

Rheumatic heart disease in Tennessee: An overlooked diagnosis

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Abstract

Rheumatic heart disease, already a major burden in low- and middle-income countries, is becoming an emerging problem in high-income countries. Although acute rheumatic fever and rheumatic heart disease have almost been eradicated in areas with established economies, the emergence of this problem may be attributable to the migration from low-income to high-income settings. Between 2010 and 2012, we diagnosed a cluster of rheumatic heart disease cases in children from the Middle Tennessee area. The goal of this report is to increase awareness among clinicians as the incidence and prevalence of acute rheumatic fever remain relatively significant in large US metropolitan areas. Although acute rheumatic fever is seasonal, a high suspicion index may lead to the early diagnosis and prevention of its cardiac complications. Furthermore, screening procedures may be recommended for populations at risk for rheumatic heart disease in endemic areas, and active surveillance with echocardiography-based screening might become very important.

Keywords

Rheumatic heart disease, high-income communities, overlooked diagnosis

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Introduction

Acute rheumatic fever (ARF) is a multi-system disease.¹ Rheumatic heart disease (RHD) occurs in up to 80% of the patients with ARF. The mechanism of how ARF leads to RHD in select patient groups may include specific group A streptococcus (GAS) strains (serotypes 1, 3, 5, 6, 7, 18, 19, and 24), inherited host susceptibility, and pathologic autoimmunity. ARF is more common in younger patients (90%) than in older adolescents and adults.²⁻⁴

Although more than 80% of the ARF cases occur in low- and middle-income communities (LMICs), there has been resurgence in high-income communities (HIC). The purpose of this article is to review a cluster of RHD cases found in children who were seen in various Middle Tennessee clinics between 2010 and 2012. The goal of this report is to increase awareness among clinicians, as the incidence and prevalence of ARF remains relatively significant in large US metropolitan areas.

Patient reports

Case 1

A 7-year-old Caucasian female, a known asthmatic, was evaluated with a 1-week history of an increasing cough and

exhibited no improvement with beta-agonist bronchodilators. She had a new grade III/VI systolic regurgitant murmur with a mid-systolic click noted at the apex, radiating to the left sternal border and the base of the heart, as well as diffuse expiratory wheezing and crepitation. The rest of her physical examination was unremarkable except for a body mass index at the fifth percentile and an elevated blood pressure of 115/74 mmHg. Her medical history included positive streptococcus pharyngitis tests at ages 5 and 6 years, which were adequately treated with antibiotics. Her echocardiogram report showed an enlarged left ventricle with decreased systolic function. There was also mild to moderate mitral valve regurgitation. She was treated with intramuscular benzathine penicillin G. She was also treated with a blood pressure-lowering agent, enalapril, to address

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Table 1. Laboratory, EKG, and echocardiographic findings in ARF/RHD cases.

Cases	ASO (IU/ mL, 0–200)	Anti-DNaseB (IU/mL, <170 or EU, <20)	ESR (mm) first hour	CRP (0– 0.79°mg/ dL)	GAS rapid antigen or culture	EKG	Echocardiogram
1	586	433 EU	1	0.4	Both negative	Normal	Moderate degree of mitral valve regurgitation, and enlargements of both the left ventricle and the left atrium
2	2238	37 EU	62	6.54	Both negative	First-degree heart block	Mild aortic and mitral valve regurgitations
3	393.7	1920	6	0.06	Both negative	Border-line increase in QT interval	Normal
4	654	1020	10	0.4	Positive culture	Multiple PACs	Moderate mitral regurgitation associated with thickened mitral valve leaflets and prolapsing of the anterior leaflet
5	981	697	Not done	44.8	Both negative	Normal	A trace of aortic valve insufficiency with moderate mitral valve insufficiency

EKG: electrocardiogram; RHD: rheumatic heart disease; ARF: acute rheumatic fever; EU: enzyme unit; GAS: group A streptococcus; ESR: erythrocyte sedimentation rate; PAC: premature atrial contraction; ASO: antistreptolysin O; CRP: C-reactive protein.

her mitral valve regurgitation. Secondary prophylaxis with intramuscular benzathine penicillin G was initiated on an every 4-week regimen. Prophylaxis for sub-acute bacterial endocarditis (SBE) for surgical and dental procedures was recommended.⁵

Case 2

A 10-year-old Hispanic male in his usual good state of health was seen in a follow-up visit for a presumed viral illness associated with intermittent fevers, body aches, joint stiffness, dizziness, and paleness. His medical history was marked by one documented episode of an appropriately treated streptococcal pharyngitis 3 years prior to the current presentation. He had a grade II/VI systolic murmur at the base and apex of the heart and I/VI diastolic murmurs at the left and right upper sternal borders. Treatment and prophylaxis were appropriately instituted.⁵

