FELLOWS CASE OF THE MONTH

COMPLEX CASE STUDY

Conduction Riddles of Lyme Carditis: A Case Series

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ABSTRACT. Carditis is one of the manifestations of the Lymes disease which can occur during the early-disseminated phase of the illness. Conduction block at the atrioventricular node level is the most common manifestation of Lyme’s carditis. We report three rare cases of multilevel conduction block secondary to lymes carditis with corresponding electrophysiologic studies. The occurrence of infrahisian block in conjunction with nodal disease has rarely been reported. The exact pathophysiology behind the predominant selection of the cardiac conduction system is unclear. In order to determine the reversibility of such a conduction disease, electrophysiology study and MIBG scan were performed. Intravenous antibiotic therapy was started and resolution of conduction disease was progressively documented. In conclusion, Lyme carditis can present early on with dizziness or syncope as the sole manifestation of a diffuse conduction disease.

KEYWORDS. Keywords: lyme infection, hisian block, AV node, MIBG scan.

Introduction

Borrelia burgdorferi is a Gram-negative spirochete bacterium identified as the causative agent for Lyme disease and is transmitted to humans by an infected tick bite. Carditis is one of the manifestations of the disease and can occur during the early-disseminated phase (Stage II) of the illness. However there have been cases with cardiac symptoms as early as 4 days to as late as 7 months after the tick bite.1

Conduction block is the most common manifestation of Lyme carditis followed by other manifestations such as heart failure, and myocarditis if not treated.2 The cause for the predilection of the disease to the conduction system is unknown. The Aschoff node is affected in most cases, resulting in atrioventricular (AV) block at that site.2

We report three cases of unusual hisian, infrahisian block along with AV node and sinus node dysfunction in the setting of early Lyme carditis.

Case 1

A 48-year-old man with no past medical history presented to the emergency room with shortness of breath and dizziness for 3 days. His examination was unremarkable except for an erythematous macular rash on the left shoulder and a marked bradycardia of 38 bpm. His initial resting electrocardiogram revealed sinus bradycardia with complete heart block and a ventricular escape rhythm of 39 bpm. An echocardiogram was normal. Retrospective detailed history revealed that the patient was on a holiday at Poconos mountains 3 weeks previously and he recalled being bitten by a tick on the abdomen and the left shoulder, where he developed a centrally clearing rash. A Lyme titer was sent and the patient was started on intravenous ceftriaxone. A transvenous pacemaker was placed after transferring the patient to the coronary care unit with an admitting diagnosis of heart block secondary to Lyme carditis.
The next day immunoglobulin (Ig)G titers against Borrelia burgdorferi were positive, which was confirmed by western blot assay. Given his presentation with positive serologic results, an electrophysiologic study without sedation was performed to rule out extensive His–Purkinje disease.

The AH interval was significantly prolonged and the His potential was fractionated (Table 1). Complete heart block with ventricular escape had improved to 2:1 block by the time the electrophysiologic study was performed. Although in most cases each A depolarization was followed by an H in both AA intervals that included a QRS complex and those that did not (Figure 1), we did notice the absence of His bundle potential in a few blocked beats while the recording catheter was stable, as well as prolonged HV (Table 1) when the beat was conducted. These findings were considered as evidence of both intranodal and infranodal block. Administration of atropine partially corrected the AV conduction: the 2:1 conduction improved to a Wenckebach block; the paced atrial cycle at which AV nodal block occurred was 640 ms. Fractionation of His potential and HV prolongation were noticed once AV nodal conduction improved and AH shortened during 1:1 conduction (Figures 1–3).

Case 2

A 62-year-old male with no significant past medical history presented to the emergency department with complaints of lightheadedness for 2 days. His rhythm was sinus bradycardia at 50 bpm with first-degree AV block and episodes of atrial tachycardia with high-grade AV block. He had developed a groin rash 4 weeks previously following a golf tournament, and had been treated with cephalaxin. Lyme carditis was suspected and the patient was admitted to the coronary care unit with ceftriaxone treatment. Twelve hours after presentation, the patient developed severe sinus bradycardia followed by sinus arrest. Administration of atropine followed by emergent transvenous pacemaker restored organ perfusion. The patient tested positive for IgM and IgG antispirochete antibodies. The echocardiogram revealed normal ejection fraction with mild mitral regurgitation and mildly dilated left atrium. During his first week of hospitalization the patient remained symptomatic with spells of dizziness. The cardiac rhythm alternated between severe sinus bradycardia, episodes of second-degree AV block along with episodes of atrial flutter conducted at a 3:1 rate.

