Acute Rheumatic Carditis: A Rare Cause for Reversible Complete Heart Block

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Abstract

A previously healthy 18-year-old man presented to the emergency department with weakness, fever, and joint pains and was found to have complete heart block with transient asystole requiring urgent transvenous pacing. After further workup, the patient was found to have complete heart block secondary to acute rheumatic carditis. The conduction system recovered in a step-wise fashion following treatment with Penicillin, and high dose Aspirin, without the need for permanent pacemaker placement. This case illustrates that acute rheumatic carditis, although rare, can present with advanced conduction system involvement, which is reversible if treatment is initiated.

Introduction

The differential diagnosis for complete heart block in a young patient is limited and includes congenital complete heart block, and acquired complete heart block secondary to multiple etiologies including: trauma, infections, and inflammatory conditions such as myocarditis, sarcoidosis, and auto-immune disorders. Muscular dystrophies are also known to be associated with AV block in young patients, particularly the Myotonic sub-type.[1]

Cardiac conduction system involvement has been reported as a feature of acute rheumatic carditis, with involvement ranging from first degree atrio-ventricular (AV) block (72.5%), second degree AV block (2.6%) and rarely complete AV block (0.6%) based on previous case reports.[2] Acute rheumatic fever (ARF) is an auto-immune sequela that occurs two to four weeks following group A streptococcal pharyngitis and involves multiple organ systems: Musculoskeletal (migratory arthritis), cardiovascular (carditis and valvulitis and conduction system disorders), central nervous system (chorea), and skin (erythema marginatum, and subcutaneous nodules). Worldwide, there are an estimated 250,000 - 470,000 new cases of rheumatic fever each year.[3,4] Most cases of ARF occur in the developing world and are relatively uncommon in the United States,[4] where the incidence of ARF is 2 to 14 cases per 100,000.[5] ARF is more common in Hawai'i with an estimated incidence of 9.5 cases per 100,000.[6] It has been suggested that this may be attributable to unique and potentially more virulent strains of group A streptococci affecting the inhabitants of the Hawaiian islands.[7]

We present a case of complete heart block secondary to acute rheumatic carditis, basic diagnosis, and management principles.

Case Description

An 18-year-old man with no significant past medical history presented to a community hospital complaining of nausea, vomiting, and weakness for five days. His symptoms began about 24 hours after consuming fish at a family picnic. He also reported history of sore throat over the past two to three weeks prior to presentation. Initial vital signs revealed a blood pressure of 126/66 mmHg, heart rate of 48 beats per minute, respiratory rate of 18 breaths per minute, and a temperature of 37.1 °C (98.8 °F). Mild tenderness and swelling was noted in the right ankle. No skin rash was noted. None of his family members experienced similar symptoms and he had no sick contacts. He did not have any travel history over the past year prior to presentation and denied any history of recent animal contacts or insect bites.

An electrocardiogram (ECG) in the emergency room revealed complete heart block with a wide complex rhythm with a left bundle branch morphology suggestive of a ventricular escape rhythm at approximately 50 beats per minute (Figure 1). While performing a second ECG the patient was noted to have a transient loss of his escape rhythm and subsequent ventricular asystole indicative of cardiac arrest (Figure 2). Shortly after a temporary transvenous pacemaker was placed, the patient was transferred to our facility for further evaluation and consideration for permanent pacemaker placement.

Initial laboratory data revealed sodium level: 128 mEq/L (normal range, 133-145 mEq/L), potassium level: 3.4 mEq/L (3.3-5.1 mEq/L), chloride level: 93 mEq/L (95-108 mEq/L), bicarbonate level: 25 mEq/L (21-30 mEq/L), calcium level: 9.2 mg/dL (8.3-10.5 mg/dL), creatinine: 0.9 mg/dL (0.6-1.4 mg/dL). White blood cell count: 11.4 X 10^3/ uL (3.84 X 10^3- 9.84 X 10^3 /uL), hemoglobin: 12.8 g/dL (13.7-17.5 g/dL), platelet count: 312 X 10^3/ uL (151 X 10^3 - 424 X 10^3 /uL), troponin level: <0.02 ng/mL (< 0.05 ng/mL), erythrocyte sedimentation rate (ESR): 127 mm/hour (0-15 mm/hour), C-reactive protein (CRP): 229 mg/L (0-10 mg/L), thyroid stimulating hormone (TSH): 1.55 uIU/mL (0.27-4.2 uIU/mL). Chest X-ray on presentation was unremarkable.

Given his antecedent consumption of fish and the presence of gastrointestinal symptoms on presentation (nausea and vomiting), as well as reports of Ciguatera poisoning in the Pacific Islands, this diagnosis was initially considered but was quickly ruled out following confirmation the fish consumed was tuna (not a reef fish associated with Ciguatera poisoning) and absence of similar symptoms in family members who also consumed the fish.

