CASE REPORT

Acute rheumatic fever presenting as complete heart block: report of an adolescent case and review of literature

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SUMMARY

A 14-year-old boy suffering from chronic rheumatic heart disease came to the emergency department with recurrent episodes of presyncope and syncope. He was found to have complete heart block (CHB) and required temporary pacemaker insertion. Further workup revealed that CHB was secondary to acute rheumatic carditis. His atrioventricular (AV) conduction abnormalities recovered in a stepwise fashion over 5 days while he was being treated with corticosteroids, without the need for permanent pacemaker insertion. This case illustrates that acute rheumatic carditis can rarely present with advanced AV conduction block, which may be reversible.

BACKGROUND

Rheumatic fever has been considered as a delayed autoimmune sequelae to group A β-haemolytic streptococcal pharyngitis, although the exact pathogenesis remains unclear. It elicits a generalised inflammatory process involving the heart, joints, central nervous system, subcutaneous tissue and skin. Except for involvement of the heart (as carditis), other manifestations of rheumatic fever are self-limiting and do not leave any significant residua. Cardiac conduction system abnormalities reported in acute rheumatic fever ranges from first-degree atrioventricular (AV) block to second-degree AV block and rarely complete heart block (CHB).[1] Here, we report a case of CHB which occurred during recurrence of acute rheumatic fever in a patient with chronic rheumatic heart disease.

CASE PRESENTATION

A 14-year-old adolescent boy suffering from chronic rheumatic heart disease with advanced valvular lesions (severe aortic regurgitation and mild mitral regurgitation) presented to the emergency department with a history of 1 week of fever and recurrent episodes of presyncope and syncope. He had no recent history of joint pains/swelling or skin rash. He had prior history of documented acute rheumatic fever 1 year ago and poor compliance to secondary penicillin prophylaxis. At presentation, his heart rate was 55 beats per minute and blood pressure was 160/50 mm Hg. He had fever (38°C, 100.4°F) but no respiratory distress (respiratory rate of 18 breaths per minute). He had peripheral signs of aortic regurgitation (water hammer pulse, bisferiens pulse, visible pulsations of carotid artery, pistol shot femorals, to and fro bruit over femoral artery and positive Hill sign of 25 mm Hg). Auscultation revealed high-pitched blowing early diastolic murmur extending into later half of the diastole at the second aortic area suggestive of severe aortic regurgitation. An ECG showed a heart rate of 55 beats per minute and CHB with wide complex rhythm of left bundle branch block (LBBB) morphology suggestive of a ventricular escape rhythm (figure 1). Patient was admitted and temporary transvenous pacemaker was placed via femoral access.

INVESTIGATIONS

Initial laboratory data revealed a haemoglobin of 11.4 g/dL, leucocytosis ($19x10^9$/L), erythrocyte sedimentation rate (ESR) of 20 mm/hour and raised C reactive protein (CRP) of 148.5 mg/L. Troponin I assay by rapid kit test was negative. Antistreptolysin O (ASO) titre was found to be elevated at 582 IU/mL (normal <200 IU/mL). His renal function tests were within normal limits and potassium level was 4.9 mEq/L. His chest X-ray showed mild cardiomegaly with enlargement of left ventricle and left atrium. Echocardiography showed severe aortic regurgitation, mild mitral regurgitation and dilated left ventricle with normal function. Mitral valve also showed multiple nodules and there was minimal pericardial effusion, both features suggesting acute rheumatic carditis.

TREATMENT

In view of one major (carditis) plus two minor components (fever, elevated CRP) of revised Jones criteria, along with confirmatory evidence of previous streptococcal infection in the form of high ASO titres, the diagnosis of acute rheumatic fever was confirmed. Since the patient had significant valvular lesions secondary to carditis, treatment with corticosteroids was initiated in addition to other supportive measures.

