Palpebral narrowing associated with peripheral facial nerve palsy showed a varying degree of drooping of the eyebrow. There were patients with a severe degree of narrowing. This indicates that the frontal muscle which has been considered as an accessory muscle, occasionally plays a very important part in eyelid elevation. We suspect that this is closely related to the skin condition of the upper face. Some Japanese people, especially elderly ones, have excessively loose skin of the upper eyelid or forehead, resulting in "masked" palpebral narrowing. In such a situation, occurrence of frontal muscle weakness can produce severe palpebral narrowing. In conclusion, the "eyebrow lifting test" is simple but very helpful in differentiating between two types of palpebral narrowing due to weakness of the frontal muscle and levator palpebrae superioris.

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1 Adams RD, Victor M. Diseases of the cranial nerves. In: Adams RD, Victor M, eds. *Principle of neurology*, 4th ed. New York: McGraw-Hill, 1989:1081.
2 Ohkawa S, Yoshida T, Yamadori A. Acute stage Bell's palsy and narrowing of the palpebral fissure caused by drooping of the eyebrow and the upper eyelid. *J Neurol Neurosurg Psychiatry* 1992;**55**:976.
3 Caplan LR. Ptosis. *J Neurol Neurosurg Psychiatry* 1974;**37**:1–7.
4 Miller NR. Anatomy and physiology of normal and abnormal eyelid position and movement. In: Miller NR, ed. *Walsh and Hoyt' s clinical neuro-ophthalmology*. 4th ed. Baltimore: Williams and Wilkins, 1985:933.
5 Averbuch-Heller L, Poonyathalang A, von Maydell RD, Remler BF. Hering's law for eyelids: Still valid. *Neurology* 1995;**45**:1781–3.

Generalised motor neuron disease as an unusual manifestation of *Borrelia burgdorferi* infection

Lyme borreliosis is a well known multisystem disease caused by the spirochete *Borrelia burgdorferi* and can produce a wide array of neurological abnormalities in humans. The most frequent are meningitis, cranial neuritis, and painful radiculoneuritis.[1] Other clinical manifestations include chronic encephalomyelitis, spastic paraplegia, and axonal polyneuropathy. Our report concerns what we think to be the first case of a patient with upper and lower motor neuron disease and *Borrelia burgdorferi* infection of the CNS. A causal relation is strongly supported by an evaluation of the *Borrelia burgdorferi* specific antibody index and the patient's favourable response to medical treatment.

Fifteen months before admission a 33 year old patient noticed weakness in his right hand followed by weakness of the left hand and a progressive gait disturbance. Although he had no pain or sensory disturbance and no history of a tick bite, an erythema migrans, or arthralgias, his physician tested him for *Borrelia burgdorferi* specific antibodies in the serum because he lived in an endemic region. The test disclosed high concentrations of specific IgG antibodies (1:1200, cut off <1:200). The patient was treated with doxycyclin for two weeks. A control examination performed in a different laboratory still disclosed high concentrations of specific IgG antibodies (1:160, cut off 1:40). Treatment was started again with cefotaxim (2g intravenously for five days). Six months later he was admitted to our hospital because of persisting paresis and muscle atrophy.

Severe atrophy of hand muscles.

On admission, clinical examination disclosed hyperactive deep tendon reflexes with a clonus in both ankles. The muscles of both hands and forearms showed atrophy and severe paresis (figure). His gait was clumsy and stiff, and there was mild spastic paraparesis. There was no sensory loss, and cerebellar function was normal.

Needle EMG disclosed severe active denervation in the small hand muscles bilaterally. Mild to moderate signs of axonal damage were seen in the right anterior tibial muscle and in the left masseter muscle. Motor and sensory nerve conduction velocities were normal. No conduction block could be detected. F waves were abolished. Visual and sensory evoked potentials were normal. Motor evoked potentials disclosed prolonged central conduction times to both anterior tibial muscles and to the left abductor digiti minimi muscle.

Routine blood chemistry was normal. Serum *Treponema pallidum* and GM1-specific antibodies were not detected. Borrelia serology tests showed slightly raised concentrations of *Borrelia burgdorferi* specific IgG antibodies in serum and clearly raised concentrations in the CSF. At the time of the first lumbar puncture (after antibiotic treatment), CSF contained five white blood cells/μl and a total protein concentration of 410 mg/l. Reiber-formula analysis[2] indicated an intrathecal synthesis of IgG and IgA. Employing a sensitive affinity blotting technique most of the oligoclonal IgG bands in the CSF were shown to be specific for *Borrelia burgdorferi* This finding was confirmed by western blotting using identical concentrations of IgG in the CSF and serum. A higher number of *Borrelia burgdorferi* specific antibody bands were found in the CSF than in serum. Calculation of the *Borrelia burgdorferi* specific antibody index from enzyme linked immunosorbent assay (ELISA) studies disclosed raised values for IgG and IgA. Specificity of intrathecally produced IgG antibodies for *Borrelia burgdorferi* was confirmed by employing a highly specific 14 kDa fragment of the flagellin as antigen in enzyme linked immunosorbent assay (ELISA).[3] The table shows the detailed data of the antibody tests on admission and on follow up examination five months later. MRI of the cervical spinal cord and the brain disclosed no abnormalities.

