Treatment of Nervous System Lyme Disease

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).¹

**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.¹

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-CM² Coding and CPT® information for Nervous System Lyme Disease related to this case.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>ICD-9² Code</th>
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<tbody>
<tr>
<td>Erythema migrans/ Lyme disease</td>
<td>088.81 Lyme disease</td>
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<tr>
<td>Lyme meningitis</td>
<td>First code the underlying disease: 088.81 Lyme disease</td>
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<tr>
<td></td>
<td>Then code: 320.7 Meningitis in bacterial diseases classified elsewhere</td>
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<tr>
<td></td>
<td>Note: The instruction to use this code combination comes from the index of ICD-9-CM.</td>
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<tr>
<td>VII Cranial neuritis</td>
<td>Use both codes in either order:</td>
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<tr>
<td></td>
<td>351.8 Other facial nerve disorders and</td>
</tr>
<tr>
<td></td>
<td>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.</td>
</tr>
</tbody>
</table>
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:

<table>
<thead>
<tr>
<th>Patient Visit</th>
<th>CPT® Code</th>
<th>Requirements</th>
</tr>
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| New patient         | 99205 – level 5 new patient   | To achieve a level 5 code, the neurologist would need to satisfy the following conditions:  
• 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:  
• expanded focus history or physical examination and  
• low complexity MDM  
99214:  
• detailed history or physical examination and  
• moderate complexity MDM |

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