OUTCOME AND FOLLOW-UP

His CHB recovered to first-degree heart block (PR interval 360 ms, figure 2) on the third day of admission followed by occurrence of ectopic atrial rhythm with PR interval of 220 ms on the fifth day (figure 3). He was advised secondary prophylaxis with long-acting intramuscular benzathine benzyl penicillin in the dose of 1.2 million units 3 weekly. He was discharged on corticosteroids with instructions for its tapering over the next 8 weeks. Periodic

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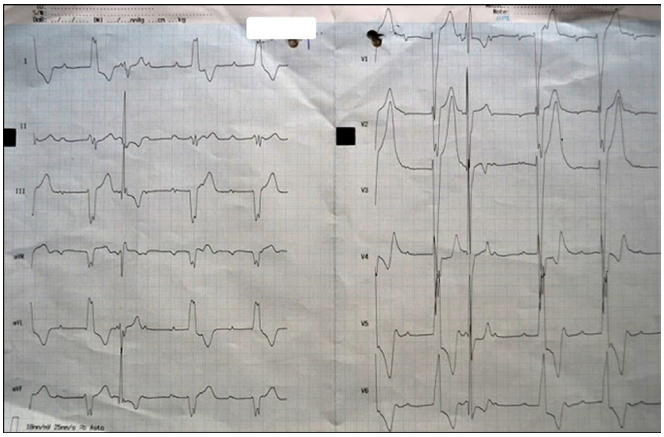

Figure 1 Initial ECG demonstrating complete heart block and a wide complex ventricular escape rhythm of left bundle branch morphology (atrial rate 110/min, ventricular rate 55/min).

ECGs were done, and 3 months later, his rhythm reverted to normal sinus rhythm (figure 4).

DISCUSSION

In 1946, Filberbaum and colleagues had reported various rhythm disturbances associated with acute rheumatic fever ranging from sinus arrest, atrial tachycardia, junctional tachycardia and junctional rhythm to AV conduction block.[2] The most common AV conduction abnormality during acute rheumatic fever, first-degree AV block (up to 72.5%) has been attributed to increased vagotonia and may not be associated with carditis. Second-degree AV block of Mobitz type I is much less frequent occurring in 0.06%–2.6% of cases and complete AV block is least common occurring in 0.016% to 4.6% cases in various case series.[1–3] In a report, Carano *et al*[4] have compiled 25 cases of complete AV block associated with acute rheumatic fever. Five more cases have been reported since then.[5–7] Most cases involving AV block associated with acute rheumatic fever occurred during the first attack with minimal valvulitis. Ours is the first case of CHB occurring during recurrence of rheumatic fever in a patient with established rheumatic heart disease. Of the total 30 cases reported earlier, 24 cases were in the paediatric and adolescent age group. Twelve cases experienced Stokes-Adams attacks. The duration of AV block varied from few minutes to 10 days in 21 of the reported cases. Of these, CHB reversed to sinus rhythm in 18 of 21 cases during treatment of acute rheumatic fever and persisted in three cases at 3 months. Temporary pacemaker implantation was necessary in 10 cases and only one patient (an adult) required permanent pacemaker implantation.[6] Initial studies have shown that complete AV block during acute rheumatic fever occurs proximal to the bundle of His, and the escape rhythm is usually narrow.[1] But our patient had CHB with wide ventricular escape rhythm of LBBB morphology suggesting involvement below the bundle of His level, although we have not done an electrophysiological study. Exact pathogenetic mechanisms underlying these rhythm disturbances in acute rheumatic fever is unknown. It has been attributed to increased vagotonia (for first-degree heart block) and/or inflammation secondary to acute rheumatic activity per se, since the inflammatory biomarkers (CRP, ESR) are elevated.[1] Stepwise reversibility from third-degree heart block to Wenckebach to prolonged AV conduction in response to anti-inflammatory medications lends credence to inflammatory hypothesis.[5]

