Acute onset of facial nerve palsy associated with Lyme disease

*Vita Komen[1,2], Vladimira Vuletic[1,2]

[1] Clinic of Neurology, Clinical Hospital Center Rijeka, Rijeka, Croatia
[2] Department of Neurology, Faculty of Medicine, University of Rijeka, Rijeka, Croatia

Abstract

Lyme borreliosis (LB) is an anthropozoonosis, caused by different genospecies of the Borrelia burgdorferi sensu lato complex. The infection has been reported in countries throughout the Northern Hemisphere. Spirochetes circulate in small amounts in the blood. They can affect muscles, joints and nervous system. Clinical presentation of Lyme disease can include extreme fatigue, headache, stiff neck, muscle soreness, joint pain, swollen lymph nodes and sore throat. Recognition of an erythema migrans (EM) rash is very important as it is a hallmark symptom of LB. The stages of disease are labeled as early localized, early disseminated, and late disseminated. In areas endemic for Borrelia burgdorferi, Lyme neuroborreliosis (LNB) is estimated to cause 2-25% of peripheral facial palsy cases. Facial palsy in LB can present as part of a triad that includes radicular pain, cranial nerve involvement and lymphocytic pleocytosis in the CSF, or the palsy can be the sole manifestation of the disease. The current guidelines indicate that the diagnosis of LB is based on a two-tier serology at all stages of the infection. The effect of corticosteroid treatment in PFP patients with LB remains uncertain. The course, duration, and success of antibiotic therapy for LB varies according to the disease stage and site of disease manifestations.

Keywords: Lyme disease, facial paralysis, cerebrospinal fluid, serology, corticosteroids

Sažetak

Akutni početak paralize facijalnog živca povezan s lajmskom bolešću

Lajmska bolest (Lajmska borelioza, LB) je zoonoza uzrokovana spiralnim bakterijama iz kompleksa Borrelia burgdorferi sensu lato. Incidencija bolesti je zabilježena u zemljama cijele sjeverne hemisfere. Spirohete cirkuliraju u malim količinama u krvi te svoj učinak mogu ostvariti, između ostalog, i na mišiće, zglobove i živčani sustav. Klinička slika lajmske bolesti može uključivati malaksalost, glavobolju, ukočenost i bolnost u vratu, bolove u mišićima i zglobovima, uvećane limfne čvorove i grlobolju. Pravovremeno uočavanje osipa zvanog erythema migrans (EM) važno je jer je to karakteristični simptom LB. Stadiji bolesti dijele se na rano lokalizirani, rano diseminirani i kasno diseminirani. U područjima endemskim za bakteriju Borrelia burgdorferi, procjenjuje se da Lajmska neuroborelioza (LNB) uzrokuje 2-25% slučajeva pareze nervusa facialisa (ličnog živca). Paraliza lica u LB može se pojaviti kao dio trijasa koji uključuje radikularnu bol, zahvaćenost kranijalnih živaca i limfocitnu pleocitozu u likvoru, ili paraliza može biti jedina manifestacija bolesti. Sadašnje smjernice pokazuju da se dijagnoza LB temelji na dvostupanjskoj serološkoj dijagnostici. Učinak liječenja kortikosteroidima bolesnika s LB i perifernom parezom ličnog živca ostaje neizvjestan. Tijek, trajanje i uspjeh antibiotske terapije u liječenj LB varira ovisno o stadiju bolesti i mjestu manifestacije bolesti.

Ključne riječi: Lajmska bolest, pareza facijalnog živca, cerebrospinalni likvor, serologija, kortikosteroidi

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Correspondence:
Vita Komen
vita.komen@gmail.com

This article was submitted to RAD CASA - Medical Sciences as the original article

Conflict of Interest Statement: The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Received: 12 November 2024
Accepted: 2 December 2024
Published: 20 December 2024

Citation:
Komen V, Vuletic V. Acute onset of facial nerve palsy associated with Lyme disease
565=68-69 (2024): 82-86
DOI: 10.21857/yq32oh8gl9

Introduction

Lyme disease (LD) or Lyme borreliosis (LB) is an anthropozoonosis, caused by different genospecies of the Borrelia burgdorferi sensu lato complex. It is a zoonotic infection transmitted to humans through the bite of an Ixodid tick. Only three genospecies, Borrelia burgdorferi sensu stricto, B. afzelii, and B. garinii, have been related to LB. Borrelia burgdorferi sensu stricto is found in North America, while five species—B. afzelii, B. garinii, B. burgdorferi, B. spielmanii, and B. bavariensis—are found in Europe and Asia. Because of host specialization and tissue tropism, genospecies heterogeneity can reflect various clinical symptoms of LB. For the three major genospecies, different clinical symptoms have been identified, despite their commonality. In the United States, for instance, B. burgdorferi affects the joints, while B. garinii mostly affects the neurological system and B. afzelii primarily causes skin infections (1).

