Discussion

Rhabdomyolysis, ARF, acute hepatotoxicity and transverse myelitis (TM) are well known complications of intravenous heroin assumption.^{1,2} A single case of acute myelopathy as a consequence of heroin inhalation has been reported following, in common with other cases, a period of abstinence preceding the event; moreover, recovery was almost complete 1 month later.¹ No cases of acute MDMA myelopathy are known; rhabdomyolysis is known to complicate MDMA consumption.³

This case particularly highlights selective involvement of the lumbar motoneurons, as supported by neurophysiological and neuro-radiological evidence. Unexcitability of motor fibres and complete denervation in the lower limbs and paraspinal muscles, with spared SAPs and SSEPs, suggest damage of SAH and/or ventral root axons. MRI confirmed selective T2 hyperintensity confined to the SAH and nerve roots, with intense enhancement in both, spreading from D10 up to the conus (fig 1). This unique MRI pattern has never been reported previously to our knowledge, and differs from MRI patterns observed in TM and radiculitis. In TM, MRI can sometimes show an inflammatory lesion localised within the cord, usually not involving more than 3–4 vertebral segments (more often at the dorsal level), hyperintense in T2 weighted images and with nerve root sparing. In the acute phase, enhancement is generally present, but not involving the nerve roots.⁴ In radiculitis (eg, Guillain-Barré syndrome), MRI signal alterations are commonly localised to spinal anterior roots, without any involvement of the spine.⁵ Moreover, the MRI pattern did not suggest a spinal infarction. The anterior spinal artery derives from a single anterior radiculo-medullary artery (Adamkiewicz artery) that arises from L3 upwards; spinal infarction is rare and occurs mostly in the cervical spine. MRI abnormalities on sagittal T2 weighted images in our patient are not consistent with typical "pencil-like" hyperintensities and cord enlargement.

Therefore, taken together, the clinical, neuroradiological and neurophysiological data suggest that the pathological process primarily affected SAH conditioning motoneuron cell death, and then nerve roots and lumbar plexus as a consequence of wallerian degeneration.

Suggested pathogenetic mechanisms of heroin associated acute myelopathy include hypotension watershed zones ischaemia, vasculitis and hypersensitivity reaction. Among the most credited hypothesis is the possibility of a direct toxic effect of heroin.^{1,2} In our case, we speculate that a pathogenetic role played by MDMA cannot be excluded and may contribute towards an explanation of some of the atypical features such as the lack of a clear period of abstinence from heroin, as described in most cases of opiate toxic myelopathy, and the unusually high dose of MDMA inhaled. Moreover, it has been proved that the lumbar motoneurons express high levels of serotonin receptors (mediating MDMA effects) whereas opiates receptors are three times more represented in the dorsal horns compared with the ventral horns of the spinal cord.⁶

This case increases our knowledge of the possible severe acute toxic effects of intranasal insufflation of heroin and ecstasy, often considered by the public as "safe" drugs. The selective involvement of the anterior horns of the spinal cord indicates a poor functional prognosis and suggests a specific, but still

unknown, pathogenetic effect on the spinal motoneurons, possibly toxic or immunopathological in nature.

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Unusual case of tick borne encephalitis with isolated myeloradiculitis

Tick borne encephalitis (TBE) virus causes the most important arthropod transmitted disease in central Europe. In endemic areas, TBE has an incidence of 1.2 per 10 000 and a mortality of approximately 1%. The TBE virus is a neurotropic human pathogen. The most common presentations are meningitis (49%), meningoencephalitis (41%) and meningoencephalomyelitis (10%).¹ Patients with concomitant spinal cord involvement are thought to be affected more severely and mechanical ventilation is often necessary.²

We present an unusual TBE case with an isolated myelitis without signs of meningitis or meningoencephalitis.

Case report

A 43-year-old man from the Black Forest area, Germany, who had not had TBE vaccination developed symptoms of gastroenteritis with fever (diarrhoea, nausea and vomiting) lasting for approximately 1 week. Two days after recovery from the gastrointestinal symptoms he experienced severe pain in his shoulders followed by proximal pareses of his brachial muscles without signs of meningism. As the symptoms were not ameliorating during the

following days, the patient was admitted to hospital. At that time he had severe pareses of the upper extremities and mild pareses of the lower extremities. All tendon reflexes appeared very brisk but pyramidal signs were absent. Sensory testing was normal. The patient recalled a tick bite several weeks ago, after having collected mushrooms in the forest.

The following laboratory findings were abnormal: increased C reactive protein 8 mg/l, leucocytosis 12 200/μl, cerebrospinal fluid with mild pleocytosis (49 cells/μl: 80% lymphocytes, 20% monocytes), a clearly increased total protein content (2070 mg/l), local IgM synthesis of 83.5% and an increased IgG index of 0.81. Serological testing of the CSF and serum showed positive IgM and IgG antibody reactivity against TBE virus. A virus specific IgG CSF/serum index of 3.69 (normal value <2) confirmed high intrathecal specific antibody production. Coinfection with *Borrelia burgdorferi* was excluded. Moreover, there was no evidence of further possible aetiologies, such as infection by *Treponema pallidum*, herpes simplex virus 1 and 2, varicella zoster virus, enterovirus, HIV, Epstein-Barr virus or cytomegalovirus, or evidence of a vasculitis or collagenosis.

On electromyography and electroneurography, axonal lesions were seen in the left deltoid (C5/6), the left triceps (C7/8) and the left abductor digiti minimi manus (C8/Th1). There was normal neurography of the right and left radial nerve, the left ulnar and the left median nerve, and thus a brachial plexus lesion was excluded. In summary, there was evidence of a polyradiculitic or anterior horn lesion from C5 to Th1 (maximum at C5 and C7).

