SHORT REPORT

Opsoclonus–myoclonus as a manifestation of Lyme disease

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Opsoclonus–myoclonus syndrome (OMS) is a rare condition that includes chaotic multidirectional saccadic eye movements associated with myoclonus and ataxia. In adults, it is usually considered to be an autoimmune disease occurring either in a paraneoplastic context or after central nervous system infection. We report the case of a patient who presented with the classic features of OMS as a manifestation of acute *Borrelia burgdorferi* infection that was shown both on serum and cerebrospinal fluid examination. The outcome was favourable after prolonged antibiotic treatment. Lyme disease could be added to the list of aetiologies to be screened in OMS, as it would allow effective treatment and avoidance of unnecessary investigations.

Opsoclonus is a disorder of saccadic eye movements that interrupts steady fixation and persists during sleep. It consists of involuntary, chaotic, multidirectional saccades without intersaccadic intervals,[1] and is often accompanied by arrythmic-action myoclonus that predominantly involves the trunk and the neck. Some patients also exhibit signs of cerebellar dysfunction such as dysarthria and ataxia. In adults, the syndrome is usually recognised as an autoimmune disease occurring either in a paraneoplastic context[2] or after central nervous system (CNS) infection.[3] We report a case of opsoclonus–myoclonus syndrome (OMS) related to recent *Borrelia burgdorferi* infection.

A 40-year-old farmer was admitted to the neurology unit of Hôpital Neurologique Pierre Wertheimer, Lyon, France for vertigo, gait disturbance and nausea. He reported no prior medical history. Ten days before admission, the patient had presented fever and headache lasting for 3 days. Subsequently, he developed walking difficulties with unsteadiness of gait. On admission, clinical examination disclosed signs of static and kinetic cerebellar dysfunction with ataxia and hypermetric movements of both upper limbs. Ocular examination showed no ocular motor palsy but intermittent bursts of multidirectional binocular saccades, often triggered by eye movements. The patient also presented axial spontaneous myoclonic jerks of the trunk and the neck muscles. No meningeal syndrome was observed and general examination was normal. Shortly after admission, the patient's clinical condition deteriorated; he became unable to walk and even to read because of his abnormal eye movements.

Cerebral magnetic resonance image (T1-weighted sequences with gadolinium injection, T2-weighted sequences, fluid-attenuated inversion recovery sequences) was normal. Lumbar puncture showed limpid cerebrospinal fluid (CSF) with normal opening pressure, lymphocytic meningitis with mild pleiocytosis (28 white cells/mm^3 with predominance of lymphocytes), normal glucose and protein levels. CSF electrophoresis showed oligoclonal banding. Usual serum tests were normal, except for a mild lymphopenia that resolved spontaneously. Emergency treatment with aciclovir was given until herpes CSF polymerase chain reaction proved negative. Screening of all the most usual CNS infectious diseases was carried out but remained negative for herpes species viruses, HIV, arbovirus, *Treponema pallidum* serum and CSF antibodies. Polymerase chain reactions for enterovirus and Whipple disease were negative in CSF. Metabolic, toxic, dysimmune and degenerative aetiologies were also examined. CSF 14.3.3 protein dosage was negative. The search for a paraneoplastic cause was negative: thoraco-abdominopelvic computed tomography scans, tumour markers and antineuronal antibodies in serum and CSF, as well as the electroencephalogram, were normal.

As the patient lived in an area endemic for Lyme disease, in northeastern France, and despite the absence of any anamnestic or suggestive clinical features—he did not report a tick bite or a rash—neuroborreliosis was considered. Serological examination was consistent with a recent *B burgdorferi* infection: immunoglobulin (Ig) M index was raised in CSF, as assessed by western blot. Evidence of intrathecal synthesis of Lyme antibodies matched the serum and CSF titres. Control serological examinations (carried out 1 and 4 months after the first serology) confirmed the acute infection, showing the disappearance of IgM and persistence of a low IgG titre in the blood. The patient improved dramatically after a few days of treatment with ceftriaxone 2 g per day intravenous, which was continued for 3 weeks. He was transferred to a rehabilitation unit; full recovery was observed at 1 month and the patient was able to resume working.

In adults the most common causes of opsoclonus include parainfectious brain stem encephalitis, paraneoplastic[4] and metabolic–toxic disturbances.[5] A case of opsoclonus after anti-rubella vaccination has been reported.[6] Infectious aetiologies include various viral agents (enterovirus, Epstein–Barr virus, poliovirus, Coxsackie virus, mumps, West-Nile virus, HIV) or bacterial diseases (*Salmonella* species and *Mycobacterium tuberculosis*).

To our knowledge, *B burgdorferi* infection has never been reported as a potential aetiology of OMS in adults. Only one such case has been reported in children, with a favourable outcome with ceftriaxone.[7] Lyme disease is known to be responsible for various neuro-ophthalmological manifestations[8 9] such as keratitis, uveitis, chorioretinis, papillitis, orbital myositis, optical neuropathy, but also cranial neuropathies (and internuclear ophthalmoplegia) or cranial nerve VI palsy in relation with intracranial hypertension. Our case suggests that Lyme disease may also be presented by OMS. The commonly accepted pathophysiological mechanism of opsoclonus is that of a dysfunction of the omnipause-inhibiting cells located in the brain stem leading to a disinhibition of the saccadic burst neurones.[1] Whether dysfunction of omnipause cells is the primary cause of OMS or whether OMS results from hyperactivation of the fastigial nucleus is still a matter of debate. In the second case, hyperactivation of fastigial nucleus would be a consequence

Abbreviations: CNS, central nervous system; CSF, cerebrospinal fluid; OMS, opsoclonus–myoclonus syndrome

of dysfunction of oculomotor dorsal vermis.[10] OMS caused by neuroborreliosis might result from a direct CNS microbiological invasion, as suggested by an autopsy report of *B burgdorferi*-positive chronic encephalomyelopathy[11] or from a dysimmune postinfectious mechanism.[3 5]

In conclusion, this report suggests that neuroborreliosis should be systematically looked for in patients with OMS, as a curative treatment is available.

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