The patient was treated with ceftriaxone intravenously for two weeks, followed by oral

Summary of antibody tests

Examination	*Normal*	*Initial study*	*Follow up**
Serum:			
B burgdorferi IgG antibodies	<1:16	1:64	<1:16
B burgdorferi IgM antibodies	<1:48	1:12	<1:12
CSF:			
Total cell count (/μl)	<5	5	3
CSF/serum albumin ratio (10^{-3})	<7.5	4.7	3.5
IgG-local (%)	0	75	65
IgA-local (%)	0	20	10
B burgdorferi IgG antibodies	<1:16	1:16	1:2
B burgdorferi IgM antibodies	<1:48	<1:2	<1:2
B burgdorferi IgG antibody index	<1.5	24.6	18.0
B burgdorferi IgA antibody index	<1.5	2.2	7.2
14 kDa fragment IgG antibody index	<1.5	35.6	Negative

*Five months after treatment.

prednisone for 10 weeks. After this treatment the patient's condition improved slowly but continuously. At the time of the last clinical control examination 18 months after hospital discharge the patient was able to work without physical impairment.

Clinical and electrophysiological findings met all the criteria for the diagnosis of motor neuron disease. Clinical signs of lower motor neuron involvement were present in both arms. Electromyographic studies disclosed axonal loss at three different levels—namely, lumbar (anterior tibial muscle), cervical (hand muscles), and supraspinal (masseter muscle). Clear signs of damage to the upper motor neuron were also present. Although the symptoms of the patient could be explained by cervical myelitis the EMG findings with evidence of axonal damage in the anterior tibial and masseter muscle as well as the lack of any sensory abnormalities argue strongly against this possibility.

In addition, signs of inflammation in the CSF were not consistent with a diagnosis of amyotrophic lateral sclerosis. We identified a *Borrelia burgdorferi* infection of the CNS as the cause of the inflammation. Evidence included a raised specific IgG and IgA antibody index, the demonstration of *Borrelia burgdorferi* specific oligoclonal IgG bands in the CSF and the predominance of individual *Borrelia burgdorferi* specific antibody bands in CSF (as indicated by western blotting). The absence of a high white cell count and protein in the CSF could be attributed to prior antibiotic treatment. Optimising dose and duration, antibiotic treatment was renewed and combined with a long term steroid therapy. Four months later a CSF examination showed a considerable decrease in specific antibody concentrations, and the patient's condition continued to improve.

In the light of the evidence, it seems safe to conclude that the patient's symptoms were due to a CNS *Borrelia burgdorferi* infection which merely mimicked amyotrophic lateral sclerosis. Several reports have been published on spirochetal diseases leading to isolated damage to the motor system. Spinal meningovascular lues has been reported to cause a clinical syndrome mimicking motor neuron disease.[4] Fredrikson and Link published a case report of a patient with isolated upper motor neuron symptoms due to CNS borreliosis who responded favourably to antibiotic treatment.[5] Cases of painful motor neuropathy due to *Borrelia burgdorferi* specific infection have also been reported.[1] Halperin *et al*[6] found serological evidence of exposure to *Borrelia burgdorferi* in nine of 19 patients with motor neuron disease. However, none of them showed signs of *Borrelia burgdorferi* specific immunoreactivity in the CSF or favourable response to treatment.

It can be speculated that the spirochete *Borrelia burgdorferi* has the ability to induce an immune reaction that specifically affects motor neurons. This reaction may mimic different, non-curable diseases, such as spastic spinal paralysis, spinal muscle atrophy, and amyotrophic lateral sclerosis. Therefore, we suggest that patients diagnosed as having progressive motor neuron disease, who live in endemic areas, should be tested for *Borrelia burgdorferi* specific antibodies in serum and in CSF. The test could reliably detect a rare, but treatable disease mimicking motor neuron disease.

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1 Pachner AR, Steere AC. The triad of neurologic manifestations of Lyme disease: meningitis, cranial neuritis, and radiculoneuritis. *Neurology* 1985;**35**:47–53.
2 Reiber H, Felgenhauer K. Protein transfer at the blood cerebrospinal fluid barrier and the quantitation of the humoral immune response within the central nervous system. *Clin Chim Acta* 1987;**163**:319–28.
3 Kaiser R, Rasiah C, Gassmann G, Vogt A, Lücking C. Intrathecal antibody synthesis in Lyme neuroborreliosis: use of recombinant p41 and a 14-kDa flagellin fragment in ELISA. *J Med Microbiol* 1993;**39**:290–7.
4 ElAlaouli F, Medejel A, AlZemmouri K, Yahyaoui M, Chkili T. Syphilitic lateral amyotrophic sclerosis. A study of 5 cases. *Rev Neurol* 1990;**146**:41–4.
5 Fredrikson S, Link H. CNS-borreliosis selectively affecting central motor neurons. *Acta Neurol Scand* 1988;**78**:181–4.
6 Halperin JJ, Kaplan GP, Brazinsky S, *et al*. Immunologic reactivity against Borrelia burgdorferi in patients with motor neuron disease. *Arch Neurol* 1990;**47**:586–94.

