BORRELIOSIS, A POSSIBLE CAUSE OF FACIAL PALSY.
MINI REVIEW

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ABSTRACT

Bell’s palsy (idiopathic facial paralysis) was considered for a long time as the most common cause of facial paralysis but the developing methods of investigation tend to decrease the rate of idiopathic cases bringing into focus pathogens like Borrelia burgdorferi that causes Lyme disease. Although neuroborreliosis can be difficult to diagnose especially when facial palsy is the only sign, for better outcomes, practitioners should keep in mind this possible etiology in order to avoid mistaking Lyme associated facial palsy with Bell’s palsy. While antibiotic treatment (not necessary for Bell’s palsy) is needed to prevent progression of disseminated borreliosis in cases of Lyme disease related facial palsy, corticosteroids, which are the first therapy choice for Bell’s palsy, have been associated with worse outcomes in Lyme disease facial palsy.

Keywords: acute facial palsy, Borrelia burgdorferi.

RéSUMÉ

Maladie de Lyme – cause possible de la paralysie faciale

La paralysie faciale idiopathique a longtemps été considérée comme la cause la plus fréquente de paralysie faciale, mais les méthodes de recherche en développement tendent à diminuer le taux de cas idiopathiques mettant en évidence des agents pathogènes tels que Borrelia burgdorferi qui cause la maladie de Lyme. Bien que la neuroborreliose soit difficile à diagnostiquer, surtout lorsque la paralysie faciale est le seul signe, de meilleurs résultats, les praticiens devraient garder à l’esprit cette étiologie possible afin d’éviter de confondre Lyme associée à la paralysie faciale avec la paralysie de Bell. Alors que le traitement antibiotique (non nécessaire pour la paralysie de Bell) est nécessaire pour prévenir la progression de la borreliose disséminée dans les cas de paralysie faciale liée à la maladie de Lyme, les corticostéroïdes, qui sont le premier choix thérapeutique pour la paralysie de Bell, ont été associés aux pires résultats dans la maladie de Lyme paralysie faciale.

Mots-clés: paralysie faciale aiguë, Borrelia burgdorferi.

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ABOUT FACIAL PALSY AND LYME DISEASE

Facial palsy is an usually suddenly installed inability, with several degrees, to move facial muscles on one side of the face, due to cranial nerve VII dysfunction. Patients or the entourage notice the facial asymmetry, the patient complains of difficulties in mastication, impossibility to whistle, impossibility to close the eyelid that leads to dryness of the eye and potential eye injury. Peripheral facial palsy occurs in the general population, with an annual incidence of 20.25 per 100,0001-4.

Facial palsy can have various etiologies like bacterial or viral infections, trauma, tumors, neurological diseases that cause nerve demyelination5,6. Bell’s palsy (idiopathic facial paralysis) was considered for a long time as the most common cause of facial paralysis but the developing methods of investigation tend to decrease the rate of idiopathic cases bringing into focus pathogens like Borrelia burgdorferi that causes Lyme disease.

Lyme disease is a multisystem illness caused by a tick-borne spirochete7 that develops when the bacteria disseminate from the local tick bite to different organs and tissues8. Lyme disease, the most common tick-borne disease in the United States, occurs most commonly between April and October9. Neuroborreliosis (NB) is the neurological manifestation of Lyme disease and may affect both the peripheral and the central nervous system, including neurologic conditions such as cranial-nerve (particularly facial-nerve) palsy and meningitis that mimics aseptic meningitis8,10,11.

Lyme disease can occur at any age, but is most common in children aged 5 to 10 years9,12,13. Peripheral facial palsy may be the only sign of Lyme borreliosis in children14. The practitioners should consider this etiology when dealing with idiopathic facial paralyses, especially in endemic areas during summer months, or when bilateral15.

The percentage of facial palsy caused by Borrelia infection varies between 6% according to a study performed in Finland in 199216 and 50% according to Cook et al. A study from the Department of Infectious Diseases, University Medical Centre in Ljubljana, published in 1999, found a percentage of Borrelia infection causing facial palsy of 19.3 of the analyzed cases17, while Cook et al concluded, after a study published in 1997, that Lyme disease may be the cause for up to 50% of all facial nerve paralyses in children for regions with endemic infections caused by Borrelia burgdorferi17.

Lyme disease was first reported in 1975 in the region Lyme, Connecticut, where a high number of children were diagnosed with a disease resembling juvenile arthritis6,18. It is a zoonosis transmitted to humans by ticks that feed on both humans and small mammals (like mice) or birds especially during the nymph stage, when, due to the small size, they go unnoticed19,20. Ticks may have appeared 225 million years ago, when they parasitized on reptiles21, but the connection between the tick and Lyme disease was made in 1982 when Willy Burgdorfer isolated the spirochete Borrelia from the blood of two patients22. Borrelia species are gram-negative microaerophilic mobile spirochetes8.

Lyme disease is transmitted to humans by the bite of infected ticks of the Ixodes genus, the deer tick Ixodes scapularis or the deer tick Ixodes pacificus23. Borrelia burgdorferi, the organism responsible for Lyme disease, is a spirochete23. In Europe and in Asia, Borrelia afzelii, Borrelia garinii, and other related species, in addition to Borrelia burgdorferi, cause Lyme disease24. Usually, the tick must be attached for 36 to 48 hours before the spirochetes can spread25, which is possible due to small size and secretion of a complex cocktail of salivary components that prevent hemostasis at the feeding site and inflammatory reactions that may trigger the sensation of pain or itching on the host26.

The natural progression of Lyme disease has 3 stages of infection8.

