Various Rhythmic Disturbance in Acute Rheumatic Fever: A Two-case Report

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Research Article

Keywords: Acute Rheumatic Fever, Complete atrioventricular block, Supraventricular tachycardia, Arrhythmias, Case Report

Posted Date: May 10th, 2022

DOI: https://doi.org/10.21203/rs.3.rs-269960/v1

Abstract

Acute rheumatic fever (ARF) remains a major health problem worldwide, especially among the endemic countries. Different kinds of rhythm and conduction abnormalities may be seen during the course of the disease. Although the first-degree atrioventricular block is the most common disturbance reported in ARF, other advanced heart block may be an exceptionally rare manifestation. We reported two cases of ARF; one presented with first- and second-degree AV block (Mobitz II), and the other one presented with supraventricular tachycardia followed by unstable bradycardia due to complete atrioventricular block and required temporary pacemaker. Both cases completely recovered with anti-inflammatory drug regimen for ARF.

Introduction

Acute rheumatic fever (ARF) is a serious health concern affecting certain populations around the world. Although the overall age-standardized death rate is decreasing throughout the past decades, mortality rate among the endemic countries is still as high as 400 per 100,000 population approximately[1, 4]. Patients with ARF may manifest in a variety of presentation, most commonly the cardiac involvement defined as carditis. Cardiac rhythm conduction abnormality is one of the prevalent clinical manifestations among patients with ARF, including the prolongation of PR interval which is considered a minor diagnostic criterion according to the Revised Jones' criteria in 2015[2]. Other rhythm disturbances such as premature ventricular contractions (PVCs) or junctional tachycardia are also reported as manifestations of ARF carditis while second- or third-degree atrioventricular (AV) block are rare. We reported two patients diagnosed with acute rheumatic fever who presented with second- and third-degree AV block. One of our cases was found to have unstable bradycardia and required a temporary pacemaker insertion.

Case 1

A 13-year-old boy suffering from obesity and hypertension was included in a weight reduction program at Naresuan University Hospital during school break. During the admission, he had pain on his right knee and ankle, and tightness on his chest. Four days prior to admission, the patient had fever and sore throat, and was given a course of antibiotics as prescribed by a physician in a private clinic.

The patient's vital signs showed temperature of 39.3°c, HR 45 beats per minute (bpm) with full and regular rhythm, and BP 113/62 mmHg. His cardiovascular exam revealed a grade II/VI systolic ejection murmur at the upper left parasternal border without any heaves or thrills and mild swelling of the right ankle with limited range of motion due to pain. Other examination was unremarkable. An electrocardiogram (ECG) showed first degree AV block with second degree AV block (Mobitz II) (Fig. 1).

Laboratory test results revealed white blood cells count (WBC) of 17,340 cell/cu.mm, C-reactive protein (CRP) of 169 mg/L and erythrocyte sedimentation rate (ESR) of 98 mm/hr. The evidence of streptococcal infection was confirmed by Anti-DNaseB of 987 U/mL, antistreptolysin O (ASO) titer of 1,650 IU/mL and a

positive throat swab culture for group A Streptococcus. Echocardiography showed mild mitral and aortic valve regurgitation with LVEF of 78%. He was diagnosed with acute rheumatic fever because of two major components (carditis and polyarthritis) and two minor components (fever and elevated acute phase reactants) of the modified Jones criteria, corroborated by a culture-positive test for group A Streptococcal infection. His hemodynamic status became stable, however, his ECG showed 2nd degree AV block with the slowest HR of 40 bpm. He was given high doses of aspirin and penicillin V. He was responding well to medications. His ECG rhythm reverted to normal sinus rhythm on the seventh day of treatment and was fully recovered without any noticeable valvular regurgitation during the follow-up serial echocardiogram.

Case 2

A previously healthy 13-year-old boy was admitted to a rural hospital due to low-grade fever for two days and two episodes of syncope. He denied any previous episodes of syncope or palpitation. On admission, his physical examination revealed a temperature of 38 °c, BP 135/60 mmHg, and HR 155 bpm with full pulse. The ECG showed supraventricular tachycardia (SVT) with rate of 150 bpm (Fig. 2). After two doses of adenosine (6 and 12 mg, respectively), ECG recorded a sinus rhythm rate of 130 bpm. The patient was then referred to a provincial hospital.
Upon admission at the second hospital, the patient developed bradycardia, and his ECG showed complete AV block with HR of 15 to 30 bpm (Fig. 3) along with cardiopulmonary compromise. Patient was transferred to our hospital and internal temporary pacemaker was installed with VVI mode HR of 80 bpm..

The patient's laboratory test results revealed WBC of 12,700/mL, ESR of 76 mm/hr, CRP of 13.3 mg/dL, ASO titer elevated at 1,120 IU/mL, and a throat swab culture with no pathogenic bacterial growth. His ECG showed mild MR, minimal pericardial effusion and normal LVEF (64%). The diagnosis of acute rheumatic fever was made. Since the patient had severe carditis with complete AV block, an anti-inflammatory agent, prednisolone (2mg/kg/day), was initiated along with penicillin V. Two days after treatment, ECG revealed first-degree AV block at a rate of 65 bpm and normal sinus rhythm at a rate of 80 bpm with stable vital signs. Pacemaker was turned off on the fifth day of treatment. High-dose of aspirin (80 mg/kg/day) was administered after a 2-week prednisolone regimen. He was discharged after 22 days of hospital stay with echocardiogram showed no evidence of persistent valvular dysfunction.

