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Title:

Cerebral sinuvenous thrombosis: a rare complication of Lyme neuroborreliosis

Thrombose der zerebralen Blutleiter: seltene Komplikation einer Neuroborreliose

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Summary

Association between neuroborreliosis and cerebral sinuvenous thrombosis is rare and it can be made only when other, more common predisposing conditions are excluded. In the case of increased intracranial pressure and confirmed neuroborreliosis, early magnetic resonance venography and combination of antibacterial therapy with anticoagulation provide better long-term outcome. We present a case of a patient with cerebral sinuvenous thrombosis who was first treated for neuroborreliosis.

Zusammenfassung

Eine Verbindung von Neuroborreliose und Thrombose der zerebralen Blutleiter ist selten und kann erst dann gemacht werden, wenn andere prädisponierende Ursachen ausgeschlossen worden sind. Bei erhöhtem inkraniellem Druck und bestätigter Neuroborreliose führen eine frühe MRI-Venographie und die Kombination von Antibiotika und Antikoagulantien zu besseren langfristigen Ergebnissen. Wir berichten über einen Patienten mit Thrombose der zerebralen Blutleiter, der anfänglich wegen Neuroborreliose behandelt worden war.

Introduction

Lyme neuroborreliosis is an infectious disease of peripheral and central nervous system (CNS) caused by the spirochete Borrelia burgdorferi. Multifarious clinical manifestations of neuroborreliosis include Bannwarth's syndrome (painful meningoradiculitis with cranial nerves palsy), myelitis, progressive encephalitis, radiculopathy, polyneuropathy, and cerebral vasculitis [1]. Cerebral sinuvenous thrombosis (CSVT) is a well-established and potentially fatal complication of bacterial meningitis. However, CSVT associated with *B. burgdoferi* infection has rarely been reported [2].

Case report

A 73-year-old woman was admitted to a local general hospital because of acute onset of double vision and thunderclap headache, grading 7/10 on visual analog scale. Her medical history included well- controlled arterial hypertension and hypothyreosis. She denied prior head trauma, diabetes mellitus, and infectious diseases. Vital signs were normal at admission, and except for a subjective sense of double vision when looking toward the left, the rest of clinical examination did not detect any abnormalities. Brain computed tomography (CT) was unremarkable, but bilateral papilledema was present on fundoscopic examination. Laboratory findings such as complete blood cell count, C-reactive protein, liver function enzymes, electrolyte panel and coagulation profile

were all within the normal range. A supra-aortic trunk Doppler sonography together with transcranial sonography showed no abnormalities. The patient developed left abducens nerve palsy 3 days after the admission. Carefully taken medical history revealed that the patient was bitten by a tick 2 weeks before hospitalization. However, no skin changes were noticed afterward. Magnetic resonance imaging (MRI) of the brain was performed, which showed no pathological substrate. In the meantime, specific IgM antibodies against *B. burgdorferi* were detected in peripheral blood by enzyme-linked immunosorbent assay. Treatment with intravenous ceftriaxone was initiated followed by doxycycline, and 14 days later the patient was discharged from the hospital without neurological sequel with the diagnosis of benign intracranial hypertension.

The patient was readmitted to the hospital 2 weeks later because of neurological deterioration characterized by prominent headache, vomiting, left abducens nerve palsy, and gait instability. Cerebrospinal fluid (CSF) analysis showed lymphomonocytic pleocytosis (44cells/mm^3), protein elevation (0.67g/L), and normal glucose and lactate levels. Serological testing for neurothropic viruses, Klebsiella pneumoniae, Toxoplasma gondii and Treponema pallidum came negative. The Western blot finding of *B. burgorferi* IgM bands showed mild reactivity. MR venography was performed, which showed left internal jugular vein thrombosis together with left transversal and sigmoid sinus thrombosis (Figs. 1 and 2). Repeated laboratory investigation showed normal coagulation profile, D-dimer value, antinuclear antibody, extractable nuclear antigen antibodies (anti Ro/SS-A and anti La/SS-B), rheumatoid factor, anticardiolipin antibodies, anti-double-stranded DNA antibody, anticardiolipin antibody, anti-β2-glycoprotein, angiotensin-converting enzyme, serum tumor markers: carcinoembryonic antigen, carbohydrate antigen 19-9, neuron-specific enolase, cytokeratin-19, alpha-fetoprotein, carbohydrate antigen 125. Tests for hereditary coagulopathies including antithrombin, Factor VII, Leiden factor, protein C, and protein S were negative, and chest CT was normal. Intravenous administration of unfractionated heparin was started immediately, followed by oral anticoagulant warfarin. Control CT venography after 13 days demonstrated complete recanalization of the internal jugular vein and sigmoid sinus with hypoplastic left transversal sinus. The patient was re-examined in outpatient clinic 1 month after discharge, and complete resolution of symptoms was observed.

