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Peripheral facial nerve palsy as a manifestation of neuroborreliosis in children

Obwodowe porażenie nerwu twarzowego jako objaw neuroboreliozy u dzieci

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Abstract

Lyme disease, also known as Lyme borreliosis, is a systemic infectious disease caused by *Borrelia burgdorferi* spirochetes. It is spread by the bites of infected ticks of the genus *Ixodes* which inhabit forest plants in the northern hemisphere, including western Asia and Europe. In the classic course of Lyme disease, the most common early manifestations include erythema migrans, a circular red skin rash around the tick bite, and general symptoms. The most prevalent clinical manifestation of late disease is neuroborreliosis. The migration of urban residents to regions endemic to ticks of the genus *Ixodes* which are vectors of *Borrelia burgdorferi* increases the risk of neuroborreliosis in all children regardless of their place of residence, especially during the summer and early autumn. The absence of characteristic manifestations in the first stage of the disease, shortly after the tick bite, involving typical skin lesions and systemic symptoms, may result in delayed diagnosis and initiation of causal treatment. In adult patients, the disease appears typically as meningoradiculoneuritis. In children, common manifestations associated with late-stage Lyme disease are meningitis and cranial nerve palsy including peripheral facial nerve palsy. Symptoms of neuroborreliosis are uncommon in the early stage of the disease. Facial nerve palsy can be a diagnostic symptom of multiple nervous system disorders, including infectious diseases. Neuroborreliosis is a form of Lyme disease that affects the structures of the nervous system, and may present as peripheral facial nerve palsy. Consequently, children with peripheral facial nerve palsy should undergo diagnostic work-up for *Borrelia burgdorferi* infection also in cases without typical erythema migrans in the early stage of the disease.

Keywords: neuroborreliosis, children, facial nerve

Streszczenie

Borelioza lub krętkowica kleszczowa, nazywana także chorobą z Lyme, jest układową chorobą zakaźną powodowaną przez krętki *Borrelia burgdorferi*. Do infekcji dochodzi po ukąszeniu przez kleszcza z rodzaju *Ixodes*, bytującego na roślinach leśnych półkuli północnej, w tym zachodniej Azji i Europy. W klasycznym przebiegu boreliozy wczesne objawy występują najczęściej pod postacią wędrującego rumienia, usytuowanego koncentrycznie wokół miejsca ukąszenia, i symptomów ogólnych. Neuroborelioza jest najczęstszą postacią późnego etapu choroby. Przemieszczanie się mieszkańców miast w rejony endemicznego występowania kleszcza z rodzaju *Ixodes*, cechujące się związaną z tym możliwością zakażenia *Borrelia burgdorferi*, zwiększa ryzyko wystąpienia neuroboreliozy u wszystkich dzieci, niezależnie od miejsca zamieszkania, szczególnie w okresie lata i wczesnej jesieni. Brak typowego przebiegu pierwszego etapu choroby występującego wkrótce po ukąszeniu kleszcza, któremu towarzyszą typowe zmiany skórne oraz objawy ogólnoustrojowe, może mieć wpływ na późne rozpoznanie choroby i wdrożenie leczenia przyczynowego. U dorosłych choroba ta objawia się najczęściej pod postacią zapalenia opon mózgowo-rdzeniowych i nerwów rdzeniowych (*meningoradiculoneuritis*). U dzieci natomiast w późnym okresie boreliozy typowe jest zapalenie opon mózgowo-rdzeniowych i porażenie nerwów czaszkowych, w tym obwodowe porażenie nerwu twarzowego. Objawy neuroboreliozy rzadko występują we wczesnym okresie choroby. Porażenie nerwu twarzowego może być wskaźnikowym objawem licznych chorób układu nerwowego, w tym chorób zakaźnych. Neuroborelioza jest postacią choroby, która dotyczy struktur układu nerwowego i może się objawiać obwodowym porażeniem nerwu twarzowego. Należy więc pamiętać, że u dzieci z obwodowym porażeniem nerwu twarzowego konieczna jest diagnostyka w kierunku infekcji *Borrelia burgdorferi*, także w przypadkach przebiegających bez typowego rumienia wędrującego we wczesnym etapie choroby.

Słowa kluczowe: neuroborelioza, dzieci, nerw twarzowy

Lyme disease, also known as Lyme borreliosis, is a systemic infectious disease caused by *Borrelia burgdorferi* spirochetes. It is also the most common infectious disease in humans which is transmitted by arachnids. Lyme disease is spread by the bites of infected ticks of the genus *Ixodes* which inhabit forest plants in the northern hemisphere, including western Asia and Europe.

The migration of urban residents to regions endemic to ticks of the genus *Ixodes* which are vectors of *Borrelia burgdorferi* increases the risk of neuroborreliosis in all children regardless of their place of residence, especially during the summer and early autumn[1,2].

In the classic course of Lyme disease, early manifestations include a distinctive bull's-eye rash located around the tick bite (erythema migrans) or – less commonly – lymphocytic infiltrate. Skin lesions are usually accompanied by general symptoms such as headaches, joint and muscle pain, high or low-grade fever, and weakness. The above symptoms resolve within 3 months, when some of the infected patients present with signs of late-stage disease including inflammation of large joints, frequently neurological disturbances, and occasionally myocarditis. Neurological symptoms are also characteristic of the later – chronic – period of the disease, occurring many years after infection. At that stage, they coexist with chronic arthritis and atrophic dermatitis[3].

