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Acute Rheumatic Fever Presenting with Transient Complete Heart Block: A Rare Cardiac Manifestation in an Adult Patient

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Abstract

Background: Acute rheumatic fever (ARF) is an autoimmune reaction to Group A β-hemolytic streptococcal infection, primarily affecting the heart, joints, and connective tissues. While conduction abnormalities in ARF are uncommon, complete atrioventricular (AV) block is an exceptionally rare manifestation.

Case Presentation: We report a case of a 33-year-old breastfeeding woman who presented with fever, sore throat, and myalgia. While under observation in the emergency department, she experienced a transient episode of syncope, during which telemetry captured a complete AV block with a 12-second ventricular pause. Diagnostic workup confirmed acute rheumatic fever based on the revised Jones criteria. Other causes of conduction abnormalities, including myocarditis, sarcoidosis, and autoimmune disorders, were systematically ruled out. The patient was treated with Amoxicillin-Clavulanate and nonsteroidal anti-inflammatory drugs (NSAIDs), leading to complete resolution of the conduction disturbance. A permanent pacemaker was not required.

Conclusion: Transient complete heart block is a rare but significant cardiac manifestation of ARF. Clinicians should consider ARF in young patients presenting with unexplained conduction abnormalities. Early recognition and appropriate management are crucial for preventing long-term complications.

Keywords: Acute rheumatic fever; Complete heart block; Atrioventricular block; Group A streptococcus; Carditis; Transient heart block; Rheumatic carditis

Introduction

Acute Rheumatic fever (ARF) is a delayed autoimmune reaction to group A β-hemolytic streptococcal pharyngitis (GAS). It triggers widespread inflammation in the body, which can affect multiple organs, including the joints, nervous system, skin, subcutaneous tissue and most importantly the heart [1]. The inflammatory response associated with ARF can cause serious complications, particularly in the cardiovascular system, leading to long-term consequences such as rheumatic heart disease [2].

However, while ARF is classically considered a delayed immune response, typically manifesting 2-4 weeks after the initial infection, there have been very few reports suggesting that symptoms, including carditis, may develop during the acute phase of GAS infection [3]. Rheumatic carditis, one of the most concerning manifestations of ARF, can affect the heart's conduction system, leading to various degrees of atrioventricular (AV) block. Studies have shown

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that first-degree AV block is the most frequently observed conduction abnormality, occurring in approximately 72.5% of cases. Less frequently, second-degree AV block occurs in 2.6% of cases, while complete AV block is rare, reported in only 0.6% of cases according to previous reports [4]. We report a case of transient high degree AV block in a young adult presenting with acute rheumatic fever in the setting of GAS pharyngitis.

Case Presentation

A 33-year-old female a history of allergic necrotizing venulitis presented to the emergency department with a five-day history of fever, sore throat, and generalized myalgia. She denied any cough, shortness of breath, chest pain, palpitations, or prior episodes of syncope. There was no recent history of travel, sick contacts, or recent infections. She was not on any regular medications and had no known drug allergies.

On arrival, she was febrile with a temperature of 38.2°C, a heart rate of 88 beats per minute, blood pressure of 120/75 mmHg, respiratory rate of 16 breaths per minute, and oxygen saturation of 98% on room air. Physical examination revealed mild pharyngeal erythema without exudates or tonsillar hypertrophy. Cardiovascular examination showed normal heart sounds with no murmurs, rubs, or gallops. Pulmonary, abdominal, and neurological examinations were unremarkable. There were no signs of arthritis, subcutaneous nodules, or erythema marginatum.

While under observation in the emergency department, the patient suddenly became unresponsive for approximately 10 seconds without any preceding symptoms such as dizziness or palpitations. Telemetry captured a transient complete atrioventricular (AV) block with a ventricular pause of 12 seconds (Figure 1). She spontaneously regained consciousness without confusion or postictal symptoms. A 12-lead ECG performed immediately after the episode showed normal rhythm with sinus tachycardia, no PR prolongation or QRS widening (Figure 2).

