A case of Lyme neuroborreliosis with bilateral recurrent laryngeal nerve palsy

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DECLARATIONS

Competing interests
None declared

Funding
None

Ethical approval
Written informed consent to publication has been obtained from the patient or next of kin

Guarantor
MF

Contributorship
All authors were responsible for patient care; MF was responsible for writing the article; MP was responsible for the online video; BF, ES and JW were responsible for the critical reading of the manuscript

Acknowledgements
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We report a case of neuroborreliosis with severe bilateral recurrent nerve palsy, which had to be treated at an intensive care unit because of acute respiratory distress.

Case history

A 66-year-old woman was transferred to our facility from a local hospital. Her only remarkable prior condition was diabetes mellitus, for which she was on oral medication. She did not recall a tick bite or any skin changes during the last months.

During the past 4 weeks, she had suffered severe lumbosacral and cervicobrachial pain with multisegmental radiation (visual analog scale 8–10). Pain had responded insufficiently to non-steroidal analgesics and opioids. Computed tomography (CT) scans of the cervical and lumbar spine had been unremarkable except for moderate spondylarthrosis. She had been hospitalized for the past 4 days due to a severe bout of pain radiating into both legs, with nocturnal accentuation. During the 24 hours prior to admission to our facility, leg weakness rapidly emerged and resulted in an inability to stand, while pain had improved.

On admission (day 1), the patient presented with distally accentuated paresis and slight muscular atrophy in arms and legs. Cranial nerves were unremarkable, as were phonation and swallowing ability. Deep tendon reflexes were absent in the lower and reduced in the upper extremities. She was unable to walk unaided, sensation in both hands and feet being diminished.

Radicular or cord compression was ruled out by magnetic resonance (MR) tomography of the entire spine.

Cerebrospinal fluid (CSF) analysis showed 57 leukocytes/μL (normal 0–4; mainly activated lymphocytes, several plasma cells), elevated protein content (103 mg/dL; normal 15–45) and IgG serum/liquor index 1.80 (normal <0.50) but normal glucose concentration. CSF serology was positive for *Borrelia burgdorferi* IgG and IgM (enzyme-linked immunosorbent assay [ELISA] and immunoblot) but negative for other neurotropic agents. Numerous oligoclonal bands were found. In the serum, *B. burgdorferi* IgG and IgM antibodies were detectable as well. Parameters for autoimmune diseases were negative.

A diagnosis of Lyme polyradiculitis was made and ceftriaxone 2 g IV once daily was started. Despite treatment, the patient's condition further worsened. On day 2, she developed asymmetrical tetraparesis (right arm power grade [PG] 3–4; left arm PG 4; right leg PG 3, left leg PG 2) and incomplete loss of sensation in all extremities. Electromyography confirmed a generalized axonal neuropathy with reduced compound muscle action potential amplitudes and loss of F waves. The most striking new symptom was dysphonia with a soft and hoarse voice. Over the following 24 hours, the speech abnormality progressed to aphonia. Simultaneously, dyspnoea developed and ambient-air peripheral oxygen saturation rapidly decreased from 98% to 80%. Laryngoscopy on day 2 demonstrated bilateral vocal cord paralysis, with both vocal cords fixed in a paramedian position (online video). She was monitored at the neurological intensive care unit (ICU) from days 3 to 6, and received nasal oxygen. Intubation as well as tracheostomy were discussed, but did not become necessary.

Differential diagnosis regarding the clinical worsening at this point included polyneuritis

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cranialis (Miller-Fisher syndrome), which could be ruled out by negative antiganglioside antibodies, and brainstem encephalitis, which was discarded after a normal contrast-enhanced brain MR tomography.

Another CSF sample was taken on day 19. Abnormalities had partly resolved (40 leukocytes/μL, protein 72 mg/dL, and IgG index 0.90); serology showed positive *B. burgdorferi* IgG and borderline-positive IgM (ELISA and immunoblot). CSF/serum Borrelia IgG index from this CSF sample was markedly elevated at 7.5 (normal, <1.3).

Ceftriaxone was administered for three weeks. During this period, the patient gradually recovered, and recurrent nerve function normalized. Residual weakness (hands, PG 4; legs, PG 4-5) and slightly reduced sensation in both hands and feet persisted. The patient was admitted to a rehabilitation facility. At follow-up after 6 months, no residual weakness, in particular, no laryngeal abnormalities were found anymore. Neuropathic pain mainly affecting the right leg was present and required treatment with gabapentin.

Discussion

Lyme borreliosis is a tick-borne infectious disease caused by *Borrelia burgdorferi*. It involves many organs (skin, heart, joints) and causes a variety of neurological manifestations. Meningopolyradiculoneuritis (Bannwarth syndrome) with severe radicular pain and/or motor symptoms represents the typical pattern in the acute stage. These signs and symptoms take 4–6 weeks to develop, and as in our patient, may be bilateral. However, with antibiotic treatment, prognosis usually is benign, and progression to tetraplegia is an infrequent event. The combination of radiculitis and cranial neuritis is common (about 60% of patients). Cranial neuritides mainly affect the facial nerve (in up to 60% of all neuroborreliosis patients, bilateral in 40%), and the abducens nerve in 10%.[1–3] Less frequently, all other cranial nerves (except the olfactory) may be affected. Recurrent nerve palsy is extremely rare, with only six reported occurrences (all unilateral), having caused a sore throat or hoarseness, but no dyspnea. Prognosis was good after antibiotic treatment in all cases.[4–7]

Our patient presented with severe pain and tetraparesis due to Lyme polyradiculitis, while recurrent nerve palsy occurred later. This case requires attention because it is the first with bilateral vocal cord paralysis due to affection of both recurrent nerves. Accordingly, critical narrowing of the glottis (online video) and dyspnoea ensued. Clinical worsening was rapid, and intubation seemed imminent. Despite immediate treatment with ceftriaxone, signs and symptoms initially worsened. There are relatively few reports on therapy efficacy or latency of response in Lyme disease.[8] In most cases, however, amelioration of symptoms, especially pain, is seen promptly after antibiotic therapy. As in our patient, response may sometimes be delayed for a few days.

It is interesting that in all six previously reported cases, recurrent nerve paralysis was an isolated symptom. A survey covering otolaryngologic aspects of Lyme disease found that 4.9% of patients reported hoarseness. There is no mention, however, if this was associated with vocal cord dysfunction.[9] The relatively long duration (four weeks) of sensory radicular symptoms before the rapid occurrence of tetraparesis and vocal cord paralysis within 72 hours deserves comment. In the literature, variable onset of paresis has been reported, which may indeed occur after several weeks of sensory symptoms.[2,3]

Summing up, this report intends to raise awareness that Lyme disease may cause bilateral recurrent nerve palsy with respiratory dysfunction. Prognosis may be favorable upon expeditious antibiotic treatment despite severe initial deficits.

Comment on the online video

The examiner (PM) is telling the patient to phonate (say 'hee'). There is bilateral vocal cord paralysis, with both vocal cords fixed in a paramedian position. The video can be seen at http://shortreports.rsmjournals.com/cgi/content/full/shorts.2010.010080/DC1.

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