Temporary transvenous pacemaker placement is indicated in cases of advanced symptomatic AV block. Our patient showed favourable response to systemic corticosteroids with reversal of CHB not requiring permanent pacemaker insertion. He is being continued on secondary prophylaxis with three-weekly long-acting penicillin.[8]

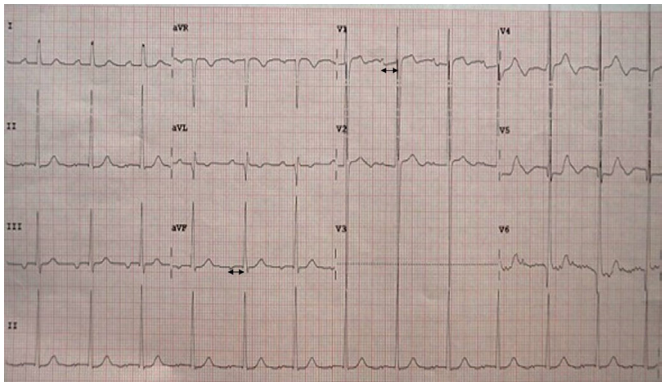

Figure 3 ECG on the fifth day demonstrating ectopic atrial rhythm with PR interval of 220 ms.

CONCLUSION

Advanced AV block (CHB) can rarely occur secondary to acute rheumatic carditis. It is often transient and reverses over the course of days. Temporary pacemaker should be considered in symptomatic cases.

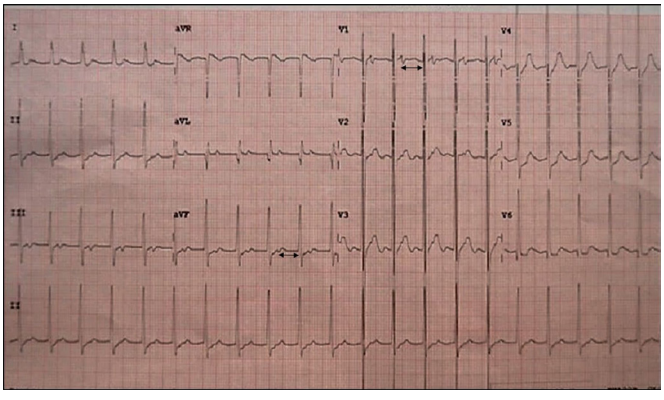

Figure 2 ECG on the third day demonstrating reversal of third-degree heart block to first-degree heart block with PR prolongation (360 ms).

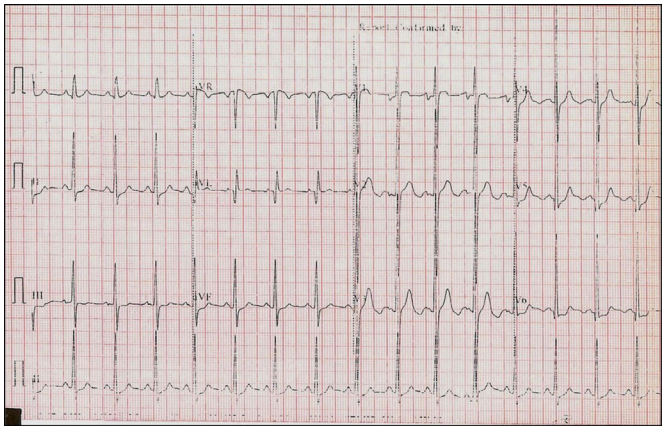

Figure 4 Follow-up ECG demonstrating reversal to normal sinus rhythm at 3 months.

Learning points

- Complete heart block (CHB) can rarely occur secondary to acute rheumatic carditis.
- CHB in acute rheumatic carditis is usually transient and reversible over few days if treated appropriately.
- Temporary pacemaker placement should be considered in symptomatic cases with CHB.

Contributors Both AS and SU equally participated in the analysis and interpretation of data, preparation of manuscript and critically reviewed it for intellectual content and gave final approval for submission.

Competing interests None declared.

Patient consent Obtained.

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