Clinician's Corner

A 10-year-old female with unilateral seventh cranial nerve palsy

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CASE PRESENTATION

A 10-year-old previously healthy female presented to her local emergency department (ED) at the end of the summer with a right-sided facial droop. She came from her family cottage in eastern Ontario, where she had spent the past few months. Upon further questioning, she endorsed a 2-month history of intermittent myalgia, arthralgias and fevers. Two weeks prior to her ED visit she started having worsening symptoms with the addition of headache and abdominal pain. In the ED, she was diagnosed with a right-sided seventh cranial nerve palsy. A neurologist was consulted and she was sent home with a 1-week course of prednisone (60 mg daily) and acyclovir (400 mg four times daily), with some blood tests sent to try and identify the possible cause.

Over the next week, she developed increasing lethargy, with neck and back stiffness, as well as abdominal and facial pain and worsening headaches. Her paediatrician referred her to the tertiary care children's hospital for evaluation in the ED. At that point, her headaches were waking her up nightly for the previous week and were most severe in the morning and with activity, particularly when touching her toes. She did not report any vision changes, sick contacts or rashes. There was no recent history of travel outside the country and her immunizations were fully up to date. She presented febrile and tachycardic and on examination she had a right facial droop but other cranial nerves were normal. She had a positive Kernig and Brudzinski test, some diffuse abdominal pain but the rest of her cardio-respiratory, neurological and dermatology exam were normal. Further investigation combined with the follow-up of some initial testing sent from her local community ED revealed her diagnosis.

DISCUSSION

Serology sent from the local ED revealed that the Lyme IgG Western blot was nonreactive, IgM was reactive and her Lyme IgG/IgM enzyme-linked immunosorbent assay (ELISA) was reactive, suggestive of a recent infection with *Borrelia burgdorferi*, the causative agent in Lyme disease. Given her clinical meningismus, a lumbar puncture was performed. Cerebrospinal fluid (CSF) showed 3.08 g/L of protein (normal <0.6 g/L) and a white cell count of 235 cells/ μ L (normal <5 cells/ μ L) of which 61% were lymphocytes and 9% neutrophils. Bacterial culture plus molecular testing for herpes simplex virus and enterovirus were negative. Her abdominal pain responded well to intravenous ranitidine. Based on the positive serology, CSF cell count and clinical presentation, she was diagnosed with neuroborreliosis or early-disseminated Lyme disease with neurological involvement. The patient was prescribed a 4-week course of parenteral ceftriaxone at 75 mg/kg daily following which she returned to her baseline condition with no ongoing facial palsy.

Lyme disease is the most common tick-borne infection in North America and familiarity regarding its clinical presentations and appropriate treatment is important for physicians and patients alike (1–3). *B. burgdorferi* is a spirochete bacteria that is transmitted to humans through the bite of *Ixodes* genus ticks. Primary hosts for these ticks include small mammals, birds, white-tailed deer, mice and other small rodents.

Clinical manifestations of Lyme disease are typically divided into early-localized disease, early-disseminated disease and late disease. In early-localized disease, a rash at the site of the tick bite, termed erythema migrans (EM) is most common (1–3). EM is often described as a ‘bull’s eye’ with central clearing; however, it is more commonly uniformly erythematous (2). For human transmission to occur, the literature suggests the tick must be attached for at least 24 hours but often in children the tick itself may not be seen (1). In a clinically consistent case, Lyme disease should not be ruled out on the basis of no history of tick bite or EM rash. The CDC estimates 71% of patients with confirmed LD present with EM and another study showed that only 36% of children with LD had a recent history of tick bite (4). Thus, understanding potential exposure in an endemic area is important even if no tick bite was documented. The epidemiology of Lyme disease in Canada is changing and the number of reported cases is increasing in most parts of the country. Clinician knowledge of up to date local epidemiology including prevalence of Lyme disease in ticks is thus prudent and is available from the Public Health Agency of Canada and Provincial and Territorial health resources. By 2017, human cases of Lyme disease have been reported in all provinces of Canada, with the highest number of cases reported by Québec, Ontario and Nova Scotia (5). Most areas endemic for *Ixodes scapularis* ticks

are found in the southern parts of these three provinces plus southern New Brunswick and southeastern Manitoba (6).

The EM at the site of the bite often spontaneously resolves in four weeks but in early-disseminated disease the EM may progress to several widespread lesions due to dissemination of the bacteria (2). Other manifestations in this stage, as seen in our case, may include neurological signs such as peripheral facial nerve palsy, papilledema and meningeal symptoms such as headache and neck stiffness (1–3). It is estimated that as many as 9% of Lyme disease cases in children present with a facial palsy (7). The most frequent clinical manifestations in the different stages of the disease are listed in Table 1 (2–4,8).

