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A Message from Cardiology Associates, LLC



About the Author

Dr. Elizabeth Kingsley practices general cardiology in Annapolis and Kent Island, Maryland. She has been a member of Cardiology Associates since 1984, where she moved directly after her cardiovascular fellowship at the Harvard University Boston Beth Israel Hospital. While there, she had special interest in myocarditis and trained under Dr. Eugene Braunwald and Dr. William Grossman. She presents a case today of myocarditis. Dr. Elizabeth Kingsley practices at Anne Arundel Medical Center and is on staff both at Anne Arundel Medical Center and Washington Hospital Center. She has been honored by the American Heart Association

for her work in bringing education and awareness to women and the cardiovascular community regarding heart disease in women. She continues to be interested in cardiovascular diagnosis and management in all populations, but has a particular interest in the geriatric age group as well.

Lyme Carditis and Manifestations of Lyme Disease



Ixodes scapularis tick



Erythema migrans

Lyme disease, or Lyme borreliosis is a systemic disease caused by the spirochete *Borrelia burgdorferi* and it is transmitted by the Ixodes tick. The disease is divided into three stages. Stage 1, or the early localized stage, occurring 2-30 days after the initial tick bite, can present as a flu-like illness and can present with an erythema migrans. Stage 2, or early disseminated stage, occurs within weeks to months and is characterized by neurologic complications, musculoskeletal pain and the cardiac manifestations can occur at this time. Stage 3

can occur several months to years later and is characterized by monoarthritis or oligoarthritis, affecting large joints and having neurologic symptoms.

The cardiovascular manifestations were first described in 1980 with a report of 20 North American cases, but cases have since been reported worldwide, including Australia and Europe. The incidence of Lyme disease is increasing substantially just based on CDC reports, and cases are underreported. It is estimated that 4-10% of patients in the United States with untreated Lyme disease develop carditis. One of the most prevalent forms of Lyme carditis is the development of atrioventricular block. The Lyme carditis prevalence is 3:1 male to female, even though there is a slight female to male predominance for the disease as a whole. The pathologic findings of Lyme disease on histology shows a transmural inflammatory infiltrate and in the hyperacute disease, small inflammatory nodules composed primarily of neutrophils and macrophages can be seen. Later, lymphoid cells infiltrate the endocardium and the characteristic band-like or plaque-like pattern can be seen. The isolated myocyte necrosis and ultimate interstitial fibrosis can be seen. The large intramyocardial vessels can be affected with scattered vasculitis. It is not clear whether the presence of live spirochetes is necessary for continued disease or whether the disease can result from immune-mediated mechanisms, but live spirochetes have been cultured from biopsy specimens of patients.

Lyme carditis usually occurs between June and December and usually after a tick bite or EM (erythema migrans) has occurred, with a range of 4 days to 7 months. The patient may or may not recall the rash or the tick bite. Cardiac conduction disease and complete heart block has been reported as the sole manifestation of Lyme carditis. Joint complaints are found in 51%, EM in 67%, neurologic illness in 27% with Lyme carditis. The patient can present with lightheadedness, syncope, dyspnea, palpitations. Only 5% had left ventricular systolic dysfunction and 21% were hospitalized. There are fatalities resulting from Lyme disease; however, they are rare. There is a report of a patient with coexistent Babesia microti infection who died. One retrospective study showed that 45-52 patients with Lyme carditis had documented AV block, 40% had Wenckebach type block and 50% had complete heart block. The conduction disturbance can be fluctuating, varying from prolonged PR interval to complete heart block. Fifty percent went on to complete heart block. There can be tachy or bradyarrhythmias. The tachyarrhythmias are usually supraventricular. Rarely ventricular and fascicular tachyarrhythmias have been reported. With the AV nodal block, many patients do not respond to atropine sulfate administration, implying that the AV nodal block was not vagotonic, but more likely direct effect of the disease on the AV node. There are reports of prolonged AH and HV intervals in patients with alternating bundle branch blocks, suggesting that Lyme disease can affect the conduction system diffusely, but that the AV node seems to be the most vulnerable. There are some cases where permanent pacing for Lyme carditis despite antibiotic therapy becomes necessary. Usually with treatment, the complete heart block generally resolves within a week, and lesser conduction disturbances may resolve within 6 weeks. In one series of 105 cases, temporary pacing was implanted in 35%, and permanent pacer was implanted in 5 patients, but only one ultimately had persistent complete heart block. Ninety-four percent of patients had complete recovery.

Myocardial Disease

Although it is considered nonspecific, myocardial involvement of Lyme disease often presents with diffuse ST-T wave changes. These were seen in 65% of patients in one series. Clinical congestive heart failure is thought to occur in 10-15% of patients with Lyme carditis. Myocardial involvement may present as cardiomegaly or left ventricular systolic dysfunction on echocardiogram due to the isolated myocyte necrosis and endocardial and interstitial fibrosis. Spirochetal forms have been found on endocardial biopsy.

