Poliomyelitis-like syndrome with matching magnetic resonance features in a case of Lyme neuroborreliosis

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In summary, our data suggest that hypophosphataemia plays a key role in the pathogenesis of neuropathy that develops in patients following IVH without IP, and highlights the importance of rapid phosphate replacement therapy in these patients.

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Lyme disease is a multisystemic disorder caused by an epizootic organism of the spirochete group, called *Borrelia burgdorferi* (Bb), which is transmitted to humans by ticks of the genus *Ixodes*.[1] Three sequential clinical stages have been described: (i) early localised; (ii) early disseminated; and (iii) late persistent disease. Lyme neuroborreliosis may occur during the early dissemination phase, most often as a painful meningo-radiculitis and very rarely as a radiculo-myelitis, whereas encephalomyelitis is observed in the late phase.[2] We report the case of a patient with an early subacute poliomyelitis-like syndrome closely matching the selective involvement of the anterior horns and roots of the cervical spinal cord seen on magnetic resonance (MR) imaging.

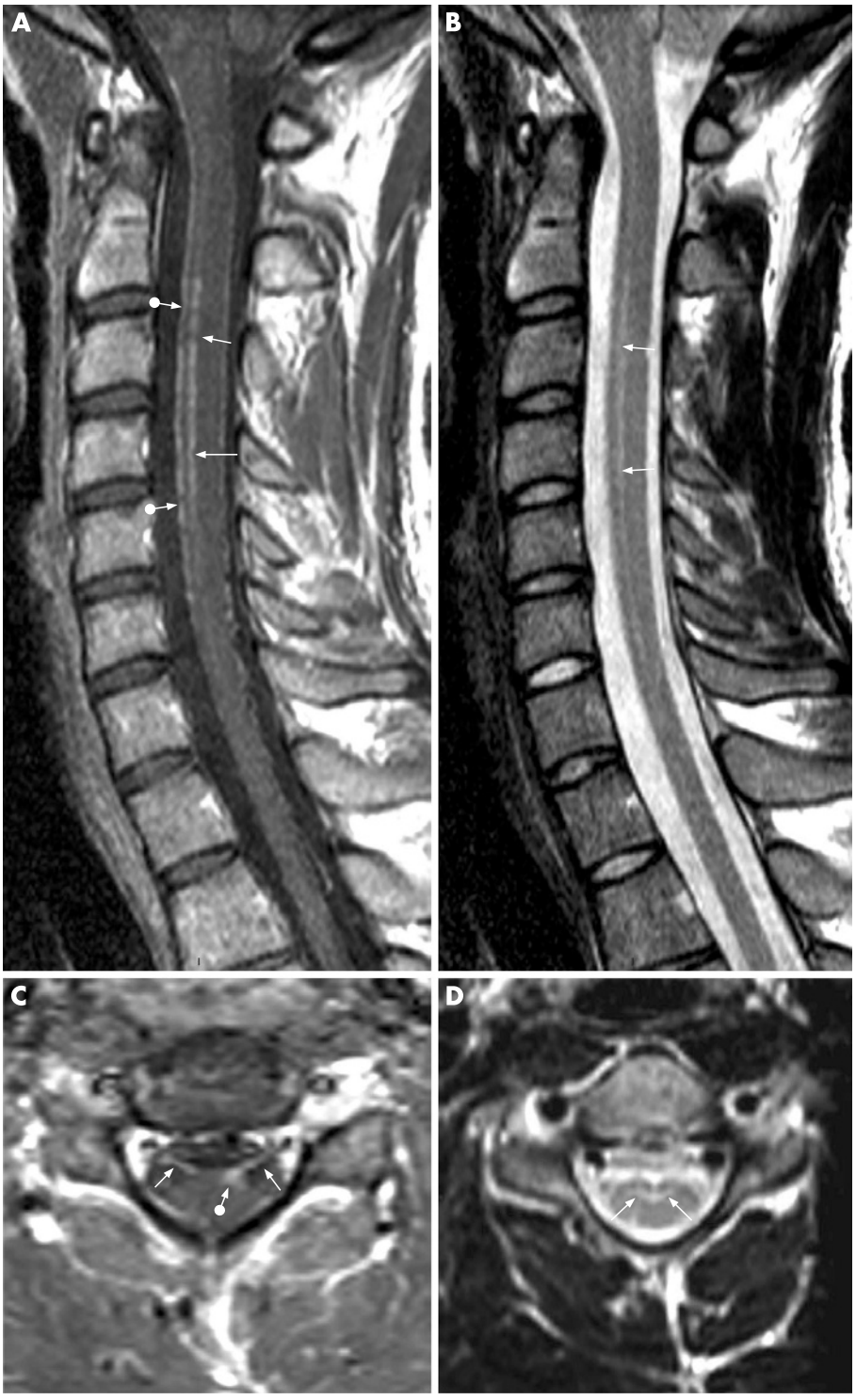

Figure 1 Magnetic resonance (MR) workup of the cervical spine at admission. (A) Post-contrast mid-sagittal T1 weighted image showing abnormal enhancement of both the anterior pia mater (ball-arrowheads) and anterior horns of central grey matter from C2 to C5 (arrows). (B) Mid-sagittal T2 weighted image (similar slice location as in (A)) showing abnormal linear hypersignal intensity in the anterior horns of central grey matter in the same C2–C5 area (arrows). (C) Post-contrast transverse T1 weighted image at the C5 level showing thickened, abnormally enhancing, anterior roots due to inflammatory blood root barrier disruption (arrows), together with enhancement within the left anterior horns (ball-arrowhead). (D) T2 weighted transverse image at a comparable level showing abnormal hypersignal intensity within the anterior horns (arrows).

A previously healthy 21-year-old man presented in September 2006 with painless weakness in both arms that had increased over a period of 2 weeks. He had a short history of neck

pain and stiff neck 2 months previously, which had spontaneously resolved. An indepth retrospective anamnesis failed to reveal any evidence of tick bites or erythema chronicum migrans, but the patient had undertaken open air activities in a woody countryside at the end of June.

Physical examination showed severe weakness (2/5) in the proximal muscles of both upper limbs. Strength was only mildly reduced in the hands (4.5/5) and was normal in the legs. No sensory deficit was present. Diffuse fasciculations were observed in both shoulders. Deep tendon reflexes were not elicited in the right arm. Of note, Lhermitte's sign was absent. There were no meningeal signs or fever.

