Case Report

An Unusual Case of Suspected Lyme Neuroborreliosis in a Patient Presenting with Ear Pain

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Abstract

We report a case of suspected Lyme neuroborreliosis (LNB) where the patient's sole symptom was chronic, intermittent episodes of unilateral ear pain. This case was unusual because this patient did not show any other neurological or extraneurological symptoms of Lyme disease. LNB is a manifestation of infection by *Borrelia burgdorferi* spirochetes that can manifest in a number of different clinical presentations depending on the nervous system dissemination of the bacteria. Most commonly, these neurological findings present as peripheral nerve radiculopathies, but rarer involvement of the central nervous system (CNS) can occur if a patient goes untreated. Most often, CNS involvement presents as meningitis with increased lymphocytes but can, in rarer cases, involve the spinal cord or brain parenchyma. The diagnosis of LNB was made after the patient was found to have suspicious lesions in the spinal cord and brain parenchyma on magnetic resonance imaging as well as cerebrospinal fluid Borrelia antibody index was consistent with CNS Lyme disease. We discuss this case as a unique clinical presentation of suspected LNB and the diagnostic findings associated with this infection.

Keywords: Cerebrospinal fluid Lyme antibody index, ear pain, encephalomyelitis, Lyme neuroborreliosis

Introduction

Lyme disease is an infection with a multitude of clinical manifestations caused by several species of the spirochete *Borrelia burgdorferi* sensu lato, most commonly *B. burgdorferi* sensu stricto (ss.) in the United States as well as *Borrelia garinii* and *Borrelia afzelii* in Europe that are transmitted by the *Ixodes* tick species. The progression of Lyme disease is currently considered as two distinct phases: acute localized infection and disseminated infection. The nervous system is the third most common site of Lyme disease (Lyme neuroborreliosis/LNB) involvement in the United States after the skin and joints, with involvement in approximately 10%–15% of infected individuals, whereas in Europe, LNB is more commonly seen than arthritis. Acute neurologic involvement is usually observed weeks to months after initial infection via tick bite, usually presenting as early manifestations of the disseminated infection stage.[1] Clinical manifestations of LNB vary depending on progression of the disease, and patients may not present with the classic erythema migrans rash which may lead to a low index of suspicion for Lyme disease infection.[2] Cranial nerve abnormalities are the most common nervous system manifestation in American Lyme disease, occurring in about 5%–10% of patients within weeks to several months of infection. The facial nerve is the most commonly affected in approximately 80% of patients with cranial nerve involvement, presenting with unilateral or bilateral facial nerve palsy. Other common clinical manifestations include brachial and lumbosacral plexopathies, radiculoneuritis, or lymphocytic meningitis characterized by headaches with waxing and waning of intensity;[1,3] however, unilateral otalgia has not been referenced as a sole presenting symptom of underlying Lyme disease. Late disseminated disease of the nervous system can manifest in both the peripheral nervous system but can also present more rarely in the central nervous system (CNS) as encephalopathy,[4,5] lymphocytic meningitis, or encephalomyelitis that can affect CNS parenchyma.[6,7] The Infectious Disease Society of America current guidelines recommend treatment of LNB

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with 2–4 weeks administration of ceftriaxone, with cefotaxime or penicillin G as alternatives.

Patient Information

Our patient is a 35-year-old male with a past medical history of insomnia, depressive disorder, and obstructive sleep apnea who lives in the Lehigh Valley area of Eastern Pennsylvania, a Lyme-endemic area. His primary complaint was of chronic, intermittent episodes of right-sided ear pain that would last for several seconds before disappearing. Acetaminophen relieved his symptoms but did not prevent them from recurring.

Clinical Findings

Physical examination was notable only for a bulging right tympanic membrane on his first visit. He did not report any signs of neurological deficits or hearing loss.