In the first stage, the early localized Lyme disease, a skin infection appears in 3 to 30 days after tick bite1,2,27,28. Erythema migrant rash occurs in approximately 70 to 80 percent of infected persons29. The lesion is an expanding circular red rash30,31, of at least 5 cm, that may have centrally located vesicles or necrotic areas30,32,33, sometimes having a bull’s-eye appearance30,35. In children, the rash is commonly found in the head and neck region36,38. Flu-like symptoms may accompany the local infection including fatigue, headache, arthralgia, myalgia, and, fever8,30,34,35,39.

In the second stage, early disseminated Lyme disease, developing within weeks to months after a tick bite, the spirochete disseminates to different areas of the skin or in myocardium, spleen, liver, muscle, bone and retina40. Extra cutaneous signs of disseminated Lyme disease that may occur, with or without erythema migrans, include neurologic conditions, such as cranial-nerve (particularly facial-nerve) palsy and meningitis that mimics aseptic meningitis, as well as carditis10,11.

The third stage, late disseminated Lyme disease, is uncommon but develops in untreated cases more than six months after a tick bite. Late neuroborreliosis and Lyme arthritis, a relapsing arthritis that usually is pauciarticular, most often affecting the knee, may develop41. Acrodermatitis atrophicans (ACA) can also occur predominantly in adults24,42.
Lyme disease is first suspected based on clinical findings. Considering that peripheral facial palsy may be the only sign of Lyme borreliosis, if the practitioners do not keep in mind this possible etiology, Lyme associated facial palsy can be easily mistaken with Bell’s palsy. It is important to early diagnose borreliosis because antibiotic treatment could prevent progression to other secondary or tertiary stage manifestations. In cases of acute isolated facial palsy, antibiotics are considered to prevent disease spreading but the influence on the clinical course of the facial palsy is not proved.

Diagnosis:

In US a 2-step approach is recommended. First, a screening Lyme IgM and IgG detection by enzyme immunoassay (EIA) or immunofluorescent antibody (IFA) assay should be performed. A positive or an equivocal result should be confirmed by a standardized Western immunoblot for presence of IgM and IgG antibodies against specific B. burgdorferi proteins because normal oral flora, other spirochetal infections like syphilis or leptospirosis, viruses like Epstein-Barr virus, may give false positive results. Lyme IgM peaks three to six weeks and IgG peaks weeks to months following tick bite. The production of IgG may last for years after the infection has disappeared.

Intrathecal antibody production is generally used for a definite diagnosis, but it has low sensitivity in the very early phase of the disease. In some patients, antibody production may be delayed for up to six weeks. PCR and culture have high sensitivity on skin samples of patients with EM but PCR on material obtained from extra cutaneous sites is in general of low sensitivity, with the exception of synovial fluid, so that both the culture of CSF for Borrelia and specific PCR analysis from CSF have a very low sensitivity and are not used in clinical routine.

For the diagnosis of neuroborreliosis, in Europe, both serum and CSF antibodies are required. The CSF/serum Borrelia sensu latu antibody index (AI) can give important information from the first week after onset of neurological symptoms. In children, confirmed neuroborreliosis requires clinical symptoms suggestive of neuroborreliosis with cranial neuropathy as the main manifestation, CSF pleocytosis and a positive Borrelia AI index.

Regarding treatment:

Treatment with doxycycline, amoxicillin, or cefuroxime is safe and highly efficacious for early Lyme disease. Recommendations of the Red Book Report of the Committee on Infectious Diseases from 2009, for treatment of isolated facial nerve palsy, without signs of meningitis, are orally administered doxycycline, amoxicillin, or cefuroxime for 21 to 28 days in order to prevent late disease. It is also mentioned that antibiotics have no effect on the resolution of the facial palsy and that corticosteroids are not indicated. For neuroborreliosis involving the central nervous system, parenteral treatment with ceftriaxone, cefotaxime or penicillin G have been used.

Most studies reported corticosteroids to be efficient and the treatment of choice for Bell’s palsy, but their role in facial palsy associated with Lyme disease is controversial. A 10-20-year follow-up study of patients who suffered of Lyme disease, reported no difference in long-term outcome between those who had received steroids for Lyme related facial palsy and those who had not. In contrast with these findings, a recent study showed a harmful effect of steroids in patients with Lyme disease-associated facial palsy. The facial function scores on long-term recovery (an average of 15 months) of 51 patients with Lyme disease-associated facial paralysis, demonstrated worse outcomes for those who received corticosteroids as part of their initial treatment. Both these findings and the recommendations for antibiotic treatment in facial palsy associated with Lyme disease argue the need to differentiate between Bell’s palsy and facial palsy as manifestation of neuroborreliosis. In an attempt to early differentiate between these entities, a study on cerebrospinal fluid parameters in patients with peripheral facial palsy, found that the time of the year, associated neurological symptoms and mononuclear pleocytosis were strong predictive factors for Lyme neuroborreliosis as a cause of peripheral facial palsy in an area endemic for Borrelia.

Conclusions

Although neuroborreliosis can be difficult to diagnose, especially when facial palsy is the only sign, for better outcomes, practitioners should keep in mind this possible etiology in order to avoid mistaking Lyme associated facial palsy with Bell’s palsy. While antibiotic treatment (not necessary for Bell’s palsy) is needed to prevent progression of disseminated borreliosis in cases of Lyme disease related facial palsy, corticosteroids, which are the first therapy choice for Bell’s palsy, have been associated with worse outcomes in Lyme disease facial palsy. Further studies are needed to conclude on the actual proportions of Borrelia species involvement in acute facial palsy and starting from these conclusions to make recommendations for diagnostic procedures and...
treatment when dealing with a initially considered Bell’s palsy.

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