Lyme borreliosis has been reported in countries throughout the Northern Hemisphere. In Europe and Asia, the reported country-wide incidence ranges from low to negligible in the United Kingdom, Turkey and Japan, to >80 cases per 100,000 individuals in the Netherlands, Belgium, Austria, Slovenia, Lithuania and Estonia. Data on the incidence of Lyme borreliosis is scarce in Asia, although cases have been reported in China and Mongolia. A higher risk of infection is linked to occupations that promote tick exposure, such as forestry workers, hunters and hikers. There has been no evidence of infection through other routes of transmission, such as blood transfusion, sexual contact, urine, or breast milk (2).

Spirochetes circulate in small amounts in the blood. Depending on the case and genospecies, they can affect muscles, joints, organs, and nervous system, breaching the blood–brain barrier. It can also be transferred to the fetus during pregnancy (3).

In the United States, the age distribution of LB is typically bimodal, with peaks among children 5–15 years of age and adults 45–55 years of age. The incidence is higher among men than among women under 60 years old, but the sex ratio is nearly equal or slightly higher in women in older age groups. June and July are the months that have the highest rates of illness occurrence (2).

The annual incidence of peripheral facial palsy (PFP) is 20-53 per 100,000. In areas endemic to Borrelia burgdorferi, Lyme neuroborreliosis (LNB) is estimated to cause 2-25% of peripheral facial palsy cases. Numerous diagnoses, including Ramsay Hunt syndrome, sarcoidosis, Sjogren's syndrome, tumors, and acute idiopathic peripheral facial palsy, commonly referred to as Bell's palsy (BP), are responsible for the remaining cases. Of these, BP constitutes by far the largest group, causing 60-75% of cases of PFP (4). The underlying etiopathogenesis remains unknown, although some authors suggest the reactivation of herpes simplex virus. Although BP can happen at any age, it is most common between the ages of 15 and 45 (5).

Stages of disease

Both innate and adaptive immune responses are capable of identifying and eliminating B. burgdorferi. It doesn't produce toxins or extracellular matrix-degrading proteases, so most of the manifestations of human Lyme borreliosis at each of the three stages of disease result from inflammation generated by these immune responses (2).

Lyme disease includes a large group of symptoms such as extreme fatigue, headache, stiff neck, muscle soreness, joint pain, swollen lymph nodes and sore throat (3). Recognition of an erythema migrans (EM) rash is very important as it is a hallmark symptom of LB, even when the patient does not recall the tick bite. According to researchers, only 20% to 60% of people with Lyme disease recall being bitten by a tick (3). Therefore, it is crucial to emphasize that LB is not excluded if there is no history of a tick bite. Although EM centrifugal evolution is the most crucial diagnostic criterion, there is considerable variation in its clinical presentation (1).

The stages of disease are labeled as early localized, early disseminated, and late disseminated. The infection typically begins during summer with erythema migrans (stage 1), which occurs at the site of the tick bite. Malaise, exhaustion, headache, arthralgia, myalgia, fever, and regional lymphadenopathy are frequently present in patients with erythema migrans. The B. burgdorferi strains spread from the tick bite site to other parts of the body in a matter of days to weeks. During early disseminated infection (stage 2), patients might develop acute Lyme neuroborreliosis. The most prevalent clinical manifestations of Lyme neuroborreliosis in the US include motor or sensory radiculoneuritis, cranial neuropathy (especially facial palsy), and lymphocytic meningitis with intermittent headaches and mild neck stiffness. Tick-borne meningopolyneuritis, also known as Bannwarth syndrome, is brought on by B. garinii infections in Europe. It starts with painful radiculoneuritis linked to lymphocytic meningitis and can be followed by cranial neuropathy or paresis of the extremities. Mostly in the US, arthritis is a defining feature of late-stage, stage 3 Lyme borreliosis. The most common late symptom of Lyme borreliosis in Europe is acrodermatitis chronica atrophicans (2).