Discussion

Acute rheumatic fever (ARF) is a multi-systemic inflammation as a consequence of group A streptococcal (GAS) infection. Several studies on the pathogenesis of ARF pointed to the molecular mimicry between GAS antigens called M-protein or carbohydrate antigen (N-acetyl-beta-D-glucosamine) that produce antibodies, and human protein, such as cardiac myosin or laminin. This mechanism results in humoral immune response injury and T-cell recruitment leading to granulomatous inflammation and Aschoff body formation, which are the hallmark of ARF[3, 4]. The organs mostly involve in ARF are the heart, joints and subcutaneous tissues.

Acute rheumatic fever (ARF) is diagnosed based on the Modified Jones' criteria, which classifies the clinical manifestations and laboratory results into major and minor criteria[2]. Major characteristics of ARF include joints involvement (mono- or polyarthritis or polyarthralgia), chorea, skin manifestation (subcutaneous nodules or erythema marginatum) and carditis[4].

Carditis is a manifestation secondary to ARF, also considered to be pancarditis, which refers to the involvement of pericardium, myocardium and endocardium[5]. The endocardium is the most common manifestation of carditis that results in chordal elongation with prolapse of the leaflet coaptation leading to permanent valvular damage[6]. Myocardial involvement with conduction pathway abnormalities is considered a minimal myocardial dysfunction that may not be detected clinically [7]. Several studies reported rhythm conduction defects in ARF for decades, and 40–60% of patients exhibit a prolonged PR interval (first-degree atrioventricular (AV) block [8–10]. Other rhythm disturbances included accelerated junctional tachycardia, premature contractions, ventricular tachycardia, Torsade de pointes due to prolonged QT interval, complete bundle branch block and rarely second or third-degree AV block[5, 10–12]. The incidence of second-degree AV block (Mobitz I) was reported as 1.5%-2.6% and third-degree or complete AV block as 0.6%-4.6% in different studies[7–8]. Mobitz II was reported in one patient on the 24-hour Holter monitoring but not on the standard 12-lead ECG[16]. A presentation with supraventricular tachycardia (SVT) followed by complete AV block, the same as the second case in our study, has never been reported. Most of the conduction abnormalities are transient. Sevket B et al. reported some short-term conduction abnormalities do not appear on the standard 12-lead ECG but reported on 24-hour Holter monitoring, including premature contraction and complete heart block. The 24-hour rhythm Holter recording is suggested to be part of the routine ARF screening to detect rhythm abnormality and prevent bradyarrhythmia[13].

Rhythmic disturbances in ARF have no correlation with valvular involvement[13–17]. Several studies reported that the occurrence of conduction defect is not a specific sign of carditis[7, 8, 10, 12–14]. Although the exact mechanism of conduction defects in ARF is unknown, they were thought to be due to localized myocardial inflammation involving AV node or due to vasculitis involving the AV nodal arteries as the consequence of immune-mediated mechanism in ARF[13]. As mentioned above, the molecular mimicry in ARF refers to a reaction between glycoprotein of GAS antigen and host tissue, especially the cardiac myosin in the heart valves resulting in valvulitis. The atrioventricular node has a very low content of glycoprotein but rich in glycogen, which is believed to be a substrate for the rheumatic process as it is implicated by the presence of short PR interval in patients with glycogen storage disease[9].

Most of the ARF rhythm conduction defects are transient, self-limiting with complete resolution at the end of the acute stage of the disease following anti-inflammatory medications. Some of the patients whose symptoms persisted or reported an Adams-Stokes attack may require a transient pacemaker[15]. One patient reported a complete heart block presented with Adam-Stokes attack had temporary pacemaker for four days[11]. The first patient in our study had a second-degree AV block that was resolved with anti-inflammatory drugs in the first week. In comparison, the second patient had a supraventricular

tachycardia and a complete AV block that resulted in unstable bradycardia requiring a temporary pacemaker for five days along with anti-inflammatory drugs.

Conclusion

Besides PR prolongation, various rhythm disturbances were frequently found during the acute phase of ARF. We recommend diagnostic work-up and ECG monitoring in patients with ARF whether or not they have signs of carditis. Cardiac dysrhythmias are usually transient and completely resolve after several days or weeks with anti-inflammatory medication. The temporary pacemaker should be considered in patients with unstable hemodynamic status.

Declarations

We wish to confirm that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome. No funding was received for this work. We further confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript. Written consent to publish potentially identifying information, such as details or the case, was obtained from their legal guardians. We attest that all authors contributed significantly to the creation of this manuscript, each having fulfilled criteria as established by the ICMJE.