On hospital day two the patient reported right knee pain and was found to have fever at 39.2 °C (102.5 °F). Physical examination revealed mild swelling, redness, and tenderness in right knee. Given his history of sore throat, subsequent development of fever, and the findings suggestive of migratory arthritis the diagnosis of ARF was considered. An Anti Streptolysin O (ASO) titer was performed and found to be elevated at 441 IU/mL (normal <200 IU/mL). An echocardiogram demonstrated

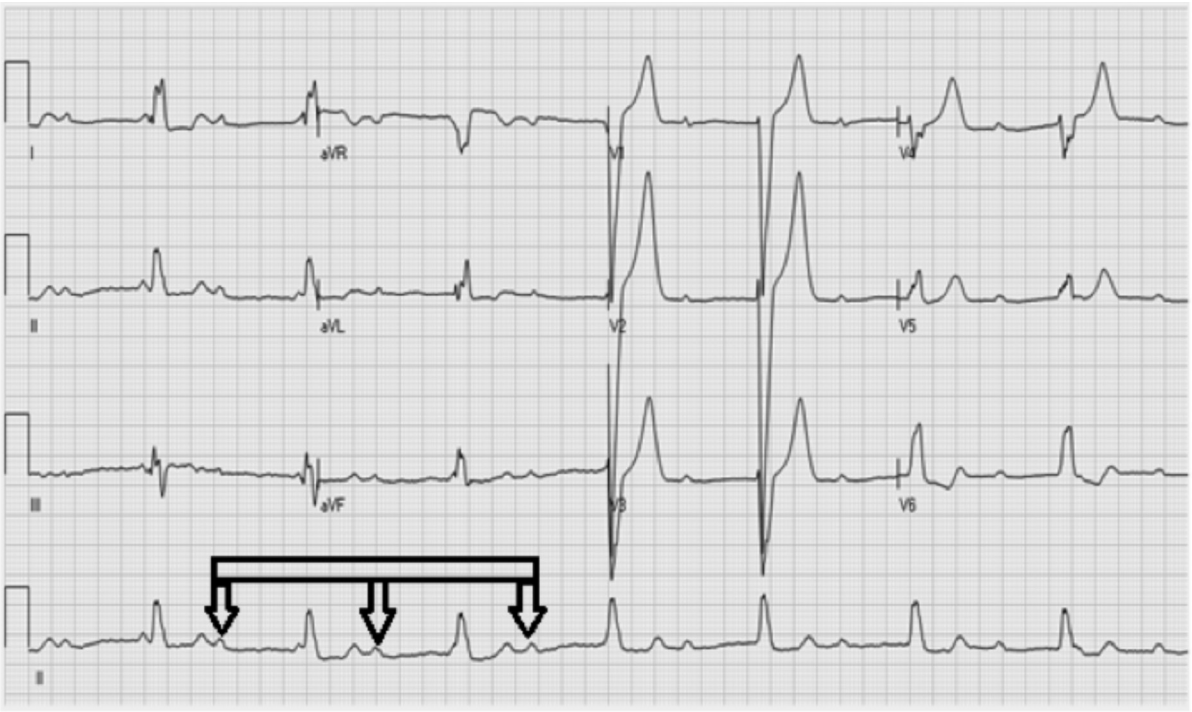

Figure 1. Initial ECG demonstrating complete heart block and a wide complex ventricular escape rhythm with a left bundle branch morphology (arrows showing atrial activity that is completely dissociated from the ventricular activity).