Diagnosis

Usually the diagnosis of Lyme carditis is made by confirming association between clinical and historical features of borreliosis with cardiac findings. Carditis can occur, however, in the absence of EM and patients may not recall the rash or the tick bite. The diagnosis should be suspected when a patient presents with symptoms of palpitations and syncope, especially if there are EKG abnormalities and especially in conjunction with neurologic complaints.

Testing

Serologic testing may support the diagnosis of Lyme carditis, but cannot make the diagnosis. Serologic tests may be negative in the first 6-8 weeks of the disease, so a negative test does not rule out the diagnosis. ELISA testing should reveal seropositivity in patients with Lyme carditis unless it occurs early in the course of the disease. Positive or equivocal results should prompt confirmation with the Western blot analysis because 5% or more of the normal population and patients with severe heart failure are positive in many assays. The echocardiogram may show multiple findings as one would expect in myocarditis, including dilatation of chambers and left ventricular systolic dysfunction.

Treatment

Antibiotic therapy in the early stages of Lyme disease is reported to prevent or attenuate later complications. Lyme carditis has not been studied in a rigorous manner, and although antibiotics are given to clear the infection, there is no evidence that it leads to more rapid resolution of the cardiac features. The most common regimen for treatment of Lyme disease includes 21 days of doxycycline 100 mg orally twice daily. Other regimens include amoxicillin, intravenous ceftriaxone, and tetracycline. If there is severe cardiac conduction disease that does not improve over 24-48 hours, the patient could be given steroids. Therapy is commonly given four weeks or longer, but shorter courses may be efficacious. Pacing is only necessary for high-grade symptomatic hemodynamically compromising heart block. Temporary pacing has been necessary in about 30% of patients. Permanent heart block rarely develops. Pericardial as well as myocardial involvement is usually mild and self-limited. The incidence in one study of cardiovascular manifestations include palpitations in 6.6%, conduction abnormalities in 1.8%, myocarditis in 0.9%, and pericarditis in 0.2%. To summarize in this case, the disease was

suspected based on symptomatology and PR interval prolongation on the electrocardiogram. The patient was initiated on treatment. The enzyme linked immunoabsorbent assay (ELISA) was positive for Lyme disease and then the confirmatory Western blot analysis was positive. With treatment, the patient has improved, he says 100%, with regard to neurologic symptomatology, arthritis, and we have seen normalization of his electrocardiogram and left ventricular systolic function. Oral therapy was all that was needed, but for severe hospitalized patients, intravenous therapy is recommended. The drug of choice is ceftriaxone, 2 grams I.V. once daily in adults. Pacemakers on a permanent basis are avoided, since there is ultimate resolution of the cardiac conduction disease in almost all cases.

Prognosis

The prognosis of Lyme carditis is good in most reports. AV block caused by Lyme disease has persisted for 3-42 days. Complete AV block typically resolves within one week and more minor conduction disturbances resolve within 6 weeks.

Lyme Carditis Case Study

Mr. WF is a 78-year-old man who presented to Anne Arundel Medical Center with atypical chest discomfort. He has a past medical history of thyroidectomy, replaced on Synthroid and a history of dyslipidemia, currently treated with Simvastatin. His only other medication was aspirin. His pain was very atypical and he was discharged from the hospital for further testing as an outpatient. While at Anne Arundel Medical Center, he was noted on his electrocardiogram to have PR interval prolongation. He was in a sinus rhythm and the PR interval was 0.46. He had left anterior fascicular block and a sinus rhythm of 79 beats per minute. He did undergo an echocardiogram, which revealed mildly reduced left ventricular systolic function with a left ventricular ejection fraction of 45-50%, mild concentric left ventricular hypertrophy and mild aortic insufficiency. His chamber dimensions were all within normal limits. He was evaluated with a bilateral duplex carotid Doppler because of a complaint of recent syncope. On review of systems, the patient had symptoms of recent arthritis involving multiple joints, including the knees and he reported symptoms where he felt as though "a fog" would come over him. Before leaving the hospital, thyroid functions were normal and a Lyme titer was drawn at my recommendation. Laboratory abnormalities revealed reactivity and positivity in 11 out of 15 B. Burgdorferi and positive for both IgG and IgM. The patient recalled a tick bite within the past 3-4 weeks, prior to hospitalization at a Department of Natural Resources function, which was held outside. Review of the patient's prior electrocardiograms revealed that the left axis deviation was not new, and PR intervals were less than 0.16 on four prior electrocardiograms performed over the last three years. There were no ST-T wave abnormalities on the acute electrocardiogram despite the finding of mild left ventricular systolic dysfunction on the echocardiogram. The patient was treated empirically and immediately with doxycycline 100 mg po daily, and Western blot confirmation of the Lyme borreliosis was made. Eight weeks after institution of antibiotic therapy, the PR interval had improved to 0.280. The left ventricular systolic function normalized. The patient ultimately did undergo nuclear stress testing because of his risk and presentation with chest pain, and that was negative for myocardial ischemia. The patient at the 8-week period post-treatment also noted significant resolution of his neurologic symptomatology and lightheadedness, as well as improvement in his joint pain.

Ixodes scapularis tick Erythema migrans

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