CASE REPORT

Emergent pacemaker placement in a patient with Lyme carditis-induced complete heart block and ventricular asystole

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SUMMARY

We report a case of a 31-year-old man who presented to the emergency department after four episodes of syncope within a 24 h time span. He was found to have symptomatic complete heart block associated with episodes of ventricular asystole lasting 5–6 s. He underwent emergent permanent pacemaker insertion during which he was found to have no underlying rhythm. He was later found to have positive serologies for Lyme disease despite no known exposure to ticks and neither signs nor symptoms of the disease. The pacemaker was ultimately removed due to resolution of his heart block with antibiotic therapy.

BACKGROUND

Lyme disease is a systemic illness caused by the spirochete *Borrelia burgdorferi*, and the most common tick-borne disease in the USA.^{1–2} First described in a cohort of 51 patients in Lyme, Connecticut, in 1977,³ it now represents a significant source of clinical and financial burden in New England and mid-Atlantic states,² with an estimated annual incidence of 329 000 cases⁴ and cost of up to \$1.3 billion each year.⁵

The natural history of Lyme disease is classically divided into three stages. The first represents localised disease in which *B. burgdorferi* spread through the dermis, resulting in the characteristic erythema migrans (EM) rash, often accompanied by a flu-like illness and, occasionally, lymphadenitis.⁶ The second stage is characterised by early-disseminated disease, in which the bacteria spread haematogenously and may cause skin lesions, migratory arthritis, cardiac arrhythmias or meningitis.⁶ The final stage is late disseminated disease, which typically manifests as arthritis and, rarely, with progression of neurological sequelae.^{1–7}

Cardiac sequelae are observed in approximately 1% of all Lyme disease cases⁸ and typically occur 2–5 weeks after appearance of the EM rash, although onset has been reported from 2 days to 7 months after initial infection.⁹ Lyme carditis usually manifests as atrioventricular (AV) conduction block, with symptoms of light-headedness, syncope, dyspnoea, palpitations and/or chest pain.⁷ A recent systematic review of the medical literature identified 45 cases of complete heart block due to Lyme carditis, of which 18 required temporary transvenous pacing. Only two cases, both reported in the 1980s, required permanent pacemakers.¹⁰

We present a case of serologically-proven Lyme disease in a young man with no known tick exposure and no rash, which manifested solely as symptomatic complete heart block requiring emergent pacemaker placement.

CASE PRESENTATION

A 31-year-old Filipino man presented to the ED in July after four episodes of syncope over the 24 h prior to admission. Three occurred during exertion (while playing with his son), and the fourth while entering the shower. He reported palpitations immediately prior to these syncopal episodes but denied chest pain and shortness of breath, as well as fever, recent illness, tick bites, rashes and history of exposure to wooded areas. The patient also denied having ever experienced symptoms of palpitations, light-headedness or syncopal events prior to the current illness.

The patient had a medical history significant for anxiety, depression and insomnia, but not for cardiac or neuromuscular disease. He was taking no medications. He smoked several cigarettes per day and used marijuana nightly to help with sleep. He denied alcohol, cocaine, amphetamine and other drug use. He had no family history of sudden death but did have an uncle with an implantable cardioverter defibrillator (ICD). Notably, he had an identical twin brother with no known cardiac issues.

INVESTIGATIONS

In the ED, the patient was anxious but in no acute distress, and afebrile with a heart rate in the 80 s, blood pressure of 170/90 and oxygen saturation of 99% on room air. Physical examination was unremarkable, with no cardiopulmonary findings, no focal neurological signs and neither jugular venous distension nor lower extremity oedema. His electrocardiogram (figure 1) revealed 2nd degree AV block, as well as T wave flattening and inversions in the inferior and lateral precordial leads, and telemetry showed multiple episodes of high-grade heart block with 5–6 s pauses of ventricular asystole. He was also noted to have a narrow complex junctional escape rhythm with a rate in the 80 s along with a sinus rate in the 100 s. The patient's electrolytes, glucose, thyroid-stimulating hormone level and troponins were all within normal limits. He had a slightly elevated alanine aminotransferase of 56 and alkaline phosphatase of 155. His complete blood count revealed an elevated white blood cell count of 14 600 cells/ μ L, and he had an elevated

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Figure 1 EKG on presentation prior to pacemaker placement, showing an episode of complete heart block.

lactic acid level at 2.1 mmol/L. A bedside echocardiogram showed a normal ejection fraction along with neither wall motion nor valvular abnormalities. Lyme serologies and an anti-nuclear antibody level were ordered.