The patient underwent an electrophysiologic study that showed prolongation of the sinus node recovery time with good response to atropine in addition to a prolonged AH interval and a fractionated His potential (Subject 2 in Table 1). Atropine partially corrected the AV conduction: the 2:1 conduction improved to a Wenckebach block. Antibiotics were continued and radioisotope scanning with an iodine-131-meta-iodobenzylguanidine (MIBG) scan showed a low heart to mediastinum ratio, suggesting low cardiac adrenergic receptor density. He was discharged home with a total intravenous course of 6 weeks of ceftriaxone as his condition improved.

Case 3

A 56-year-old previously healthy man presented to the hospital with dizziness and exertional dyspnea. The patient had developed a rash 4 days previously after

**Table 1: Intracardiac intervals.** This table represents the results obtained from both electrophysiologic studies

<table>
<thead>
<tr>
<th>Block site</th>
<th>Initial</th>
<th>Initial</th>
<th>Initial</th>
<th>Initial</th>
<th>Post-atropine</th>
<th>Post-atropine</th>
<th>Post-atropine</th>
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<tr>
<td></td>
<td>AH</td>
<td>HV</td>
<td>CSNRT</td>
<td>His</td>
<td>WCL</td>
<td>WCL</td>
<td>AH</td>
<td>HV</td>
</tr>
<tr>
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<td>59</td>
<td>150</td>
<td>34</td>
<td>640</td>
<td>550</td>
<td>446</td>
<td>63</td>
</tr>
<tr>
<td>Nodal and infranodal</td>
<td></td>
<td></td>
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<td></td>
<td></td>
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<tr>
<td>Subject 2</td>
<td>478</td>
<td>46</td>
<td>650</td>
<td>30</td>
<td>780</td>
<td>560</td>
<td>180</td>
<td>50</td>
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<td>Nodal and sinus</td>
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AH: A to His interval; CSNRT: correct sinus node recovery time; HV: His to V spike; WCL: Wenckebach cycle length.

**Figure 1:** Intracardiac electrograms. Fractionated His potential into HH ´ with an isoelectric line in between indicating infranodal block.

**Figure 2:** Post atropine shortening of the AH interval suggesting suprahnial block.
of tick bite and denied any other systemic symptoms. No pertinent physical findings were noted except for jugular cannon A waves. His blood pressure was 100/60 mmHg with a heart rate of 50 bpm. His electrocardiogram showed a sinus rhythm with third-degree heart block and junctional escape rhythm. A transvenous pacemaker was inserted. The patient was admitted to the critical care unit and ceftriaxone treatment was started. The initial laboratory work, including cellular blood count, cardiac enzymes and electrolytes, was normal except for an elevated sedimentation rate. Echocardiogram findings were normal. Further laboratory testing included ELISA testing of IgG titers against *Borrelia burgdorferi* and Western blot assay. Levels were abnormally elevated and positively correlated with the severity of the AV block. MIBG was also performed and revealed a decreased heart to mediastinum ratio. The patient refused to have an electrophysiologic study performed for the two patients, electrophysiologic studies performed for the two patients, the absence of the His bundle in most of the cycles with skipped beats favored a nodal or infrahisian block. The administration of atropine shortened the refractoriness of the AV node, thus improving the conduction at that level to a 1:1 response. As a consequence, the disease at the infrahisian fibers was unmasked and a His bundle spike in the skipped beats was consistently seen. His–Purkinje disease was initially unrecognized due to the coexistent block at a higher level.