Neuroborreliosis is the most prevalent clinical manifestation of late disease. In adult patients, neuroborreliosis appears typically as meningoradiculoneuritis. In children, common manifestations associated with late-stage Lyme disease are meningitis and peripheral facial nerve palsy. Symptoms of neuroborreliosis are uncommon in the early stage of the disease[4,5].

In children, the clinical picture of neuroborreliosis tends to vary widely. Neuroborreliosis has been reported to be accompanied by damage to other cranial nerves, often involving the motor nerves of the eyeball and the optic nerve[6–9]. Other reports have included acute, frequently transient ischaemic central nervous system disorders as well as acute cerebellar symptoms, and isolated intracranial hypertension[10–12].

Neuroborreliosis in children often presents with symptoms of meningitis, sometimes with partial seizures[13]. It is also thought that meningitis in children in endemic areas is most typically caused by *Borrelia burgdorferi* infection[14]. Occasionally, meningitis is accompanied by encephalitis[15]. However, *Borrelia burgdorferi* infection is also often confirmed in children who present only with apathy and headaches[16].

Facial nerve palsy can be a diagnostic symptom of various diseases of the nervous system, including proliferative disorders of the central nervous system, systemic disorders, vascular diseases, congenital anomalies, injuries, polyneuritis and infections. It can also be a result of iatrogenic damage after parotid and middle ear surgery in children[17].

Facial nerve palsy secondary to neuroborreliosis is very often reported in patients of developmental age, with prevalence reaching 50% in these cases[18]. *Borrelia burgdorferi* infection is also recognised as the most common cause of infectious peripheral facial nerve palsy in children, occurring more frequently than infections caused by the chickenpox virus, influenza A virus, herpes virus, enteroviruses and *Mycoplasma pneumoniae*[19]. Tveitnes and Øymar suggest that facial nerve symptoms are very common in late Lyme disease. According to these authors, facial nerve palsy was accompanied by headaches and neck aches more frequently in boys than in girls. Also, boys diagnosed with neuroborreliosis with peripheral facial nerve palsy were statistically older, and had higher levels of inflammation markers determined in blood plasma and cerebrospinal fluid (CSF)[5]. There have also been reports of bilateral facial nerve palsy in late-stage Lyme disease in children[18].

The absence of characteristic manifestations in the initial phase of the disease, shortly after the tick bite, including typical skin lesions and systemic symptoms, may result in delayed diagnosis and initiation of causal treatment. In some cases, early diagnosis of Lyme disease is hindered by negative results of immune tests for *Borrelia burgdorferi* infection[4].

A key aspect in the diagnosis of neuroborreliosis in paediatric cases is obtaining medical history from caregivers and – if possible – also from children, focusing on reports of a tick bite followed by erythema migrans and general symptoms. Typical physical examination findings in patients with neuroborreliosis also include meningitis and cranial nerve palsy, though these occur mainly in the second stage of untreated disease, a few months after the appearance of skin manifestations. Additional examinations include basic blood plasma tests with the assessment of white blood cell count and proportions, and acute inflammatory phase markers as well as Elisa and Western Blot tests to detect IgG and IgM antibodies against *Borrelia burgdorferi*. An important role is also attributed to the results of CSF analysis revealing significant mononuclear pleocytosis and the presence of IgG and IgM antibodies against *Borrelia burgdorferi*. Occasionally, CSF in children with neuroborreliosis also shows prominent eosinophilia[20]. CSF markers – including CSF-CXCL13 – have also been suggested as helpful in the diagnostic work-up for Lyme disease[21]. Computed tomography and magnetic resonance imaging findings show no abnormalities[7].

Neuroborreliosis should be suspected in all children with headache growing in intensity for more than 7 days, peripheral facial nerve palsy, and the proportion of mononuclear cells in the CSF assessment exceeding 70% (so-called "rule of sevens")[22].

American and European standards for the causal treatment of neuroborreliosis with peripheral nervous system symptoms recommend intravenous ceftriaxone therapy or oral doxycycline treatment in children aged over 8–9 years as equally effective methods regardless of the severity of disease manifestations[23]. An effective course of antibiotic therapy should last 2–4 weeks[23,24]. Blin-Rochemaure

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and Quinet suggest oral antibiotic therapy in children over 8 years of age with neuroborreliosis accompanied by facial nerve palsy without CSF abnormalities. In turn, intravenous antibiotic therapy is advised in cases of neuroborreliosis with changes found in CSF[25]. The term "post-Lyme disease syndrome" (PLDS) has been suggested to refer to patients with symptoms of the disease persisting for longer than 6 months. It has been suggested that antibiotic therapy is not effective in PLDS[24].

The House–Brackmann score is a widely recognised clinical grading scale for evaluating the function of facial nerve motor fibres, with grade I indicating normal motor function, and grade VI representing total absence of motor function in the cranial nerve VII fibres. Drack and Weissert report that in more than 90% of children with neuroborreliosis accompanied by facial nerve palsy the motor function of nerve VII is fully restored (House–Brackmann grade I) after causal treatment without steroid therapy[2]. Similar outcomes in children treated for neuroborreliosis with facial nerve palsy are reported by Skogman et al.[26].

Neurological and psychological deficits after neuroborreliosis are rare. In the majority of children, treated neuroborreliosis does not leave any cognitive, emotional or behavioural effects[27].

Summing up, it must be emphasised that children with peripheral facial nerve palsy should undergo diagnostic workup for *Borrelia burgdorferi* infection also in cases without typical erythema migrans in the early stage of the disease.

Conflict of interest

The authors do not declare any financial or personal links with other persons or organisations that might adversely affect the content of the publication or claim any right to the publication.

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