Severe but transient parkinsonism after tetanus vaccination

A 38 year old metal worker with a history of hypertension and hyperthyroidism presented with fluctuating fever and sweating, palpitations, tremor of the upper parts of both legs, and diplopia. These symptoms had been present for five days and had started within hours after he had received the last of three vaccinations for tetanus (TE anatoxal berna, contents: 20 LF tetanus toxoid, 2 mg aluminium phosphate, and 0.1 mg thiomersal, Primmed BV, Almere, The Netherlands). These vaccinations were given because of an injury to his right index finger one month before. There was no family history of movement disorders. Physical examination showed profuse sweating, normal consciousness, a temperature of 37.3°C, symmetric rigidity of all four limbs, and a painful tremor in the upper parts of his legs. Muscle strength, tendon reflexes, and sensation were normal.

Within a week he progressed to severe hypokinetic dysarthria, a mask-like face, and a resting tremor of both hands, and he had bradykinesia and generalised rigidity, together with a cogwheel phenomenon in the arms.

Laboratory examination showed a creatine phosphokinase activity of 2682 U/l (normal <190 U/l) and normal blood concentrations of manganese, copper, ceruloplasmin, and carbon monoxide. His CSF showed 50 lymphocytes (normal range 0-3), slightly raised total protein (0.54 g/l; normal range 0.15-0.45 g/l), normal IgG index (0.43; normal <0.66), and negative serological tests on Epstein-Barr virus, herpes zoster virus, herpes simplex virus, syphilis, *Borrelia burgdorferi*, and *Mycoplasma pneumoniae*. Brain MRI was normal. Single photon emission computed tomography (SPECT) with ^{123}I-iodobenzamide (IBZM), specifically binding to the cerebral dopamine receptor (D2), showed a decreased ganglia:cortex ratio, indicating a postsynaptic disorder. Nevertheless, biperidine, levodopa and carbidopa, and pergolide were prescribed, resulting in gradual but impressive clinical improvement within several weeks.

The clinical syndrome was unclear during the first few days after admission, but gradually developed into a hypokinetic rigid syndrome with resting tremor, generalised bradykinesia, and rigidity. This responded well to treatment with levodopa/carbidopa and a dopamine agonist.

Possible causes of a rapidly progressive form of parkinsonism are encephalitis, intoxication, head trauma, tumour, ischaemia, or hydrocephalus.[1] Imaging studies showed no abnormalities, thereby excluding the last three possible causes. Repeated history taking failed to disclose head trauma. The profession of the patient might suggest poisoning, but blood concentrations of manganese and copper were normal. The CSF showed mild pleiocytosis, but serological testing for various specific microorganisms did not show any recent infection. Radionuclide imaging showed a pattern similar to that seen in progressive supranuclear palsy or multiple system atrophy.[2]

The sequence of events strongly suggests a relation between the vaccination and the neurological syndrome, although the causal nature is difficult to prove.[3] To our knowledge, there are no reports of parkinsonism after tetanus immunisation. Alves *et al* reported a 5 year old boy who developed a postencephalitic rigid akinetic syndrome 15 days after vaccination for measles with live attenuated virus. Again, cause and effect remained open for debate.[4 5]

The tetanus vaccine used in our patient does not contain any living microorganisms. However, repeated injections with the tetanus toxoid might have caused hypersensitivity, and also an immunological cross reaction of antibodies with neuronal tissue directly after the last injection. This might also explain the pleiocytosis and raised protein and IgG content in CSF. The alternative explanation is that one of the substances in the vaccine vehicle, thiomersal or aluminiumphosphate, had a neurotoxic effect.

Although we are aware that a causal relation between the vaccine and the hypokinetic rigid syndrome is far from established, we have no better explanation. We wish to record the patient history as a reference, in case analogous patients might be seen in the future.

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1 Fahn S. Parkinson's disease and other basal ganglion disorders. In: Asbury AK, McKhann GM, McDonald WI, eds. *Diseases of the nervous system*. London: Heinemann, 1986;**98**:1217-28.
2 Buck A, Westera G, Sutter M, Albani C, Kung HF, von Schulthess GK. Iodine-123-IBF SPECT evaluation of extrapyramidal diseases. *J Nucl Med* 1995;**36**:1196-200.
3 Stratton KR, Howe CJ, Johnston RB, eds. *Adverse events associated with childhood vaccines. Evidence bearing on causality.* Washington, DC: *National Academy Press,* 1994.
4 Alves RSC, Barbosa ER, Scaff M. Postvaccinal parkinsonism. *Mov Disord* 1992;**7**:178-80.
5 Fenichel GM. Postvaccinal parkinsonism [letter]. *Mov Disord* 1993;**8**:253