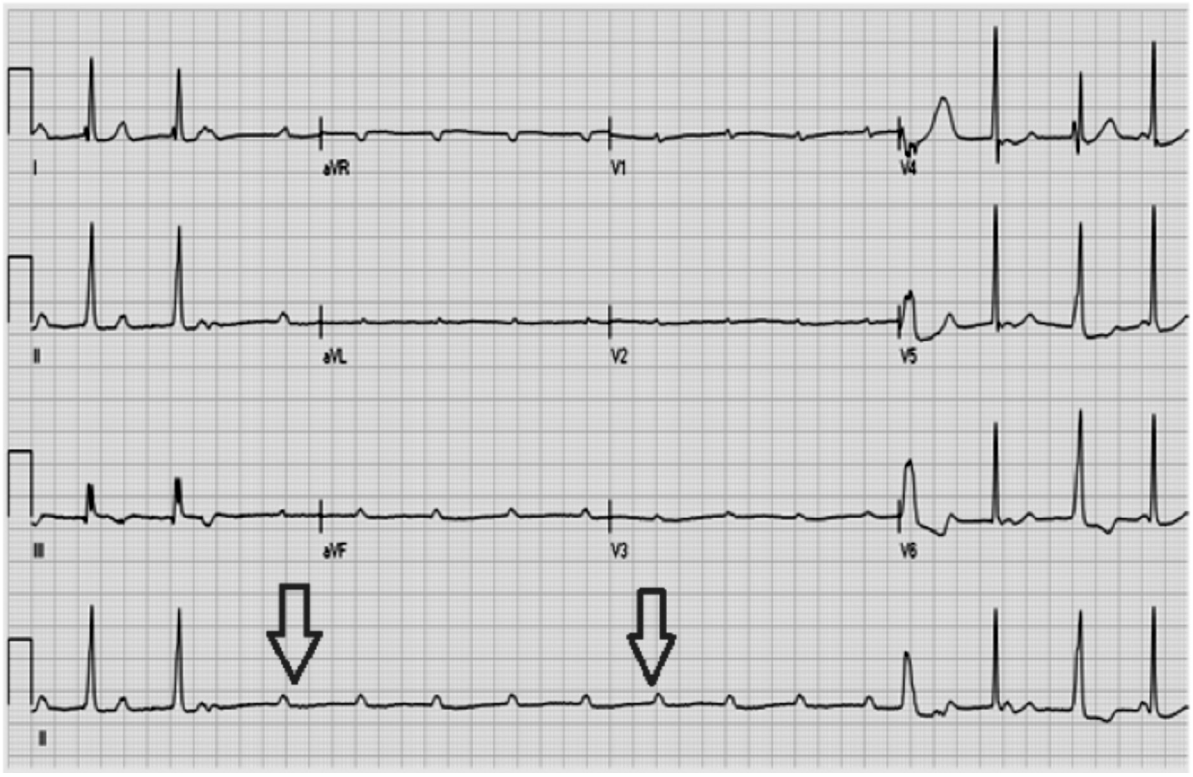

Figure 2. Transient ventricular asystole (cardiac arrest) shortly after initial ECG (arrows showing P waves documenting atrial activity with no subsequent ventricular conduction).

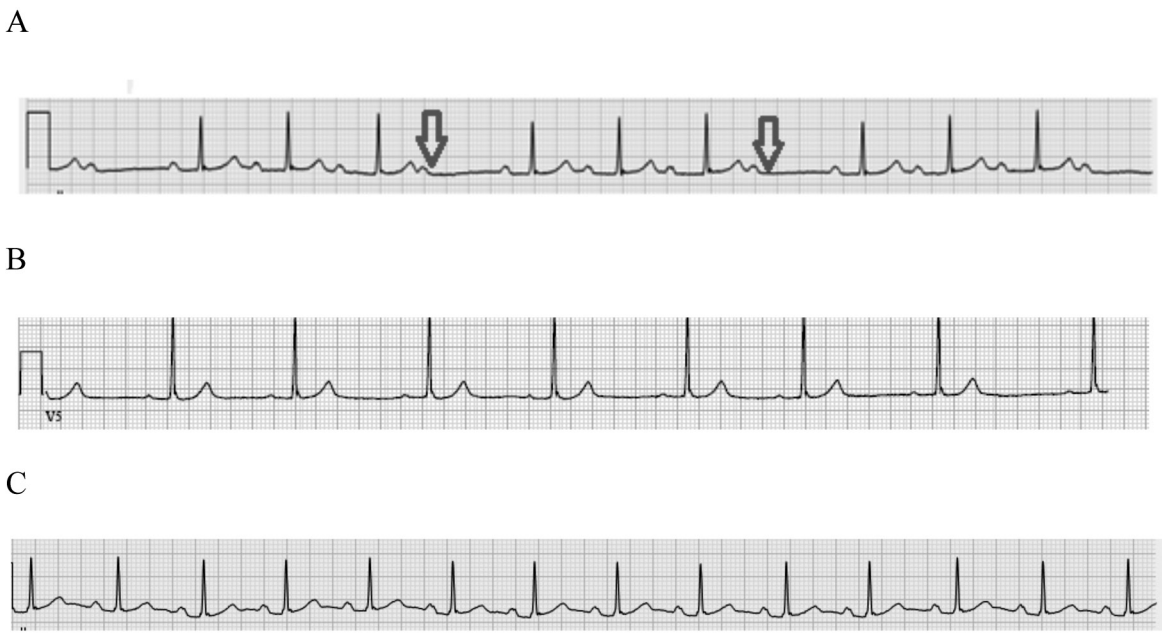

Figure 3. Rhythm strips following treatment for acute rheumatic fever: (A) Arrows showing second degree type I (Wenkebach) AV block, (B) Prolonged PR (230 msec), (C) Near-normalization of PR (200 msec).

preserved left ventricular ejection fraction with a trace aortic regurgitation. Our patient had two major components (carditis and migratory arthritis) as well as multiple minor components (fever, elevated ESR, and CRP) of the Jones criteria.[8] Thus, the diagnosis of ARF was established.