The exact pathophysiology behind the predominant selection of the cardiac conduction system is unclear since cardiac biopsies are rarely available. The spirochete may affect the entire cardiac tissue, but the inflammation is found predominantly in the connective tissue at the base of the heart, the base of the interventricular septum, and perivascular regions. Endocardial fibrosis and lymphocytic infiltration of the AV node is usually noted. These histopathologic findings indicate that conduction disease results from invasion by the spirochete or from the inflammatory response at the block site rather than through an indirect vagotonic effect. It is not clear why the organism predominantly affects the conduction system, and more specifically the AV nodal tissue. Several hypotheses can be postulated: first, the histological location of the conduction system could determine the degree of involvement. The sinus node is subepicardial, perivascular, and in continuation with the rest of the atrial cells. These properties make it vulnerable to the Lyme spirochete, though to a lesser extent than the AV node, which is located at the base of the heart in the endocardium of the atrial septum. The specialized myocytes at the AV node have been described to be separated from the contracting myocytes by a thin sheet of connective tissue without being insulated. The His bundle is in contrast insulated by connective tissue from the surrounding myocardium. These anatomical and histologic differences could explain the predilection of the spirochete to the conduction system and more specifically to the AV nodal tissue. Second, the unique metabolism of conducting cells could also explain the taking a trip into woody areas. He did not recall a history of Lyme carditis is a self-limiting AV conduction abnormality presenting as dizziness, syncope, and shortness of breath with or without chest pain. The spirochete can affect any layer of the heart causing pericardial effusion, congestive heart failure, tachyarrhythmia and QT prolongation. The AV conduction abnormalities fluctuate in severity but have mostly been described in the literature as nodal. Van Der Linde et al. reviewed a total of 105 cases, out of which 19 had an electrophysiologic study. Among those 19 patients, 10 patients (53%) had a prolonged AH interval, suggesting a proximal block of the AV conduction system. Three patients (16%) had only HV interval prolongation, indicating a more distal localization to the block. Only six patients (31%) had a prolongation of intra-atrial conduction and delayed AH and HV conductions, implying a diffuse disease similar to the patients described in this report. In the series described, two of the patients accepted to undergo electrophysiologic study. His bundle electrograms were recorded with a 6 French bipolar catheter to determine the site of the block. The recordings were made at a speed of 100 and 200 mm/s. For each pacing cycle AH intervals were measured in a standard fashion. The local His A was measured as the earliest reproducible rapid deflection. Nodal involvement was seen in all cases in conjunction with a diseased Purkinje system, sinus node disease, or atrial tachyarrhythmia. The earlier occurrence and diffuse nature of these presentations could be an indicator either of the virulence of the pathogen or of the vulnerability of the conduction disease of the host. In the electrophysiologic studies performed for the two patients, the spirochete to the conduction system and more specifically to the AV nodal tissue. The degree of involvement. The sinus node is subepicardial, perivascular, and in continuation with the rest of the atrial cells. These properties make it vulnerable to the Lyme spirochete, though to a lesser extent than the AV node, which is located at the base of the heart in the endocardium of the atrial septum. The specialized myocytes at the AV node have been described to be separated from the contracting myocytes by a thin sheet of connective tissue without being insulated. The His bundle is in contrast insulated by connective tissue from the surrounding myocardium. These anatomical and histologic differences could explain the predilection of the spirochete to the conduction system and more specifically to the AV nodal tissue. Second, the unique metabolism of conducting cells could also explain the...
tropism of the spirochete to the electrical system. These specialized cells contain numerous glycogen granules, a component not present as abundantly in the contracting myocytes, which rely on fat oxidation for energy production. The natural predilection of spirochetes for glucose could explain the manifestation of Lyme carditis with conduction disease.\textsuperscript{10}

Another ancillary test that can be performed in the setting of Lyme carditis is MIBG scintigraphy study. The lower than normal heart to mediastinum ratio has been documented in previous subjects with Lyme’s disease, with a mean of 1.4.\textsuperscript{11} It indicates an impairment of neuronal cardiac uptake in the setting of acute myocarditis. Though none of the patients had evidence of clinical heart failure, the decreased myocardial MIBG accumulation indicates early myocardial involvement.\textsuperscript{11} Treatment depends primarily on the severity of the conduction disease: it includes systemic ceftriaxone or penicillin for second/third-degree AV block or prolonged PR >300 ms along with close monitoring.\textsuperscript{12} For milder cases, oral doxycycline or amoxicillin is appropriate. The duration of therapy depends on adequate shortening of the PR interval and the normalization of AV block. Therapy is usually discontinued after 4–6 weeks, as complete heart block is usually transient and only rarely requires the placement of a permanent pacemaker.\textsuperscript{13–16} The resolution of the AV block has been attributed to the regression of inflammation with early initiation of antibiotics, despite the absence of clinical studies that test this hypothesis.\textsuperscript{9} In this case series, the reversibility was proven with atropine injection during electrophysiologic study, consequently conservative management was adopted with temporary pacemaker placement, antibiotics and continuous monitoring.

References