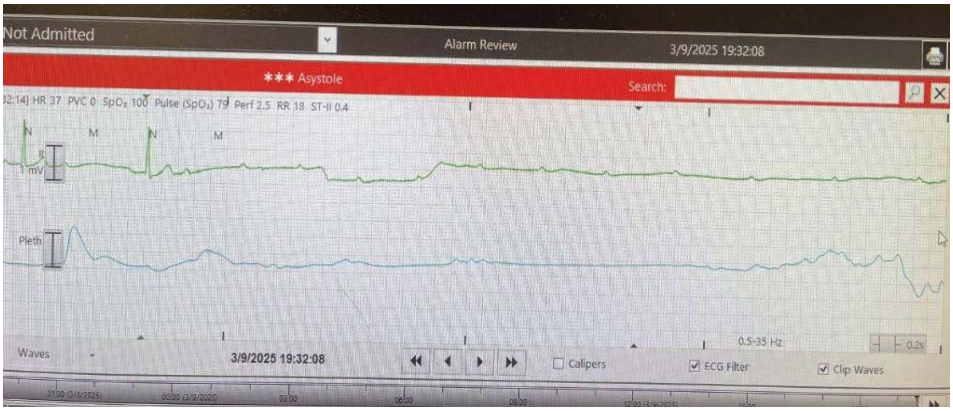

Figure 1: Telemetry showing a transient complete atrioventricular block with a ventricular pause of 12 seconds.

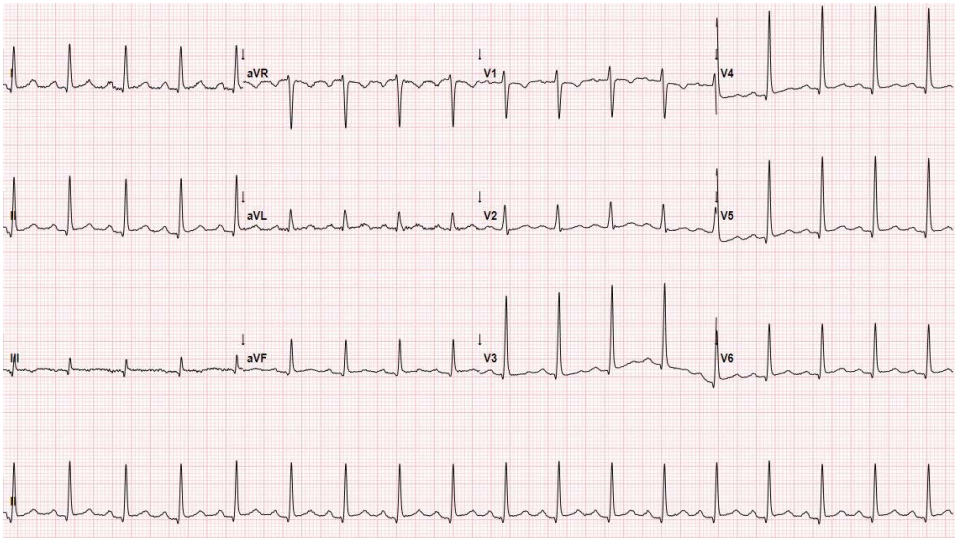

Figure 2: Electrocardiogram showing sinus tachycardia at rate of 105 beats/min, PR interval 175 msec and a QRS interval of 95 msec.

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Initial laboratory investigations, including a complete blood count (CBC), renal function, liver enzymes, and electrolytes, were within normal limits. However, further testing revealed a positive throat swab for Group A Streptococcus, an anti-streptolysin O (ASO) titer of 330 IU/mL (Normal <220 IU/mL) and a positive rheumatoid factor of 23.5 IU/mL (Normal <14.0 IU/mL). Inflammatory markers including C-reactive protein (CRP) of 112.5 mg/L (Normal <2.5 mg/mL) and erythrocyte sedimentation rate (ESR) of 62 mm/hr (Normal <20 mm/hr) were significantly elevated. Troponin levels remained negative.

She was admitted to the coronary care unit (CCU) for continuous cardiac monitoring and further evaluation. Transthoracic echocardiography demonstrated normal left ventricular function with no evidence of valvular disease, pericardial effusion. Cardiac MRI did not reveal any signs of myocarditis, fibrosis, or infiltrative disease.