Neurologic involment

Involvement of the nervous system occurs in up to 15% of patients with untreated LB. One of the most common symptoms is a headache. There may be involvement of the cranial nerves, especially the facial nerve (80%). There is evidence of ocular neuritis and paralysis of the III, IV, and VI cranial nerves. Meningopolyneuritis (Bannwarth syndrome) with radicular pain and sometimes paresis of extremities or the abdominal wall, neurologic bladder, and paresthesia can be observed. Speech problems, recent cognitive and emotional issues, mental health conditions, anxiety, depression, and panic attacks can all be linked to neuroborreliosis (1).

Facial palsy

Abnormalities of the cranial nerves are probably the most common focal nervous system abnormality in Lyme neuroborreliosis. Facial nerve palsy has been found in 11% of patients with Lyme disease. Facial palsy in LB might be the only symptom of the condition, in which case it is challenging to differentiate it from Bell's palsy. It can also occur as a component of a triad that involves cranial nerve involvement, radicular pain, and lymphocytic pleocytosis in the CSF (5). The incidence of LB is higher in children with PFP than in adults with PFP. Bilateral occurrence is a special feature of the PFP caused by LB, in 18% to 25% of the cases (6).

Even in the absence of antibiotic treatment, 98% of patients with facial palsy recover completely or almost completely, indicating an excellent prognosis (7). However, 16% to 23% of patients will have residual deficits (8).

Lyme disease can lead to Bell's palsy via swelling and impingement of the seventh cranial nerve as it tracks through the narrow bony Fallopian canal in the skull beneath the ear. Bell's palsy from Lyme disease is usually considered occurring in the early disseminated disease. Clark et al. found that the erythema migrans rash precedes Bell's palsy by a median of 20 days and 81% of patients have the onset of Bell's palsy within 4 weeks of the appearance of the rash (9).

Lyme disease should be considered in any patient with a new onset of Bell`s palsy. But making the right diagnosis is a challenge in itself. Patients with facial palsy caused by Lyme disease can present without any other signs or symptoms, and Bell`s palsy might be the first diagnosis that comes to mind. If left undetected, it could lead to early or late disseminated Lyme disease, which is more difficult to treat (10).

A journey to diagnosis

Unlike many other infectious diseases, where detection by culture, PCR, or serology is usually more dependable and definitive, LB is more difficult to diagnose.

Cerebrospinal fluid (CSF) analysis in the majority of patients shows lymphocytic pleocytosis, damage to the blood brain barrier, and an intrathecal synthesis of immunoglobulin IgM, IgG, and sometimes IgA. During facial nerve palsy, the CSF frequently exhibits lymphocytic pleocytosis even in the absence of signs and symptoms of meningitis. After the onset of neurological symptoms, for a short time, intrathecal synthesis may not be detectable and CSF pleocytosis may be absent. Even after recovery, intrathecal antibody production may remain. Though it has low sensitivity in the very early stages of the disease, intrathecal antibody production is typically used for a definitive diagnosis. In general, patients with LNB display intrathecally produced antibodies within two weeks of the onset of symptoms, but in some patients antibody production may be delayed for up to six weeks (3).

According to the guidelines, a two-tier serology is currently used to diagnose LB at all stages of the infection, except of erythema migrans. The two-tier testing procedure includes ELISA as the first test and a Western Blot/Immunoblot assay as a confirmatory test. The test kit manufacturers define the interpretation for positive, negative, and equivocal samples. Although a set of eight bands was identified as significant in each participant laboratory, no guidelines were developed for usage throughout Europe. According to one meta-analysis, the serologic test's mean sensitivity was 59.5% (range: 30.6–86.2%).

The only reliable and accessible way to confirm a Lyme borreliosis diagnosis is through serological testing. During the initial weeks of infection, when the majority of Lyme borreliosis patients are diagnosed with erythema migrans, serodiagnostic tests are insensitive. During acute, early infection, 20–50% of patients have positive responses, typically to the IgM isotype. During the convalescent period at the end of 2–3 weeks of antibiotic treatment, 70–80% of patients have seroreactivity, still usually of the IgM isotype. However, after 4 to 8 weeks of untreated infection, almost all patients have IgG antibody responses.

IgG and even IgM antibodies against B. burgdorferi might persist for months or years after near or complete spirochetal elimination with antibiotics, which is the principal drawback of serological testing. Although the amount of antibody gradually decreases after treatment, results from the non-quantitative western blot test do not significantly alter in the post-antibiotic phase. Thus, serological testing cannot be used to determine active infection or the adequacy of antibiotic therapy (2).