History and Presenting Complaints

Our patient reported unilateral right ear pain for 8 months that began in early June of 2018. He described the location of the pain as inside the ear canal by the eardrum and reported the pain as a stabbing, shooting sensation that would occur sporadically every few hours. The pain would last briefly, roughly ½ a second to approximately 3 s, and would persist for several cycles before subsiding until the next episode onset anywhere from a day to weeks later. The patient first noted his symptoms in June of 2018 and saw two neurologists in the upcoming months when the symptoms did not improve. The second neurologist recommended the patient receive magnetic resonance imaging (MRI). Two MRIs conducted without contrast on 1-8-19 and 1-10-19 were notable for white matter lesions at the left parietooccipital and mid-left corona radiata areas, a cord signal abnormality of high signal intensity at C3, and cervical spine disc degeneration at C4–C7 discs. Serological testing demonstrated positive Borrelia IgG on enzyme-linked immunosorbent assay that was confirmed with a Western blot. The patient underwent a lumbar puncture on 2-13-19, and cerebrospinal fluid (CSF) Borrelia antibody index was positive, confirming the diagnosis of CNS Lyme disease. A CSF Borrelia polymerase chain reaction (PCR) tested concurrently was negative; however, the sensitivity of this particular test is variable depending on clinical presentation, disease duration, and CSF white blood cell counts. In the United States, an analysis of six studies displayed a median sensitivity of CSF Borrelia PCR at 78% but with a range from 25% to 93%.[7] The patient was noted to live in a Lyme endemic area of the US and had outdoor exposure to ticks. CSF protein quantification was within normal range (34 mg/dl, 15–45 normal), and there was no leukocytosis noted on CSF analysis.

Therapeutic Intervention

The patient was initially treated with doxycycline 100mg PO BID upon diagnosis of Lyme disease; however, this was switched to intravenous (IV) ceftriaxone 2g IV that was administered for 6 weeks from 3 to 8-19-4-19-19.

Follow-up and Outcomes

During the administration of antibiotics, the patient reported symptomatic improvement of his ear pain, noticing a decrease in the intensity and frequency. Follow-up imaging showed radiologic improvement of white matter lesions on brain MRI done on 4-17-19. He continued to be followed as of 7-3-19, and he reported his last episode of ear pain around the end of May 2019 before it resolved completely. His only remaining symptom as of 7-3-19 was that of occasional mild tinnitus. There were no adverse events or unexpected outcomes as a result of treatment.

Discussion

The patient's description of intermittent shooting/stabbing unilateral ear pain is a unique presentation for LNB given the lack of any other typical Lyme disease symptoms. The patient showed no signs of discrete cranial nerve palsies, meningitis, or later signs such as encephalitis. The serologic and CSF testing were performed due to the white matter lesions shown on MRI; however, the location of the white matter lesions may not explain the patient's otalgia.[8] The ear pain may more likely be explained by the involvement of the sensory branches of cranial nerves V or VII.[9] Diagnosis of LNB requires objective evidence of Lyme disease infection via CSF pleocytosis and a positive antibody index, neurological symptoms, and a relationship between these symptoms. While there is some evidence of LNB through the positive CSF Borrelia antibody index as well as the parenchymal inflammation noted on MRI[10], in the absence of classical clinical signs such as erythema migrans or arthralgias and a lack of CSF pleocytosis, we cannot label this unusual presentation of symptoms as LNB, seeing as it is possible for the antibody index to remain positive for years after a prior Borrelia exposure. CSF testing for the Borrelia antibody index can be used to evaluate for CNS Lyme disease when a patient with known Lyme disease infection presents with neurologic symptoms. The difficulty in establishing a diagnosis of CNS Lyme disease by intrathecal antibody production is primarily due to the difficulty in distinguishing whether the presence of antibodies in CSF is due to antibody diffusion from serum to the CSF or from local production of Lyme antibodies intrathecally. The CSF Lyme antibody index can confirm if there is a higher concentration of antibodies in the CSF by the ratio of the CSF/serum antibody optical densities. A result of >1.3 suggests local production of Lyme antibodies intrathecally and is consistent with LNB. Lyme CSF PCR may be helpful in diagnosing CNS Lyme disease if positive, but because of its low sensitivity[11], it should not be the only criteria for diagnosis.[7] Lyme disease can be considered in the differential diagnosis for patients with tick exposures, living in Lyme endemic areas, with persistent and unexplained ear pain. Careful evaluation of patients with ear pain or other neurological signs and symptoms with MRI

and CSF testing is essential for proper disease management and treatment.

Research Quality and Ethics Statement

The authors of this manuscript declare that this scientific work complies with reporting quality, formatting, and reproducibility guidelines set forth by the EQUATOR Network. The authors also attest that this clinical investigation was determined to not require Institutional Review Board/Ethics Committee Review, and the corresponding protocol/approval number is not applicable. We also certify that we have not plagiarized the contents in this submission and have done a plagiarism check.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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