Case 3

A 4¼-year-old Caucasian female presented with a 1 year history of recurrent fevers, sore throat, joint pain, and joint swelling, suggesting both arthralgia and arthritis. Her medical history was significant for recurrent episodes of streptococcal pharyngitis, which were adequately treated with amoxicillin or benzathine penicillin injections. On examination, she had a typical rash of multiple flat, painful, bluish-purple, and red bumps over both anterior knees consistent with erythema nodosum. On auscultation of her heart, she had a grade II/VI systolic murmur at the left sternal border. A presumptive diagnosis of ARF was made based on two major criteria (erythema nodosum and arthritis) and one minor criterion (fever).⁵ The patient's family, however, refused secondary antibiotics prophylaxis treatments but *did* agree to

continue close follow-ups with pediatric cardiology on a 6- to 12-month basis.

Case 4

On a routine well-child exam, an 8½-year-old African-American female with mild developmental delay was noted to have an irregular heart rhythm, a new II–III/VI systolic regurgitant heart murmur, an active precordium, and an erythematous throat. Her echocardiogram showed mitral valve prolapse and mitral valve regurgitation. The degree of mitral valve prolapse seen on her echocardiogram (Table 1), however, did not clinically justify the degree of mitral valve regurgitation. She was appropriately treated, and a secondary prophylaxis was instituted with intramuscular benzathine penicillin injections. The pediatric cardiology recommendation included an angiotensin-converting enzyme inhibitor (enalapril) for her moderate mitral valve regurgitation.

Case 5

A 9-year-old Hispanic female presented with a new grade II/VI systolic murmur in the setting of a routine physical examination. Her medical history was remarkable for one episode of pharyngitis and arthralgia for which she received amoxicillin 3 months prior to her clinic visit. She was treated with an intramuscular injection of benzathine penicillin, and a secondary prophylaxis of the same was continued.

Discussion

Although ARF and RHD have been almost eradicated in areas with established economies, the emergence of ARF may be attributable partly to migration.⁶ The prevalent hypotheses may include a high influx of individuals

harboring “rheumatogenic” strains of the bacteria. These concerns also involve frequent global travelers, including active military personnel. Cases 1, 2, and 3 were seen in a suburban pediatric clinic with a high influx of the dependents from active duty military personnel housed at a nearby military base. Case 4 was a Hispanic girl seen in a clinic located in an urban area of Middle Tennessee. Although our report concentrated on only five cases from a small region of Tennessee, the burden of ARF may not be negligible in the United States. We hypothesize rheumatic fever cases in Middle Tennessee areas may be on the rise, warranting further epidemiological studies.

In this report, the diagnostic criteria for ARF and RHD are based primarily on clinical presentations (Jones criteria) and the echocardiogram results. Although case 1 did not strictly meet Jones minor criteria, a diagnosis of RHD was made based on echocardiogram results and evidence of past GAS infection. Therefore, it is important to note that a diagnosis of RHD can be made, or the suspicion of RHD can be raised based on the echocardiogram results, despite absence of the clinical Jones Criteria. With the exception of case 4, none of the cases had evidence of a positive *Streptococcus pyogenes* pharyngitis at the time of presentation. Laboratory test results for antistreptolysin O (ASO) titers and the anti-DNaseB were often delayed. Thus, heightened suspicion for early diagnosis is critical, as the condition can lead to cardiac complications that can be crippling to pediatric patients.

The prevalence of RHD in US populations is reported at less than 0.05 per 100,000, with rare outbreaks reported in Tennessee in the 1960s.⁷ The reported cases in this article may suggest that the actual incidence and prevalence of rheumatic fever may be underestimated in the United States, warranting further research. Based on our clinical cases, it may be recommended that high-risk population, including returning military personnel from deployment (or on ongoing assignments) should promptly seek medical attention in case of symptoms suggestive of streptococcus pharyngitis. Cost-benefit for asymptomatic individuals is prohibitive. Identification of GAS colonization or infection with “rheumatogenic” strains of the bacteria (In asymptomatic family members of index cases), and echocardiographic screening might prove beneficial in patients with newly detected heart murmurs.⁸ These tests are undoubtedly simple and cost-effective. Additional studies might be needed to assess the epidemiology of ARF in at risk middle-income communities and HICs, which may help in the development and implementation of a continuous quality improvement strategy.⁹ This will further aid in the implementation of comprehensive strategies for the prevention

and/or early detection of GAS infection in other vulnerable populations as well at risk for rheumatic fevers and rheumatic heart disease.

Declaration of conflicting interests

The authors declare that there is no conflict of interest.

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