

A presentation of Lyme disease: pseudotumor cerebri

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Lyme disease is caused by a tick-transmitted spirochete, *B. burgdorferi*. It can present with both central and peripheral nervous system manifestations, including aseptic meningitis, meningoencephalitis, Bell's palsy and other cranial neuropathies, radiculoneuritis, and myelitis. However, pseudotumor cerebri associated with Lyme disease is rare. Here, we report a eight-year-old girl with the unusual manifestation of pseudotumor cerebri associated Lyme disease.

Key words: pseudotumor cerebri, Lyme disease, children.

Lyme disease is caused by a tick-transmitted spirochete, *Borrelia burgdorferi* that may have cutaneous, cardiac, rheumatologic and neurologic symptoms. Clinically apparent central nervous system (CNS) involvement which is consisted of aseptic meningitis or meningoencephalitis is seen in more than 10% of patients¹. Neuroborreliosis may present with unspecific signs and symptoms. Pseudotumor cerebri (PTC) is an unusual presentation of Lyme disease²⁻⁴. We reported a 8-year-old girl with PTC secondary to Lyme disease.

Case Report

A 8-year-old girl admitted to our hospital with a complaint of frontal headaches, vomiting and diplopia for two days. There was no history of fever, rash, joint symptoms, recent immunizations, tick bites, or exposures to infectious diseases. Her medical history was unremarkable.

Vital signs and physical examination were normal. The patient was afebrile, with neither rash nor meningismus. On neurological examination, her higher mental functions and upper-extremity motor and sensory examination were normal. There was bilateral papilledema and a left sixth cranial nerve palsy. The remainder of the physical and neurological examination were unremarkable.

The results of laboratory examinations (complete blood cell count, liver enzymes,

kidney function parameters, serum electrolytes, C-reactive protein, erythrocyte sedimentation rate) were normal. Magnetic resonance imaging (MRI) of the brain was normal. We performed lumbar puncture. The cerebrospinal fluid (CSF) showed no pleocytosis and normal protein and glucose concentrations. CSF opening pressure was 55 cm/H₂O. We diagnosed PTC and started to acetazolamide treatment at a dosage of 10 mg/kg.

Serum markers for herpes simplex virus, cytomegalovirus, varicella zoster, mumps, rubella, rubeola, Epstein-Barr viruses, and mycoplasma were all negative. Serologic evaluation of serum and CSF for *Borrelia burgdorferi* was positive for pathogen-specific IgM but negative for IgG. Serologic testing for *B. burgdorferi* was performed with an enzyme-linked immunosorbent assay (ELISA) and was confirmed by Western blot (Duzen Laboratories). The viro-immun micro ELISA test was used. The ELISA test was done by Triturus Grifols micro ELISA system (Spain). The specificity and sensitivity of test are 96.4% and 100%, respectively. The ELISA test, while not suitable for CSF, it can be used for human serum and plasma. Because of PTC secondary to Lyme disease, she was treated with ceftriaxone per day for the next 4 weeks. At the time of discharge on 1 month, her neurologic examination was normal. Serum was positive for both *B. burgdorferi* IgM and IgG antibodies on day 30 but only for IgG on

day 120(Table I).

Discussion

Pseudotumor cerebri is characterized by signs and symptoms of increased intracranial pressure, such as headache and papilledema, absence of an intracranial mass lesion or ventricular dilatation, usually normal findings on neurological examination except for papilledema, and an occasional abducens nerve palsy. PTC may be primary or occur secondary to certain conditions. In secondary cases, some conditions have been identified as: causative agents, including certain medications, endocrine abnormalities, autoimmune disorders, anemias, infectious or postinfectious processes and cranial venous outflow abnormalities⁵.

