# Lyme Disease Associated With Postganglionic Horner Syndrome and Raeder Paratrigeminal Neuralgia

*Marjorie A. Murphy, MD, Erica M. Szabados, MD, and Jennifer A. Mitty, MD*

**Abstract:** A 55-year-old woman developed severe unilateral headaches, periocular numbness, and Horner syndrome after presenting with symptoms consistent with Lyme disease. The combination of Horner syndrome and periocular headache and numbness constituted a diagnosis of Raeder paratrigeminal neuralgia. Although the headaches resolved with antibiotic treatment, the Horner syndrome persisted for at least 1 year. This case expands the spectrum of neurologic manifestations of Lyme disease to include postganglionic Horner syndrome as well as Raeder paratrigeminal neuralgia.

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Lyme disease is a multisystem infection caused by the tick-borne spirochete *Borrelia burgdorferi*. It is the most common vector-borne disease in the United States and is endemic in Europe and Asia (1). Rhode Island has the second highest rate of Lyme disease infection in the United States (2) and is one of 12 states reporting an incidence of Lyme disease higher than the national average (3). These 12 states—Connecticut, Delaware, Maine, Maryland, Massachusetts, Minnesota, New Hampshire, New Jersey, New York, Pennsylvania, Rhode Island, and Wisconsin—account for 95% of cases reported nationally (3).

Early infection with Lyme disease consists of erythema migrans (stage 1), during which most patients present with a single (primary) lesion. Other patients have additional (secondary) skin lesions that are believed to arise through hematogenous dissemination from the site of primary infection (4). Disseminated infection (stage 2), which occurs days to weeks after the onset of disease, may also involve the nervous system, heart, or joints, and is followed within weeks or months by late stage infection (stage 3). Neurologic manifestations include lymphocytic meningitis, encephalitis, intracranial hypertension, cranial neuritis (particularly facial palsy), motor or sensory radiculoneuritis, cerebellar ataxia, and myelitis (1). In 1989, Glauser et al (5) reported a case of a reversible preganglionic Horner syndrome associated with Lyme disease.

We report a patient with Lyme disease and a persistent postganglionic Horner syndrome and Raeder paratrigeminal neuralgia, a previously unreported association.

## CASE REPORT

A 55-year-old woman from Rhode Island who had acquired Lyme disease 6 years earlier was referred for neuro-ophthalmologic evaluation of a right Horner syndrome. She had no prior history of migraine or cluster headache.

She had developed right-sided headache, anorexia, joint pains, fever, chills, and night sweats 2 months before presentation. She had removed a tick from her clothing shortly before these symptoms appeared but had not noted a tick bite. Within 4 days of the onset of her symptoms, she had also developed a rash on her lower extremities that was similar to the one she had had during her previous bout of Lyme disease. Her prior rash had been verified by a physician as being consistent with erythema migrans and early disseminated disease.

She was treated with a 6-day course of doxycycline [versus standard treatment for 14–21 days (1)] and experienced only slight improvement of her symptoms. Three weeks later she noted right upper lid ptosis and continued to have severe right-sided pain involving the right ear and the frontal, temporal, and vertex portions of the head. She noted numbness around her right eye and tenderness in the vertex area of her head. Two weeks later, a right Horner syndrome was diagnosed clinically by her internist.

MRI and MRA of the head and neck and a chest x-ray were normal. A lumbar puncture revealed one white cell, no red cells, and normal cerebrospinal fluid (CSF) glucose and protein levels. There were positive Lyme enzyme immunoassays in both the serum [index ratio of 6.56 (positive values greater than 1.1)] and the CSF

Departments of Ophthalmology (MAM), Neurology (EMS), and Infectious Disease (JAM), Rhode Island Hospital, Brown Medical School, Providence, Rhode Island.

Address correspondence to Marjorie A. Murphy, MD, Rhode Island Hospital, Department of Ophthalmology, APC 7, Providence, RI 02903; E-mail: margiemurphy@cox.net

[2.6 U/mL (positive values 0.8 U/mL or greater)]. Serum Lyme Western blot IgM (bands 23 and 41) and IgG (bands 18, 23, 28, 39, 45, and 58) were both positive. CSF Lyme Western blot IgM was negative, and CSF Lyme Western blot IgG showed reactivity to 4 of 10 *B burgdorferi* proteins. Although the CSF antibody was positive, the CSF/serum index was not consistent with intrathecal production of antibody.

Given her history of recent tick exposure, erythema migrans, inadequate initial treatment with doxycycline, and new neurologic findings, she began the standard treatment for neuroborreliosis with a 1-month course of 2 g intravenous ceftriaxone daily (1). She stated that her headache and joint pain resolved within 1 week of starting this treatment.

When we examined her 3 weeks into treatment, she continued to have right Horner syndrome and she acknowledged right forehead anhidrosis. Best-corrected visual acuity was 20/20 in both eyes at distance and near. Color vision, extraocular motility, confrontation fields, intraocular pressures, slit-lamp examination, and ophthalmoscopy were normal. Pupils measured 3 mm in the right eye and 5 mm in the left eye in very dim illumination and 2.5 mm in the right eye and 3.5 mm in the left eye in bright illumination. There was no afferent pupillary defect. There were 3 mm of right upper lid ptosis and 1 mm of right lower lid elevation ("inverse ptosis"). Decreased sensation was noted in the right V1 distribution involving the right periocular area and vertex of the scalp. Corneal sensation was intact bilaterally.

Instillation of 1% hydroxyamphetamine in both eyes produced dilation of the left pupil to 7 mm but no change in the size of right pupil, consistent with a diagnosis of postganglionic right Horner syndrome. On follow-up 9 months after presentation, the patient had a right Horner syndrome and right periocular and vertex numbness but had remained free of headache.

## DISCUSSION

This patient's constellation of postganglionic right Horner syndrome with right-sided headache, numbness in the first trigeminal segment, and scalp tenderness also involving that segment is consistent with Raeder paratrigeminal neuralgia. First reported by J.G. Raeder in 1924 (6), this syndrome has been associated with lesions in the middle cranial fossa and the gasserian ganglion, with etiologies including tumor, aneurysm, trauma, and infection (7). Given the onset of both our patient's headache and Horner syndrome concurrent with her other signs and symptoms of Lyme disease and her epidemiologic risk factors, it is likely that the Horner and Raeder syndromes are attributable to Lyme disease. The serology was difficult to interpret in this patient, but her previous history of Lyme disease and the fact that her headache and joint pain resolved within 1 week of starting intravenous antibiotic treatment support a diagnosis of neuroborreliosis.

Glauser et al (5) reported a patient with a reversible preganglionic Horner syndrome associated with Lyme disease. Similar to our patient, that patient had a positive Lyme serology along with a history of a tick bite and erythema migrans. That patient also had a lack of pleocytosis on lumbar puncture, and it was postulated that the Horner syndrome was caused by spirochetal invasion of the sympathetic pathway (5). It is possible that a similar mechanism caused the findings in our patient.

To our knowledge, neither Raeder paratrigeminal neuralgia nor postganglionic Horner syndrome has previously been reported with Lyme disease. This case expands the spectrum of neurologic manifestations associated with Lyme disease in individuals from endemic areas.

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