DIFFERENTIAL DIAGNOSIS

The patient's clinical history allowed us to reject several of the common causes of AV block, including: increased vagal tone, which is unlikely in someone with no history of athletic training; idiopathic progressive cardiac conduction disease, which is not consistent with the acute nature of this presentation; ischaemia, which more commonly presents with chest pain and ST/T wave changes on EKG; cardiomyopathy due to infiltrative diseases such as sarcoidosis or amyloidosis, which would produce

an abnormal echocardiogram and evidence of systemic illness; and, finally, myocarditis due to Lyme, systemic lupus erythaematosus, or other infectious/inflammatory aetiologies, which would be associated with systemic symptoms such as fever or rash. Sudden unexplained nocturnal death syndrome, known as Bangungut in the Philippines, was entertained based on the patient's family history of an uncle who required ICD placement and the patient's Filipino heritage, but this syndrome is more classically associated with ventricular arrhythmias.¹¹ In addition, the characteristic EKG findings of Bangungut, including right bundle branch block and ST elevations in the precordial leads (also seen in Brugada Syndrome¹¹), were absent in this patient. Nevertheless, given that the patient had an identical twin, concern for a genetic cause of his heart block was expressed to

Figure 2 EKG after pacemaker removal showing complete recovery of electrical conduction.

him, with encouragement that his twin seek evaluation for preventative medical care.

TREATMENT

Given the patient's continued episodes of symptomatic complete heart block, the decision was made to admit him for emergent implantation of a dual chamber pacemaker, with one lead to the right ventricular apex and one to the right atrial appendage. Notably, throughout the procedure, the patient experienced multiple episodes of complete heart block with asystole lasting 4–6 s. After each episode of pacing, it took several minutes to re-establish a junctional escape rhythm. When the pacemaker was connected to the leads, the patient had no underlying rhythm and thus was completely pacemaker-dependent.

After insertion of the pacemaker, the patient was completely asymptomatic and was discharged with follow-up. One week after presentation, IgG and IgM Lyme serologies were found to be positive and the patient was started on a 30-day course of doxycycline. Although symptomatic heart block due to Lyme carditis is typically treated with intravenous ceftriaxone, oral doxycycline was deemed appropriate therapy given that the patient had already received a pacemaker and was to be treated as an outpatient.¹²

OUTCOME AND FOLLOW-UP

Approximately 3 weeks after discharge, the patient's EKG revealed an intrinsic PR interval of 190 ms, indicating significant improvement in his electrical conduction abnormality. The dual chamber pacemaker was removed within 3 months of placement, after which his EKG (figure 2) showed complete recovery of electrical conduction.

DISCUSSION

The patient's acute presentation of complete heart block in the setting of positive Lyme borrelia IgM and IgG serologies points to a diagnosis of Lyme carditis. While our patient did not exhibit manifestations of localised Lyme disease and had not had a recent flu-like illness, heart block is a well-known manifestation of early-disseminated Lyme disease. Furthermore, our patient was residing in Connecticut, a state with the sixth highest incidence of Lyme disease in 2014, in the USA,¹³ and developed symptoms in July, which is in the middle of the 3-month period when 69% of Lyme carditis patients are diagnosed.⁸ However, it must be noted that IgM and IgG antibody responses to Lyme borrelia can persist for 10–20 years, and may therefore represent prior exposure rather than active infection.¹⁴ Nevertheless, we feel confident in attributing the patient's arrhythmia to Lyme disease because of the timing and setting of his acute presentation, and because of his dramatic response to antibiotic therapy.