Authors' contributions

CS analyzed patients' history and investigations. WJ performed the interpretation of the electrocardiogram and echocardiogram. CS was a major contributor in writing the manuscript. All authors read and approved the final manuscript.

Funding sources: None

Ethical approval: Naresuan University Institutional Review Board (IRB no. P3-0119/2563)

No financial or non-financial benefits have been received or will be received from any party related directly or indirectly to the subject of this article.

The authors have each completed the International Committee of Medical Journal Editors Form for uniform Disclosure of Potential Conflicts of Interest. No authors have any potential conflict of interest to disclose potential conflict of interest.

The manuscript has been read and approved by all the authors.

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Figures

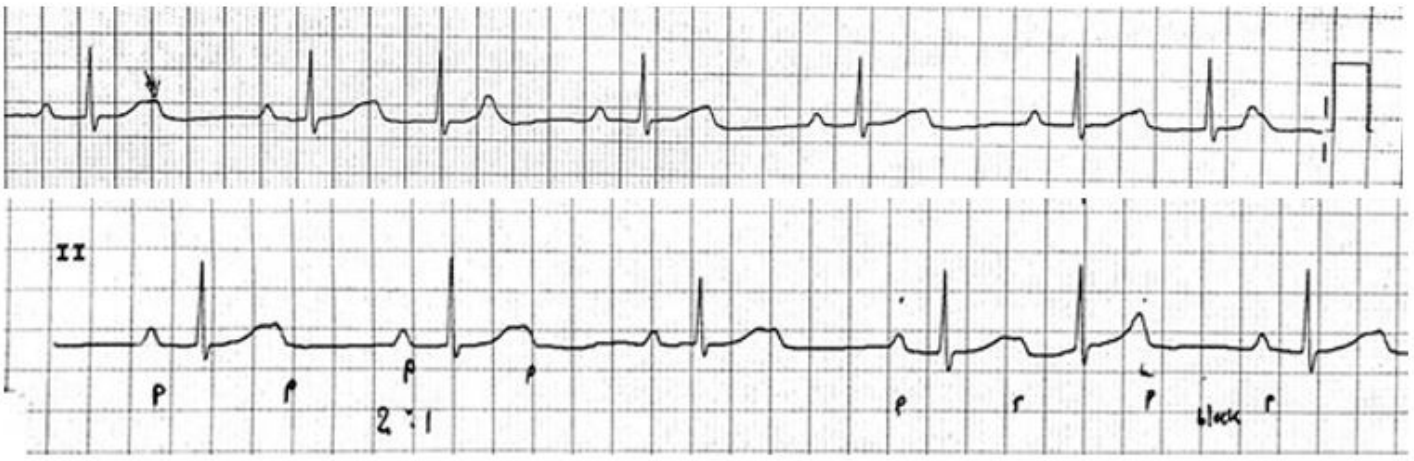

Figure 1

ECG indicating second degree AV block (Mobitz II) with 2:1 conduction

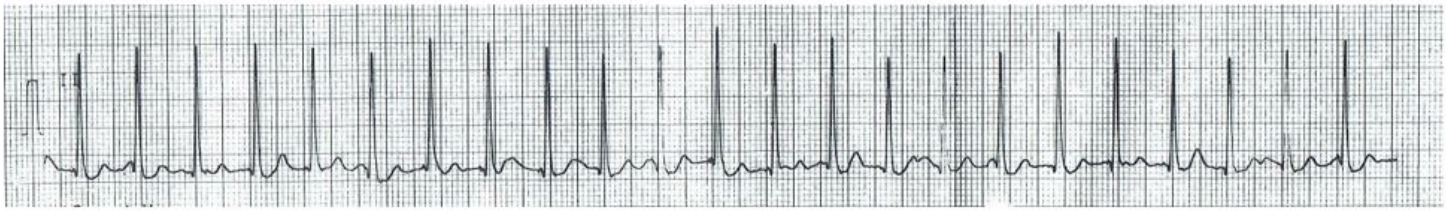

Figure 2

ECG showing supraventricular tachycardia (rate 150 bpm)

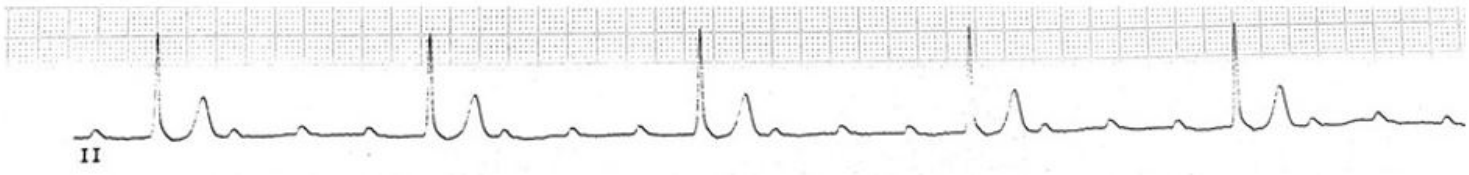

Figure 3

ECG showing complete atrioventricular (AV) block

Supplementary Files

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