Given her young age and transient conduction disturbance, further investigations were conducted to rule out other potential causes of conduction abnormalities in young adults. Cardiac sarcoidosis was considered but deemed unlikely due to a negative cardiac MRI, normal serum angiotensin-converting enzyme (ACE) levels and chest computed tomography with no evidence of bilateral hilar adenopathy or lung lesions. Additionally, systemic autoimmune diseases were ruled out with negative antinuclear antibody (ANA), Sjögren's antibody panel (SSA/Ro, SSB/La) and other relevant serological tests including double stranded DNA antibodies, C3 and C4 complement levels.

During her stay in the CCU, continuous telemetry monitoring did not reveal any further conduction abnormalities, arrhythmias, or recurrent AV block.

Based on the revised Jones criteria, the presence of carditis evidenced by the transient AV block qualified as a major criterion. Additionally, fever and elevated inflammatory markers fulfilled two minor criteria, and the positive ASO titer and throat swab for Strep A provided supporting evidence of recent streptococcal infection. Therefore, she was diagnosed with acute rheumatic fever.

She was started on Amoxicillin-Clavulanate for streptococcal eradication and non-steroidal anti-inflammatory. Given the transient nature of the conduction disturbance, a permanent pacemaker was not indicated.

Upon discharge, she was prescribed monthly penicillin for long-term secondary prophylaxis to prevent the recurrence of rheumatic fever. She was scheduled for a close follow-up with her cardiologist in two weeks.

Discussion

According to the revised Jones criteria, our patient met the diagnostic criteria for acute rheumatic fever (ARF) with one major and two minor criteria [5]. Other potential causes of conduction system abnormalities in young patients were systematically ruled out. Myocarditis was excluded based on negative troponin levels and a normal cardiac MRI. Sarcoidosis was considered unlikely given the absence of abnormalities on cardiac MRI, chest CT, and normal ACE levels. Autoimmune disorders were also ruled out based on a negative autoimmune laboratory panel. Given these findings, ARF was determined to be the most likely cause of the patient's transient complete heart block.

To our knowledge, 38 cases of atrioventricular (AV) block in the setting of ARF had been reported by Umapathy et al. [6] and Oba et al. [11], with two additional cases documented since then, bringing the total to 40 [7,8]. Among these, 26 patients were adolescents or children. AV block in ARF typically occurs during the first attack and is associated with minimal valvular involvement, though a case of recurrence has been reported [6]. Stokes-Adams attacks which are syncopal episodes due to transient AV block were documented in 12 of these cases, including our patient, whose episode in the emergency department ultimately led to the diagnosis of ARF [9].

The exact mechanism of AV block in ARF remains unclear, but increased vagal tone and immune-mediated effects have been proposed. Transient first-, second-, and third-degree AV blocks in ARF do not necessarily indicate generalized carditis and typically do not lead to long-term cardiac sequelae [10]. However, a single case required permanent pacemaker implantation due to prolonged and severe inflammation [11]. Management of ARF-related conduction abnormalities follows the same principles as general ARF treatment, with most cases responding well to nonsteroidal anti-inflammatory drugs (NSAIDs) [12]. While corticosteroids have been suggested as beneficial [13], strong evidence is lacking. In cases of advanced symptomatic AV block, temporary transvenous pacemaker placement should be considered. Additionally, antibiotic therapy for Group A streptococcal infection is recommended, regardless of the presence of active pharyngitis. It has been reported that conduction system involvement is considered a form of carditis, necessitating secondary prophylaxis with daily penicillin for at least 10 years after the initial attack or until the age of 40, whichever is longer [14]. Our patient met the criteria for long-term prophylaxis and was counseled accordingly.

We report the first case of a 33-year-old breastfeeding woman presenting with an initial attack of ARF complicated by transient complete heart block. Following treatment with a course of Augmentin and symptomatic management with NSAIDs, the AV block did not recur, and temporary pacemaker placement was not required.

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Conclusion

Transient complete heart block is a rare but important cardiac manifestation of ARF, particularly in adults. Our case highlights the importance of considering ARF in young patients presenting with unexplained conduction abnormalities, even in the absence of significant valvular disease. A thorough diagnostic workup is essential to exclude alternative causes, and early recognition allows for appropriate management with anti-inflammatory therapy and secondary prophylaxis. Long-term follow-up is necessary to monitor for potential recurrence and valvular complications.

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