Cervical spine MR examination showed abnormally increased signal intensity within the anterior horns of the cord ''grey butterfly'' from C2 to C5 on the T2 weighted images (fig 1B, D), together with increased thickness and strong contrast enhancement of the anterior radicelles of the same dermatoma on post-contrast T1 weighted images (fig 1A, C). Additional abnormal post-contrast enhancement was also observed within the pia mater (fig 1A) and the anterior horns (fig 1C). Brain MR examination was unremarkable.

Neurophysiological studies demonstrated elective involvement of motor nerves at the radicular or motoneuronal level. Needle electromyography showed signs of acute denervation with spontaneous fibrillations in both deltoid and biceps brachii, with a right-sided predominance. Abnormalities were also present in the first dorsal interosseous muscles and in the extensor digitorum communis, but to a lesser degree. Sensory and motor conduction velocities of the ulnar and median nerves were normal, as well as the latency and the amplitude of the F waves.

Standard haematological tests and biochemical assays were normal. Serology was negative for HIV, hepatitis B and C, and syphilis (TPHA and VDRL tests). However, serological tests for Lyme disease (Euroimmun Anti-Borrelia plusV1sE ELISA IgG and Euroimmun Anti-Borrelia ELISA IgM, Lübeck, Germany) were strongly positive in serum for both IgM (IgM ratio of 5.74, normal value <1.0) and IgG (133 relative unit (RU)/ml, normal value <20) antibodies. The IgG and IgM positive samples were confirmed by a commercial western blot assay (Euroline-WB; Euroimmun AG) showing specific patterns (OspC for IgM antibodies, V1sE, p83, p39, p31, 30, OspC and p17 for IgG antibodies).

CSF analysis showed 79 cells/mm^3 with 79% lymphocytes, 12% reactive lymphoblasts and 9% monocytes. The total protein content was elevated (98 mg/dl; normal range 20–55). Considerable intrathecal synthesis of IgM was present (intrathecal fraction at 76%) with a lower synthesis of IgG (intrathecal fraction at 17%). However, several CSF restricted oligoclonal IgG were present. Bb PCR was negative. However, native CSF contained 58 RU/ml of anti-Bb IgG antibodies confirmed by Euroline-WB assay, whereas serum diluted at the same IgG concentration was negative. Such results demonstrated intrathecally restricted production of these antibodies, which is a key feature of Lyme neuroborreliosis.[3]

Intravenous ceftriaxone (2 g daily) was given for 2 weeks and the weakness slowly improved thereafter. The clinical examination normalised 6 weeks after the start of treatment. A follow-up MR examination performed 6 months later demonstrated complete normalisation of the spinal status.