Fig. 1: Contrast-enhanced T1 image shows low signal intensity within the left transverse sinus, sigmoid sinus and jugular vein.

Fig. 2: The post contrast magnetic resonance venography reveals non visualisation of left transverse and sigmoid sinus.

Discussion

CSVT is a rare type of cerebrovascular disease that accounts for 0.5% of all strokes [4]. A variety of disorders can cause or predispose to CSVT such as genetic or acquired prothrombotic conditions, cancer, hematological diseases, inflammatory systemic disorders, head trauma, drugs (oral contraceptives, hormonal replacement therapy), and both systemic and CNS infections. Septic CSVT is usually associated with Staphylococcus, Streptococcus and Klebsiella species. Association with neuroborreliosis is extremely rare, and to our knowledge, there is only one similar report so far [3]. Causal relationship between neuroborreliosis and CSVT in our case was based on CSF findings of lymphomonocytic pleocytosis, western blot detection of IgM antibodies, and exclusion of other more common causes of CSVT. Mild reactivity of IgM antibodies detected by western blot in our patient can be explained by prior antibiotic treatment.

Common pathophysiological mechanism of CSVT following underlying infection is a direct extension of the infection from adjacent structures, usually arising from focal thrombophlebitis with propagation to larger dural veins and sinuses, most commonly affecting lateral and superior sagittal sinuses [4]. In addition, infection can cause hypercoagulable state, endothelial dysfunction, and activation of inflammatory and procoagulant cascades [5]. The aforementioned findings support the approach of combined therapy with antibiotics and anticoagulant heparin as the treatment mainstay.

Our report expands the clinical spectrum of neuroborreliosis, and highlights the importance of recognizing and investigating signs of raised intracranial pressure, as early detection of CSVT may enable adequate treatment and prevent fatal complications.

Conflict of interest

The authors declare that there is no conflict of interest.

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Figure 1.

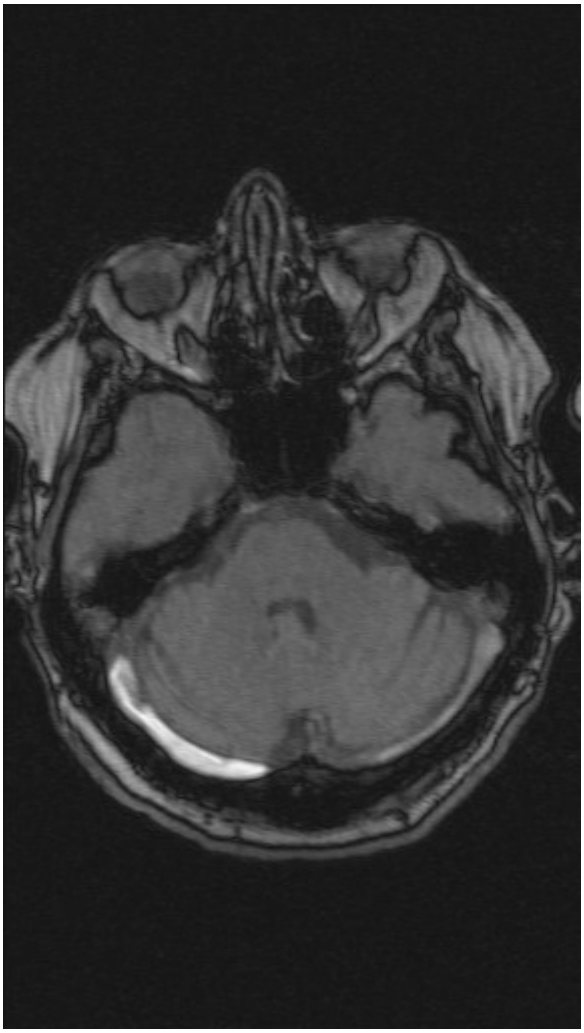

Figure 2.