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

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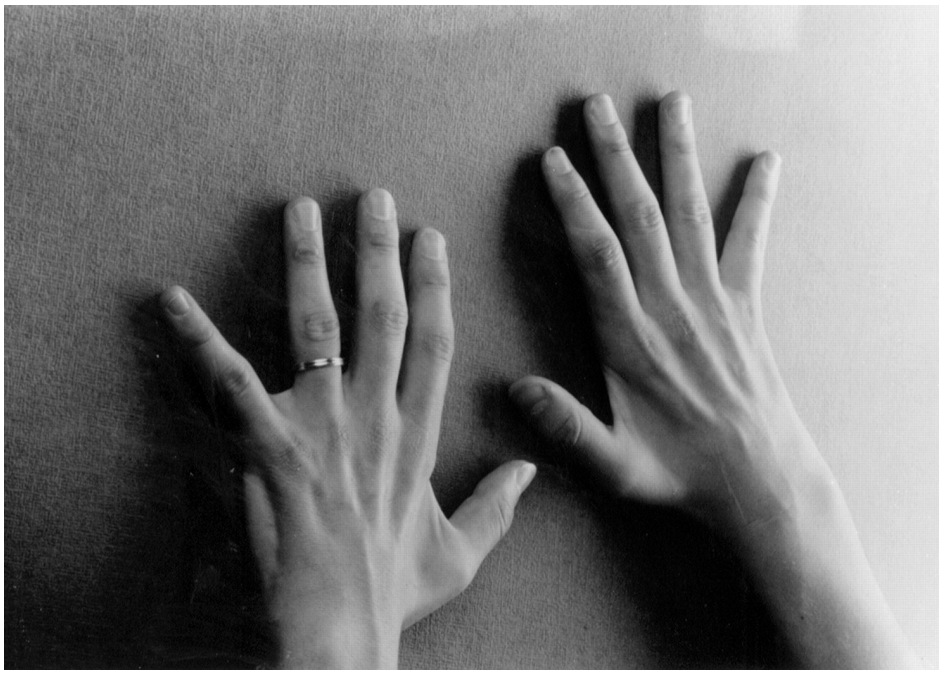
ABSTRACT

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 toBorrelia burgdorferi specific infection have also been reported. 1 Halperin et al 6 found serological evidence of exposure to Borrelia burgdorferiin nine of 19 patients with motor neuron disease. [...]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.

FULL TEXT

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.



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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.

Examination	Normal	Initial study	Follow up 1-150
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 (/[mu]l)	<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

Five months after treatment.

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

Subject:	Amyotrophic lateral sclerosis; Lyme disease; Rodents; Hospitals; Antibiotics; Immunoglobulins; Infections
Publication title:	Journal of Neurology, Neurosurgery and Psychiatry; London
Volume:	63
Issue:	2
First page:	257
Publication year:	1997
Publication date:	Aug 1997
Publisher:	BMJ Publishing Group LTD
Place of publication:	London
Country of publication:	United Kingdom, London
Publication subject:	Medical Sciences--Psychiatry And Neurology, Medical Sciences--Surgery
ISSN:	00223050
CODEN:	JNNPAU
Source type:	Scholarly Journals
Language of publication:	English