

A Case Presentation on the Treatment and Coding of Nervous System Lyme Disease

The AAN is invested in the development of clinical guidelines based on evidence based medicine. The guidelines help to clarify the scientific literature that supports our practice choices. They also provide an impetus for more research in areas where the literature is unclear or lacking. In an effort to highlight the clinical application of these AAN practice guidelines we will develop case presentations with coding and CPT information. These cases will be available on the Practice Section of the AAN web site at www.aan.com.

Lyme disease is a multisystem infectious disease caused by the tick-borne spirochete, *Borrelia burgdorferi*, which frequently affects the nervous system. There is considerable controversy and uncertainty about the best approach to treatment of neuroborreliosis.

Case Presentation

A 65-year-old male presents to the neurology clinic in the early summer after a hiking trip in the Northeast, with complaints of headache and right facial palsy. A thorough history reveals that he was bitten by a tick three weeks prior. He reports that he had an enlarging circular lesion on his thigh (ultimately 5 inches in diameter), first appearing 6 days after the bite. On examination he appears to be in pain from his headache, has photophobia, nuchal rigidity, and evidence of a right peripheral facial nerve palsy. What is the most likely diagnosis?

Lyme disease is due to infection by a spirochete, *Borrelia burgdorferi* and the body's immune response to the infection. The rash is called erythema migrans; it develops in the majority of patients after the tick bite and may last several weeks. The term 'migrans' refers to the fact that it slowly enlarges (the border 'migrates') over the course of days to weeks. Meningitis and cranial neuritis are the most frequent neurologic manifestations of the infection. In addition, radiculoneuritis occurs and is frequently misdiagnosed. Although Lyme disease has been reported from virtually every state, endemic cycles only exist in specific regions – primarily the Northeast (from New Hampshire to Maryland), the upper Midwest (Minnesota, Wisconsin), and areas of northern California. In patients who have not been in such endemic areas, the probability of contracting Lyme disease is quite remote.

What further tests would confirm the diagnosis?

CSF would reveal a mononuclear pleocytosis. Intrathecal production of antibody to *B. burgdorferi*, determined by comparing Lyme-specific antibody in CSF to that in serum, is helpful diagnostically when present. If performed properly, false positives occur only in neurosyphilis. If performed without comparing to serum Lyme-specific antibody, false positives are common in patients with positive peripheral blood Lyme serologies, or in any patient with CNS inflammation or other blood brain barrier breakdown. The biggest technical limitation of this measure is that, once present, the elevated index may remain 'positive' for many years following successful treatment.

Seroreactivity to *B. burgdorferi*, typically measured initially by ELISA, is usually present at time of presentation, except in a minority of patients for whom a convalescent phase serum sample (2-4 weeks later) is required for detection of antibody to *B. burgdorferi*. False positives occur in syphilis, in which case a reaginic test for syphilis, such as the VDRL, will

usually differentiate between the two spirochetal infections. Nonspecific elevations of serum immunoglobulin, such as in lupus, endocarditis, parvovirus or other infections, can result in false positives as well. These can generally be distinguished by performing a Western blot, interpreted using the CDC consensus criteria. Importantly, following *B burgdorferi* infection, serologies can remain positive for an extended period of time despite successful treatment, and should not be used as a measure of treatment success or failure.

What type of treatment is indicated?

Parenteral ceftriaxone, cefotaxime and penicillin are probably safe and effective treatments for peripheral nervous system Lyme disease and for central nervous system Lyme disease with or without parenchymal involvement (Level B recommendation).

Would an oral agent be acceptable?

Oral doxycycline is probably a safe and effective treatment for peripheral nervous system Lyme disease and for central nervous system Lyme disease without parenchymal involvement (Level B recommendation). Amoxicillin and cefuroxime axetil may provide alternatives but supporting data are lacking.

The patient is discharged. On a follow-up evaluation at 1 year he complains of fatigue, some memory slowing, and disturbed sleep. What is the diagnosis?

Post Lyme Disease Syndrome.

What treatment is indicated?

Prolonged courses of antibiotics do not improve the outcome of Post Lyme Disease Syndrome and are frequently associated with adverse events and are therefore not recommended (Level A recommendation).

The following is ICD-9-CM Coding and CPT information for Nervous System Lyme Disease related to this case.

Diagnosis	ICD-9 Code
Erythema migrans/ Lyme disease	088.81 Lyme disease
Lyme meningitis	First code the underlying disease: 088.81 Lyme disease Then code: 320.7 Meningitis in bacterial diseases classified elsewhere Note: The instruction to use this code combination comes from the index of ICD-9-CM.
VII Cranial neuritis	Use both codes in either order: 351.8 Other facial nerve disorders <i>and</i> 88.81 Lyme Disease Note: The index also instructs to use code 351.8 rather than the code for Bell's Palsy (351.0) when the term "neuritis" is used.
Radiculoneuritis	There are many different codes for radiculoneuritis (indexed under "radiculitis"). Choose the code appropriate to the site.

The following CPT Codes would be used for the neurological visits.

Patient Visit	CPT Code	Requirements
New patient	99205 – level 5 new patient	To achieve a level 5 code, the neurologist would need to satisfy the following conditions: <ul style="list-style-type: none"> • chief complaint • 4 bullets in the history of present illness • 3 bullets in the past medical history, family history, and social history • at least 10 bullets in the review of systems • at least 23 bullets in the single system neurologic examination • sufficient points in the MDM to warrant a highly complex score
Consult	99245 – level 5 consultation	
Return visit	99213 or 99214 – established office visit	99213: <ul style="list-style-type: none"> • expanded focus history or physical examination <i>and</i> • low complexity MDM 99214: <ul style="list-style-type: none"> • detailed history or physical examination <i>and</i> • moderate complexity MDM

Disclaimer

This statement is provided as an educational service of the American Academy of Neurology. It is based on an assessment of current scientific and clinical information. It is not intended to include all possible proper methods of care for a particular neurologic problem or all legitimate criteria for choosing to use a specific procedure. Neither is it intended to exclude any reasonable alternative methodologies. The AAN recognizes that specific patient care decisions are the prerogative of the patient and the physician caring for the patient, based on all of the circumstances involved.