Lyme disease is an infectious disease caused by *Borrelia spirochetes*, bacteria that are usually

transmitted to human beings by *Ixodes* ticks. It is manifested by a wide spectrum of clinical symptoms that vary according to the time elapsed after infection. Various neurologic complications of borreliosis occur, including aseptic meningitis, meningoencephalitis, Bell's palsy and other cranial neuropathies, and radiculoneuritis^{1,6}. PTC in Lyme disease is a rare manifestation in pediatric patients. There have been only a few reports of PTC in association with borreliosis in children^{2-4,7-10}. Table II presents an analysis of cases of PTC with *B. burgdorferi* infection from the literature and our patient²⁻⁴. The association between Lyme disease and PTC was first described in 1985 in 2 children². All presented with headache, papilledema, and diplopia associated with sixth cranial nerve palsy. Eleven presented with systemic findings and signs of Lyme disease

Table I. Lyme Serologic Study Results

At diagnosis Serum and CSF were positive IgM (+), IgG (-) (both ELISA and WB)

On day 30 Serum was positive IgM (+), IgG (+) (ELISA)

On day 120 Serum was positive IgM (-), IgG (+) (ELISA)

ELISA: Enzyme-linked immunosorbent assay, WB: Western blot

Table II. Clinical and Laboratory Data of Lyme Disease with PTC in Children
ICP: Intracranial pressure

	Belman et al. ⁷	Ellermann and Hjelt ⁸	Jacobson and Frens ³	Jonsell ⁹	Raucher et al. ²	Wu et al. ¹⁰	Kan et al. ⁴	Our patient
Patients (n)	6	1	1	1	2	1	1	1
Headache	6/6	+	+	+	2/2	+	+	+
Diplopia	2/6	-	+	-	2/2	-	+	+
Abducens nerve palsy	2/6	-	+	-	2/2	-	+	+
Papilledema	6/6	+	+	+	2/2	+	+	+
Increased ICP	6/6	+	+	+	2/2	-	+	+
Pleocytosis in CSF	2/6	+	+	-	0/2		+	-
Increased protein in CSF	4/6	-	-	-	1/2	-	+	-
Neuroimaging abnormality	2/6	-	-	-	2/2	-	+	-
Systemic complaints (arthralgia/myalgia)	2/6	-	+	-	2/2	-	-	-

ICP: Increased intracranial pressure, CSF: cerebrospinal fluid

such as a viral type of illness, arthralgia, myalgia, arthritis, or erythema migrans before the onset of neurologic findings. By contrast, our patient presented with acute neurologic signs without systemic complaints. As similiar, Ellerman et al.⁸, Raucher et al.², and Kan et al.⁴ reported patients without systemic complaints. Six patients had pleocytosis and an increased CSF protein. Brain imaging studies, including CT and MRI, were abnormal in five cases. There have been reported slit-like ventricles, deep white matter changes, and enhancement of the dura²⁻⁴.

The pathophysiology of PTC in Lyme disease is undefined. At least two primary mechanisms for the development of increased CSF pressure in idiopathic intracranial hypertension have been postulated and supported by experimental data. The first mechanism is secondary to an autoimmune process that includes autoantibodies to myelin basic protein, autoreactive T-cell lines in CSF, and a shared antigenic determinant between *B. burgdorferi* and human tissue, including myelinated fibers of peripheral nerve, nerve cells, and axons of the central nervous system^{11,12}. The other mechanism is direct infectious or inflammatory processes¹³. Both mechanisms may ultimately impair the conductance of CSF flow and cause vasogenic extracellular brain edema. In patients without meningeal inflammation a postinfectious CNS autoimmune process as mentioned above may be contributory. It is postulated that some patients with neuroborreliosis without an inflammatory reaction may have been identified at the beginning of the infectious process in which no tissue infection or reaction had yet occurred¹⁴. In our patient, there was no pleocytosis in CSF analysis. As similiar, Jonsell⁹ reported a case with PTC without pleocytosis in CSF.

In conclusion, PTC is a rare *B. burgdorferi* related neurologic complication in childhood. Lyme disease should be included in the differential diagnosis of any child who presents with PTC. Even if the patient does not report or remember a tick bite, serum Lyme serologic study should be performed in endemic regions in any child with unexplained PTC.

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