Currently, there are limited literature and guidelines as to further management and prevention of additional future recurrences. As in this case, we recommend an additional 10 years of secondary prophylaxis to prevent further episodes of recurrence and progression of RHD.

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KEY WORDS acute rheumatic fever, group A streptococcus, recurrent acute rheumatic fever, rheumatic heart disease, valvular heart disease

APPENDIX For supplemental videos, please see the online version of this paper.