It is also possible to perform a PCR for the detection of Borrelia DNA in CSF as well as an ELISA for Chemokine 13. But, for now, these methods are solely used in research studies (1).

There are few studies on CSF findings in peripheral facial palsy. One study systematically researched CSF in 509 patients and the usefulness of this approach in differential diagnosis was assessed. Of 383 patients with idiopathic palsy, 25.6% had a compromised CSF/blood–brain barrier (BBB), and 7.3% had pleocytosis. Weber et al. studied CSF samples of 59 patients with BP and found an impaired BBB in 15%, whereas pleocytosis was detected in approximately 12% (5).

Regarding the best way to diagnose Lyme facial nerve palsy, there is a lot of disagreement. On CSF investigation, the majority of patients show signs of a CNS infection; but specific marks predictive of a poor prognosis are lacking (11).

Corticosteroids - yes or no?

Adults with Bell's palsy benefit from corticosteroid treatment given within 72 h after the onset of the palsy, but there is no reliable evidence that this treatment is beneficial or harmful for LB patients who require antibiotic treatment. The effect of corticosteroid treatment in PFP patients with LB remains uncertain. Although corticosteroids reduce inflammation, they can also

weaken the immune system and increase the spirochete load (5). In contrast, individuals may experience serious late sequelae if they do not receive the proper treatment (12).

No established role for corticosteroids in the setting of acute LB exists. Two retrospective studies have led to the consensus that corticosteroid treatment in LDFP patients receiving appropriate antibiotics has not demonstrated any discernible benefit or harm. A study published by Clark et al., demonstrated no difference in facial outcomes among 101 patients with LDFP who received antibiotics alone, both antibiotics and steroids, steroids alone, or no treatment whatsoever. One limitation of this study was that the follow-up length was not adequately documented; the text implied that the majority of patients were monitored for less than six months. A more recent retrospective cohort study of 51 patients with LDFP found that patients who had received antibiotics and corticosteroids had worse outcomes compared with patients who had received antibiotic therapy alone (3).

The possible lack of benefit from corticosteroids in LDFP may imply that the mechanism of action of corticosteroids is not simply a reduction in nerve swelling within a closed compartment. Conversely, if that is the mode of action, the kind of inflammation linked to facial neuropathy in Lyme disease patients might be different from that of Bell's palsy patients. Treatment with corticosteroids would probably have varying effects on different forms of inflammation. Steroids might make the infectious process in LDFP worse (13).

Antibiotic therapy

Depending on the illness stage and location of symptoms, antibiotic therapy for LD has different courses, duration, and success rates. However, oral doxycycline, amoxicillin, cefuroxime, or intravenous ceftriaxone are the usual therapeutic options. Additional research has shown that antibiotic therapy works best when it is administered early in the course of the illness, and that treatment failure is more likely to occur when a diagnosis is delayed. Patients treated within 4 weeks of the onset of the disease had a higher chance of a successful treatment outcome, while those treated later experienced a higher frequency (up to 50% or more) of cognitive and musculoskeletal impairment, according to retrospective studies of LD patients conducted at least a year after treatment (2).

Patients with neurological or cardiac involvement of Lyme borreliosis can typically be successfully treated with the same oral antibiotics used to treat erythema migrans, depending on the severity of the condition. However, with neurological or cardiac manifestations, a parenterally administered antibiotic might be used in certain cases. The most commonly prescribed parenterally administered antibiotic is ceftriaxone, as it is administered once a day, but cefotaxime and intravenous penicillin are also highly effective. Antibiotics are useful in reducing subsequent clinical sequelae in patients with seventh nerve palsy, but they do not speed up the recovery of facial weakness.

Most Lyme borreliosis patients respond well to antibiotic therapy and recover fully, regardless of the disease's presentation (2).

Conclusion

Peripheral facial palsy is a frequent manifestation of early LNB, and it should be included in the differential of patients presenting with acute onset peripheral facial palsy. When assessing patients with peripheral facial palsy, it is important to actively look for the numerous hints from the medical history and physical examination that aid in the diagnosis of LNB. When a patient has bilateral facial nerve palsy, LNB should always be included in the differential diagnosis. Antibiotic medication is an effective treatment, and the majority of patients will restore facial nerve function. Neither the overall result nor the rate of recovery seems to be impacted by adjunctive corticosteroid medication.

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