Credo Reference

Lyme Disease

by , |

DESCRIPTION

Lyme disease is a tick-borne disease caused by a spirochete transmitted to humans through bites of blacklegged (or deer) ticks. The disease was identified after a number of children living near Lyme, Connecticut, presented with a unique symptom cluster in the 1970s. Although the disease has been reported in all 50 states, the incidence is highest in New England and Mid-Atlantic states, Minnesota, Wisconsin, and northwestern California. Initial symptoms often include a rash at the site of the bite, malaise, and fatigue; later manifestations may include neurologic, cardiovascular, and musculoskeletal symptoms (Bratton, Whiteside, Hovan, Engle, & Edwards, 2008; Centers for Disease Control and Prevention, 2008).

NEUROPATHOLOGY/PATHOPHYSIOLOGY

Lyme disease is transmitted to humans by blacklegged ticks (***Ixodes scapularis***) in New England and the Great Lakes regions, and western blacklegged ticks (***Ixodes pacificus***) in the West (Bratton et al., 2008). In larval and nymphal stages of the life cycle, ***Ixodes*** ticks feed on the white-footed mouse, which is the primary host for the spirochete (***Borrelia burgdorferi***) responsible for Lyme disease. Adult ***Ixodes*** ticks feed on deer, which do not carry ***B. burgdorferi***. Infection with ***B. burgdorferi*** initially occurs at the site of the tick bite, and the most common initial symptom is a distinctive rash (erythema migrans) that gradually expands over the course of several days. Transmission occurs only after prolonged attachment and feeding (greater than 24 hours).

Histologic examination of erythema migrans lesions reveals immune response to the infectious spirochete including increased lymphocytes, macrophages, proinflammatory cytokines, and antibody production (Steere, Coburn, & Glickstein, 2004). After an interval (ranging from 3 to 30 days), the organism may spread through the lymph system and bloodstream with a propensity for the central nervous system (CNS), joints, heart, and eyes. In many ways it mimics syphilis in that after inoculation, spirochetemia transpires with widespread dissemination, which when involving the CNS, presents as meningitis (Roos, 2007).

NEUROPSYCHOLOGICAL/CLINICAL PRESENTATION

The course of Lyme disease has been classified into three stages (Steere et al., 2004). During the first stage, the most common sign of infection is the development of a spreading skin lesion (erythema migrans) at the site of the tick bite within 1–2 weeks. The characteristic presentation of erythema migrans is a bull's-eye or target-shaped rash that is not painful but may itch. Additional symptoms of fatigue, malaise, elevated temperature, and joint pain are also found in one-third of

cases or fewer. After the initial localized infection, the second stage is generalized infection in which ***B. burgdorferi*** is found in the cerebrospinal fluid (CSF) and blood as well as spread through organs including the heart, retina, liver, spleen, meninges, and brain. During the second stage, additional erythema migrans lesions may develop, joint or muscle pain may be present, and neurological signs may occur. Neurologic symptoms of disseminated infection can include meningitis, neuropathy (especially, facial nerve palsy), radiculopathy, and subtle encephalopathy with memory and concentration difficulties and changes in mood and sleep. Elevated protein levels in CSF differentiate disseminated infection from viral infection. In children, the primary neurologic symptom is acute facial paralysis. If left untreated, transition to a third stage will occur 6 months to several years after infection and includes primarily rheumatologic (chronic arthritis, especially in the knees) or neurologic symptoms. Third-stage neurologic manifestations may include chronic meningitis resulting in narrowing of leptomeningeal arteries and subsequent infarction (Miklossy, Kuntzer, Bogousslavsky, Regli, & Janzer, 1990).

A minority of patients who have been treated for Lyme disease continue to report neurocognitive and musculoskeletal symptoms in a syndrome referred to as ***posttreatment chronic Lyme disease*** (PCLD; Radolf, 2005). PCLD is controversial due to differences of opinion on whether it represents persistent infection or a noninfectious condition akin to chronic fatigue syndrome (Halperin et al., 2007; Radolf, 2005). The neurocognitive features of Lyme encephalopathy may include difficulty with verbal memory, naming, and attention in addition to changes in mood and sleep patterns (Bratton et al., 2008). Although these symptoms persist in a number of patients, studies have failed to consistently evidence objective neurocognitive deficits despite lowered positive affect (Elkins, Pollina, Scheffer, & Krupp, 1999) and subjective memory complaints (Kaplan et al., 2003). In a comparison of patients with PCLD and patients with abnormal findings in CSF, patients with CSF abnormalities demonstrated objective memory deficits, and both groups of patients reported greater memory complaints than controls (Kaplan et al., 1999). At long-term follow-up, patients previously diagnosed with Lyme disease scored within normal limits on measures of attention, processing speed, verbal memory, and verbal fluency (Kalish et al., 2001). Neuropsychological outcomes in children treated for Lyme disease consistently demonstrate a good prognosis with no neurocognitive differences found between children treated for Lyme disease and controls at 2- and 4-year follow-up (Adams, Rose, Eppes, & Klein, 1994, 1999) and across a range of follow-up intervals from 7 to 161 months (Vázquez, Sparrow, & Shapiro, 2003).