Treatment with Penicillin and high dose Aspirin was initiated. Shortly thereafter, the patient's AV conduction improved in a stepwise fashion with Wenkebach periodicity, followed by prolonged AV conduction, and then near-normalization of the PR interval (Figure 3). The patient was then placed on secondary prophylaxis with monthly intramuscular Penicillin. He was discharged on high dose Aspirin with instructions for tapering and was then seen in the clinic one month later with improvement of his symptoms.

Discussion

Our patient's history, presentation, as well as diagnostic workup was consistent with acute rheumatic carditis. He did not have any recent travel history to areas endemic with Lyme disease and he had no associated skin lesions; thus, this diagnosis was excluded. He did not have associated muscle weakness or family history to suggest muscular dystrophies. He also did not have respiratory, renal, or skin features to suggest sarcoidosis or other autoimmune diseases. Myocarditis was also less likely with negative troponin markers and no history of chest pain.

Ciguatera poisoning is a toxin-related illness resulting from the consumption of infected reef fish in endemic regions. It is commonly seen in the Pacific Islands and the Caribbean. It is estimated that 10,000-50,000 people per year who live in or visit tropical and subtropical areas suffer from Ciguatera Poisoning.[9] In Hawai'i, there is a total of 3 to 69 cases per year, averaging 28.5 total cases per year from 2002-2011,[10] but the true incidence is difficult to ascertain due to under-reporting. Poisoning typically causes gastrointestinal symptoms (nausea, vomiting, cramping, and diarrhea), neurological symptoms (paresthesia), and cardiovascular symptoms (hypotension and severe bradycardia and AV block).[9,11] Ciguatera Poisoning was initially considered given his recent ingestion of fish but was excluded given the type of fish consumed and the absence of similar symptoms in his family members.

To our knowledge there have only been a total of 25 published case reports of AV block in the setting of acute rheumatic fever, five of which were adult patients.[12] Of these, only four were from the United States. Filberbaum and colleagues described cases of ARF associated AV block as early as 1945 with no clear description regarding the severity of conduction involvement.[13] In subsequent years there have been sporadic reports of AV block associated with ARF in the United States.[14-16] Of the 25 reported cases, 15 were found to be reversible following treatment of ARF and one case persisted at 3 months.[12] Carano and colleagues presented a case of a 14 year old boy who had a very similar presentation and history as our case.[12] That patient first presented with acute rheumatic fever and carditis. The patient's echocardiographic study also documented minimal aortic regurgitation which, like our case, suggests that typical valvular involvement in cases of rheumatic carditis may not necessarily be present and features of carditis may only be the conduction system involvement. Lenox and colleagues presented a case of complete heart block in acute rheumatic carditis.[15] This was the patient's second attack of rheumatic fever, which

suggests that this type of conduction system involvement does not necessarily happen with first attacks of rheumatic carditis.

Treatment of conduction system involvement with ARF resulting carditis follows the same basic principles of ARF treatment. Consideration should be given to the use of a temporary transvenous pacemaker placement in patients with advanced symptomatic AV involvement. Treatment for Group A streptococcal infection should be initiated regardless of the presence of active pharyngitis. Treatment includes either oral Penicillin for 10 days or single dose of intramuscular Penicillin.[17] High dose Aspirin (4-8 g/24 hours) is the most effective anti-inflammatory agent for active arthritis. The role of systemic corticosteroids is less clear and may not provide additional benefit compared to Aspirin monotherapy.[18] Carano and colleagues' patient received steroid as part of his treatment regimen,[12] but our patient showed favorable response to Aspirin and Penicillin only. Secondary prevention for recurrent ARF should also be provided after treatment during the acute phase. Daily Penicillin treatment or monthly intramuscular injections should be used. For patients with a history of carditis (including conduction system involvement) the treatment duration is at least 10 years after the initial attack or until age 40 years, whichever is longer. It has been suggested that patients with a history of severe carditis should receive lifelong prophylaxis.[17]

Conclusion

We present a rare case of complete heart block associated with acute rheumatic carditis. Although rare, this diagnosis should be considered in patients with complete heart block particularly when it is associated with other features of ARF. As evidenced by this and several other cases, conduction disorders associated with ARF often resolve following appropriate treatment without the need for permanent pacemaker placement.

Conflict of Interest

None of the authors identify a conflict of interest.

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