The diagnosis of Lyme neuroborreliosis was therefore biologically supported by the positive blood IgM and IgG serology for Bb, together with the strong intrathecal synthesis of IgM and IgG antibodies seen on analysis of CSF. In addition, the patient's status dramatically improved with ceftriaxone antimicrobial treatment. The clinical presentation was, however, atypical, because of the absence of pain with nocturnal exacerbation, the purely motor involvement with absent tendon reflexes in the upper right limb and the diffuse fasciculations in the shoulders. These atypical signs and symptoms nicely corresponded to the cervical spinal cord imaging showing selective inflammatory involvement of anterior grey matter and spinal roots.

Recently, eight documented cases of early subacute Bb myelitis have been reported.[2] In six of these cases, painful radicular symptoms appeared before spinal cord signs. Most patients exhibited CSF mononuclear pleocytosis and cervical spinal cord lesions on MRI. All but one patient had a favourable outcome with appropriate intravenous antibiotherapy. Absence of encephalitic involvement and cranial nerve palsy, frequent combination with meningoradiculitis and good response to antibiotherapy are key features allowing discrimination between early and late variants of Bb myelitis.

Leptomeningeal enhancement and nerve root enhancement of the cauda equina on post-contrast T1 weighted MR images have been reported in spinal Lyme neuroborreliosis.[4] Enhancement of the cervical nerve roots has also been reported in this condition.[5] Concomitant enhancement of the leptomeninges, anterior horns and anterior radicelles of the cervical spine has never been described to date. The close correlation between clinical and MR features was a prominent feature of this clinically and radiologically atypical case of Lyme neuroborreliosis.

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The vestibulo-ocular reflexes during head impulse in Wernicke's encephalopathy

Ocular motor findings in Wernicke's encephalopathy (WE) include gaze evoked nystagmus (GEN), central positional nystagmus, weakness of abduction, internuclear ophthalmoplegia and horizontal or vertical gaze palsy to total ophthalmoplegia. Another feature of WE is vestibular paresis.[1 2] Previous studies documented hypoactive vestibular responses to both caloric and rotational stimuli, and a short vestibulo-ocular reflex (VOR) time constant. To address differential susceptibility of individual semicircular canals (SCC) according to stimulation frequency, we measured high acceleration VOR of the individual SCC using head impulse manoeuvres, and the low frequency VOR using bithermal caloric and rotatory chair tests in two patients with WE.

Case reports

Patient No 1

A 63-year-old woman had undergone Whipple's operation because of carcinoma of the ampulla of Vater 1 month previously and had received total parenteral nutrition (TPN). Two weeks after initiation of TPN, she began to suffer from anorexia, vomiting and vertigo which progressed to psychomotor slowing, apathy, forgetfulness and ataxia. On examination, she was awake but not attentive. She was fluent, but comprehension was impaired. The pupils were normal. Horizontal saccades were slowed and limited, and the limited ocular motor range did not improve with oculocephalic stimulation of the VOR. Other findings included horizontal GEN, limb dysmetria and severe truncal ataxia. T2 weighted MRIs showed hyperintense lesions at the periaqueductal gray matter, medial thalami and dorsal medulla (fig 1A). Three days after thiamine supplementation (100 mg intravenously daily), mental status, ataxia and horizontal gaze palsy began to improve.

Patient No 2

A 40-year-old man with diabetes mellitus presented with a 4 day history of progressive vertigo, ataxia, apathy and psychomotor slowing. He had been a heavy drinker for several years. Examination showed decreased alertness, perceptual disturbance and impaired memory. Horizontal saccades were slow and limited to approximately 30° from the primary position in both directions and the ocular motor range did not improve with oculocephalic manoeuvres. He also showed horizontal GEN and bilateral limb dysmetria. He could not stand unaided. Brain MRI was normal. With parenteral thiamine supplementation, mental status, ocular signs and ataxia recovered rapidly. However, memory impairments and GEN remained for 2 weeks after symptom onset.