Diagnosis is generally made with a positive *B. burgdorferi* serology in a clinically and epidemiologically consistent case (1). The Canadian Public Health Laboratory Network currently recommends a two-tier test method. The first tier uses a C6 ELISA. If the first tier test is positive or equivocal, the sample will be further tested for IgM and IgG specific antibodies using a Western blot assay. If only the first tier ELISA test is positive, Lyme disease is not diagnosed. In conjunction with a positive or equivocal ELISA, a positive IgM alone by Western blot in an acute case (i.e., <6 weeks post onset of symptoms) is consistent with a recent infection; a positive IgG alone by western blot is consistent with an infection of greater than 4 weeks duration, and if IgM and IgG are both positive the results indicate a recent or previous infection with *B. burgdorferi*. Not following the two-tiered testing approach increases the frequency of false-positive results and is not recommended (3).

When faced with a patient presenting with a facial palsy and a suspicion of Lyme disease, it is important for clinicians to assess if signs of neurological involvement are present, such as

Table 1. Main clinical manifestations of the different stages of Lyme disease

Clinical manifestation	Early localized	Early disseminated	Late disease
Single erythema migrans	+++	-	-
Multiple erythema migrans	-	+++	-
Fever	+	++	+
Fatigue and myalgia	++	+++	+
Headache	++	+++	+
Neck pain	+	++	+
Arthralgia	+	++	+++
Pauciarticular arthritis of large joints	-	-	+++
Carditis	-	+	-
Aseptic meningitis	-	+	-
Facial palsy	-	+	-
Papilledema and conjunctivitis	-	+	-
Peripheral neuropathy	-	-	+
Encephalitis	-	-	+

meningismus and severe headaches. Most experts recommend a lumbar puncture only in the presence of these signs (1,9,10). In patients with neuroborreliosis, lumbar puncture will generally reveal elevated white blood cells with lymphocytic predominance and elevated protein.

In patients with a facial palsy and positive serology for Lyme disease but without other neurological involvement, the recommended treatment is a course of oral amoxicillin 50 mg/kg/day in three divided doses (maximum, 500 mg three times a day) for 14 to 21 days. In children aged more than 8 years old, a regimen of oral doxycycline 4 mg/kg/day in two divided doses (maximum, 100 mg two times a day) for 14 to 21 days can also be considered (1). If a patient shows signs of neurological involvement and the lumbar puncture is positive with elevated white blood cells, the diagnosis of neuroborreliosis is confirmed. An intravenous regimen of ceftriaxone 75 mg/kg q24h (maximum, 2 g four times a day) for 10 to 28 days is then recommended (1). Prognosis of patients with neuroborreliosis if adequately treated is usually excellent (1,7,10,11).

CLINICAL PEARLS

- Even if there is no known history of a tick bite, it is important to establish if your patient has been in an area endemic for Lyme disease.
- If a patient comes from an area endemic for Lyme disease and presents with a facial palsy, we suggest clinicians consider Lyme disease and order blood serology.
- A lumbar puncture should be considered in patients with signs of neurological involvement of Lyme disease, such as

meningismus, severe headaches or other neurological symptoms beyond facial palsy. The result of the lumbar puncture is critical in determining the treatment agent and duration.

Consent for this publication was obtained from the patient and her parents.

References

1. Wormser GP, Dattwyler RJ, Shapiro ED, et al. The clinical assessment, treatment, and prevention of Lyme disease, human granulocytic anaplasmosis, and babesiosis: Clinical practice guidelines by the Infectious Diseases Society of America. *Clin Infect Dis* 2006;43(9):1089–1134.
2. Shapiro ED. Lyme disease. *N Engl J Med* 2014;371(7):684.
3. Onyett H; Canadian Paediatric Society, Infectious Diseases and Immunization Committee. Lyme disease in Canada: Focus on children. *Paediatr Child Health* 2014;19(7):379–88.
4. Gerber MA, Shapiro ED, Burke GS, Parcells VJ, Bell GL. Lyme disease in children in Southeastern Connecticut. Pediatric Lyme Disease Study Group. *N Engl J Med* 1996;335(17):1270–4.
5. Statistics Canada. Surveillance of Lyme disease [Internet] 2017; updated July 4, 2017; cited October 8, 2017. Available from: <https://www.canada.ca/en/public-health/services/diseases/lyme-disease/surveillance-lyme-disease.html> (accessed May 14, 2018).
6. Ogden NH, Koffi J, Pelcat Y, Lindsay LR. Environmental risk from Lyme disease in central and Eastern Canada: A summary of recent surveillance information. *Can Commun Dis Rep* 2014;40(5):40–5.
7. Hildenbrand P, Craven DE, Jones R, Nemeskal P. Lyme neuroborreliosis: Manifestations of a rapidly emerging zoonosis. *AJNR Am J Neuroradiol* 2009;30(6):1079–87.
8. Poehling KA, Edwards KM, Griffin MR, et al. The burden of influenza in young children, 2004–2009. *Pediatrics* 2013;131(2):207–16.
9. Morrison C, Seifter A, Aucott JN. Unusual presentation of Lyme disease: Horner syndrome with negative serology. *J Am Board Fam Med* 2009;22(2):219–22.
10. Halperin JJ, Shapiro ED, Logigian E, et al. Practice parameter: Treatment of nervous system Lyme disease (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology* 2007;69(1):91–102.
11. Jowett N, Gaudin RA, Banks CA, Hadlock TA. Steroid use in Lyme disease-associated facial palsy is associated with worse long-term outcomes. *Laryngoscope* 2017;127(6):1451–8.