Also consistent with a diagnosis of Lyme carditis are the patient's demographic characteristics; a prior systematic review of 45 cases of Lyme carditis causing complete heart block showed that 84% of cases were males with a median age of 31 years. Further, more than 40% presented after a syncopal episode, 40% required pacing for management of the heart block and 56% lacked the characteristic EM rash, as in our patient's case.¹⁰ Similarly, among Lyme case reports submitted to the Centers for Disease Control and Prevention from 2001 to 2010, only 42% of Lyme carditis patients had an accompanying EM rash.⁸ Overall, it is estimated that 70–80% of patients with Lyme disease present with the EM rash.¹ As with our patient, it is also estimated that 30–50% of patients do not recall a tick bite.¹⁵

A unique aspect of this case, however, was the rapidity with which the patient progressed from onset of symptoms to

complete AV block (1–2 days); the median reported in the literature is 14 days (range of 2–24 days). Typically, Lyme carditis-induced AV block resolves within 1–6 weeks on antibiotic therapy in a stepwise progression from complete block (which generally resolves within 1 week), to first degree heart block, to normal sinus rhythm.⁹ The recommended regimen consists of 14–21 days of antibiotics, starting with intravenous ceftriaxone and then transitioning to oral doxycycline therapy. However, as mentioned above, doxycycline alone can also be used for outpatient treatment in select circumstances.¹² According to the 2008 American College of Cardiology/American Heart Association/Heart Rhythm Society Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities, Lyme carditis-induced AV block is not an indication for permanent pacemaker therapy given that it is expected to both resolve completely and to not recur with appropriate antibiotic therapy.¹⁶ When Lyme is suspected as a potential cause for complete heart block in patients presenting in endemic regions, temporary pacemaker placement and antimicrobial therapy are appropriate. Permanent pacemaker therapy is therefore only indicated if the heart block fails to resolve after 6 weeks.⁹ However, this is exceedingly rare, as there are only two prior reports of patients requiring permanent pacemakers due to persistent AV block.^{17 18}

AV block in Lyme carditis most commonly occurs above the bundle of His in the AV node but can also occur within the atrium, bundle of His, bundle branches and fascicles.⁹ Our patient's initial narrow complex junctional escape rhythm indicated block at or above the bundle of His, yet his multiple episodes of ventricular asystole and lack of an underlying rhythm during pacemaker insertion suggest possible diffuse conduction system involvement.¹⁹ In line with this possibility is a prior study of patients with Lyme carditis, which suggested that the T wave flattening and inversions in the inferior and lateral leads in these patients (also observed in the present case) represent evidence of diffuse cardiac involvement.³ Several prior episodes of Lyme-induced complete heart block leading to the failure to generate an escape rhythm have been reported,^{19–23} yet, unlike in our patient, most of these cases were in patients with other clinical manifestations of Lyme disease.

High-grade heart block is a severe life-threatening condition, as it can lead to cardiac arrest via ventricular tachycardia and ventricular fibrillation¹⁰ or, as in our patient, periods of ventricular asystole. Brief asystole in Lyme carditis is associated with a worse prognosis, along with escape rhythms with a wide QRS complex and fluctuating bundle branch blocks.⁶ It is interesting to note our patient's relatively quick progression from an adequate junctional escape rhythm in the 80 s to asystole observed during the pacemaker implantation procedure. This appears to be consistent with prior reports showing that heart block due to Lyme carditis can fluctuate and advance rapidly within minutes to hours.¹⁰

Lyme carditis resulting in AV block is believed to be due to the host's immune response to *B. burgdorferi* in the myocardial tissue.^{7 10} Patients with complete AV block due to Lyme disease have demonstrated lymphoplasmacytic infiltration of the endocardium and perivascular myocardium along with myocyte injury on cardiac biopsy.⁹ Mouse models have helped elucidate the pathogenesis of conduction abnormalities in Lyme carditis. Immunofluorescence studies of murine cardiac tissue after Lyme inoculation have shown borrelia to be present at the AV junction as well as the epicardium and less commonly in the myocardium.⁹ These findings, as well as the rapid clinical response to antibiotics and minimal efficacy of steroids, suggest that the pathogenesis of heart block in Lyme carditis hinges on local

effects of spirochetes on the conduction system rather than induction of an autoimmune reaction against myocardial tissue. Nevertheless, further elucidation of the immunopathogenic mechanisms underlying Lyme borrelia-induced conduction disease is certainly needed.⁹

Learning points

- ▶ Lyme carditis must be considered in the differential in all patients who present with syncope and heart block even in those without antecedent or concurrent signs or symptoms of Lyme disease or known exposures, especially in endemic areas.
- ▶ Heart block due to Lyme carditis can progress quickly from second degree atrioventricular block to high-grade block, requiring a high degree of clinical suspicion to ensure timely intervention.
- ▶ Lyme carditis can cause complete heart block with associated ventricular asystole, requiring emergent temporary pacemaker therapy.

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