DIAGNOSIS

Diagnosis of Lyme disease relies heavily on clinical features and is supported when the presentation includes an erythema migrans lesion and history of living or traveling in an endemic region (Bratton et al., 2008; Wormser et al., 2006). History of a tick bite can be helpful to ascertain, but patients frequently do not recall a tick bite. Typical laboratory tests (e.g., blood counts) are nonspecific for Lyme disease, and serologic tests (e.g., antibody titers) are often inconclusive as seroconversion may occur late, be absent, or indicate prior exposure rather than recent infection.

TREATMENT

Treatment of Lyme disease may include oral or parenteral antimicrobial regimens (for medications and dosages see Halperin et al., 2007). Adjustments in the pharmacotherapeutic regimen are made when neurologic symptoms are present. Treatment guidelines for PCLD are variable, and there is no strong evidence for continued antimicrobial therapy in those patients. Oral doxycycline (100 mg twice daily for 2 weeks) is useful in addressing the facial palsy that may occur in the absence of CSF abnormalities whereas intravenous ceftriaxone for 4 weeks is preferred for other neurological features related to Lyme disease (Roos, 2007). As suggested, transmission is usually dependent upon prolonged attachment and feeding over a 24-hour period. Consequently, prevention becomes an essential intervention. When going into areas that are more likely to have ticks, individuals may use repellants. In addition, clothing that reduces the chances of ticks getting on the person or into hidden areas is recommended. Carefully checking one self for ticks after leaving such higher-risk areas is recommended.

Bibliography

- Adams, W. V.; Rose, C. D.; Eppes, S. C.; Klein, J. D. (1994). Cognitive effects of Lyme disease in children. Pediatrics, 94, 185-189.
- Adams, W. V.; Rose, C. D.; Eppes, S. C.; Klein, J. D. (1999). Long-term cognitive effects of Lyme disease in children. Applied Neuropsychology, 6, 39-45.
- Bratton, R. L.; Whiteside, J. W.; Hovan, M. J.; Engle, R. L.; Edwards, F. D. (2008). Diagnosis and treatment of Lyme disease. Mayo Clinic Proceedings, 83, 566-571.
- Centers for Disease Control and Prevention. (2008). Surveillance for Lyme disease—United States, 1992-1996 (Surveillance Summaries, Morbidity and Mortality Weekly Report, Vol. 57, SS-10). Author Atlanta, GA.
- Elkins, L. E.; Pollina, D. A.; Scheffer, S. R.; Krupp, L. B. (1999). Psychological states and neuropsychological performances in chronic Lyme disease. Applied Neuropsychology, 6, 19-26.
- Halperin, J. J.; Shapiro, E. D.; Logigian, E.; Belman, A. L.; Dotevall, L.; Wormser, G. P., et al. (2007). Practice parameter: Treatment of nervous system Lyme disease (an evidence-based review). Neurology, 69, 91-102.
- Kalish, R. A.; Kaplan, R. F.; Taylor, E.; Jones-Woodward, L.; Workman, K.; Steere, A. C. (2001). Evaluation of study patients with Lyme disease, 10-20 year follow-up. The Journal of Infectious Diseases, 183, 453-460.
- Kaplan, R. F.; Jones-Woodward, L.; Workman, K.; Steere, A. C.; Logigian, E. L.; Meadows, M. E. (1999). Neuropsychological deficits in Lyme disease patients with and without other evidence of central nervous system pathology. Applied Neuropsychology, 6, 3-11.
- Kaplan, R. F.; Trevino, R. P.; Johnson, G. M.; Levy, L.; Dornbush, R.; Hu, L. T., et al. (2003). Cognitive function in post-treatment Lyme disease: Do additional antibiotics help? Neurology, 60, 1916-1922.
- Miklossy, J.; Kuntzer, T.; Bogousslavsky, J.; Regli, F.; Janzer, R. C. (1990). Meningovascular form of neuroborreliosis: Similarities between neuropathological findings in a case of Lyme disease and those occurring in tertiary neurosyphilis. Acta Neuropathologica, 80, 568-572.
- Radolf, J. (2005). Posttreatment chronic Lyme disease—What it is not. The Journal of Infectious Diseases, 192, 948-949.

- Roos, K. L. (2007). Viral infections. In Goetz, C. G. (Ed.), Textbook of clinical neurology (3rd ed.,pp. 919-968). Saunders Elsevier Philadelphia, PA.
- Steere, A. C.; Coburn, J.; Glickstein, L. (2004). The emergence of Lyme disease. The Journal of Clinical Investigation, 113, 1093-1101.
- Vázquez, M.; Sparrow, S. S.; Shapiro, E. D. (2003). Long-term neuropsychologic and health outcomes of children with facial nerve palsy attributable to Lyme disease. Pediatrics, 112, e93-e97.
- Wormser, G. P.; Dattwyler, R. J.; Shapiro, E. D.; Halperin, J. J.; Steere, A. C.; Klempner, M. S., et al. (2006). The clinical assessment, treatment, and prevention of Lyme disease, human granulocytic anaplasmosis, and babesiosis: Clinical practice guidelines by the Infectious Diseases Society of America. Clinical Infectious Diseases, 43, 1089-1134.

Citation Information:

Noggle, C. A., & Davis, J. (2011). Lyme disease. In C. Noggle, *The Encyclopedia of Neuropsychological Disorders* (1st ed.). Springer Publishing Company. https://search.credoreference.com/articles/Qm9va0FydGljbGU6MzE1OTEy?aid=103259