Motor evoked potentials showed delayed central motor conduction to both legs and the right arm. Tibial somatosensory evoked potentials were pathological on the left side, also with a central delay. The clinical and electrophysiological pattern was compatible with an incomplete myelitis involving the anterior horn cells.

Spinal MRI performed 25 days after symptom onset was normal. To exclude mild encephalitis, cranial MRI and electroencephalography were performed. Both examinations showed normal findings.

The patient recovered slowly and incompletely over the following weeks. The pareses were slightly improved, in particular the pareses in the distal upper extremities. The weakness of the proximal arm muscles remained almost unchanged.

Discussion

The most frequent symptoms of TBE are meningitis, meningoencephalitis or meningoencephalomyelitis. Polio-like syndromes with polyradiculitic manifestations can complicate the disease.^{3–5} These reported cases of myeloradiculitis in TBE virus infections, however, showed additional signs of meningoencephalitis or cranial nerve involvement.⁵ To the best of our knowledge, only one case with an isolated anterior horn lesion of the C3 to the Th1 level has been reported.⁶ Even in this case, cranial MRI showed two cerebral lesions.

The prognosis of these cases with polio-like symptoms seems generally to be poor, with only slow and minor recovery, and persistent severe amyotrophy and pareses.^{4,6}

We have presented the second case of a patient with an isolated myelitis caused by a TBE virus infection. In contrast with the patient described by Beer *et al.*,⁶ our patient

not only showed impairment of the anterior horn neurons but also involvement of the descending spinal pathways suggestive of incomplete transverse myelitis. This presumption was supported by the clinical finding of brisk tendon reflexes and a central conduction delay demonstrated by motor evoked potentials. Respiratory muscles were not affected. An additional polyradiculitis cannot be differentiated from an anterior horn lesion by clinical symptoms or electrophysiologically. One argument in favour of a polyradiculitis was the clearly increased total protein in our patient. MRI did not disclose any spinal cord lesion. A low incidence of cerebral or spinal MRI lesions in TBE infection has also been reported elsewhere.¹ Thus MRI is of limited value in the diagnosis of TBE.¹ This may be explained by the rapid resolution of early T2 hyperintensities. Resolution of MRI lesions despite the limited clinical improvement after 6 weeks has been described by Beer *et al.*⁶ This seems to be in contrast with lesions in brain parenchyma in which a good correlation between clinical improvement and restitution of T2 abnormalities has been found.²

This unusual case shows that even in patients presenting with incomplete transverse myelitis without signs of meningitis or meningoencephalitis, a TBE virus infection should be considered as a differential diagnosis.

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Mild clinical expression of Lambert–Eaton myasthenic syndrome in a patient with HIV infection

Neuromuscular complications of HIV are related to immunodeficiency, direct cytotoxicity of the virus or side effect of the treatments. Autoimmune disorders involving the nervous

system, including Guillain–Barre syndrome, myositis and vasculitis, have been described in association with HIV. Neuromuscular junction autoimmune diseases such as myasthenia gravis have been occasionally reported in patients with HIV, whereas the Lambert–Eaton myasthenic syndrome (LEMS) has never been described. We report an unusual case of paucisymptomatic LEMS in a patient with HIV infection.

Case report

A 42-year-old non-smoker, HIV positive, black African male was referred to us with a 2 year history of progressive but fluctuant weakness of the lower limbs. Comorbidities included chronic inactive hepatitis B with undetectable viraemia and a history of L4–5 laminectomy for a spinal canal stenosis. Treatment with highly active antiretroviral therapy (HAART) was initiated 6 months before hospitalisation, although neither biological nor clinical signs of immunosuppression were detected.

At the time of admission, he did not complain of diplopia, speech or swallowing, but had a dry mouth as the only autonomic symptom. Examination of the cranial nerves revealed a bilateral ptosis that was not modified by Simpson's test (ie, ptosis worsened by effort). Examination of the upper limbs was normal except for symmetrical weak tendon jerks. Examination of the lower limbs revealed a paresis, which predominated on the iliopsoas muscles (grade 4 on the Medical Research Council scale). The muscle palpation and the sensory multimodal examination were normal. The findings from the remainder of his neurological and general examination were normal.

A repetitive low rate stimulation (3 Hz) test revealed a significant decrement of up to 40% in the upper limb muscle (fig 1A). High rate (50 Hz) stimulations, in contrast, induced an incremental response, from mild (+80%, fig 1B) to marked (+750%, fig 1C). Single fibre electromyography performed in the left extensor digitorum communis confirmed a severe defect of neuromuscular transmission with a marked increase in jitter and numerous blockings (fig 1D). These results were highly suggestive of LEMS.¹

Serum autoantibodies directed against voltage gated calcium channels (VGCC) were positive (72 PM, normal range ≤ 45 PM), and their presence were confirmed 1 year after hospitalisation (107 PM). Other autoantibodies which tested as negative were antiacetylcholine receptor, antimuscle specific tyrosine kinase, antismooth muscle, antistriated muscle, antithyroglobulin, antithyroperoxidase and antinuclear antibodies.

Paraclinical investigation, including total body CT and positron emission tomography scan, prostate specific antigen, carcinoembryonic antigen, total testosterone, alpha-fetoprotein, beta2-microglobulin and immunoelectrophoresis of serum protein were normal. HTLV-1 serology was negative. Total CD4⁺ cell count was $>400/\text{mm}^3$, and viraemia was <50 copies/ml. Brain and spinal cord MRI and quadriceps surgical biopsy were also normal.

The patient received intravenous immunoglobulin G infusions (0.4 g/kg) over 5 days, associated with pyridostigmine (up to 600 mg/day), with no effect on symptoms. Introduction of 3,4-diaminopyridine was refused by the patient. At the 1 year follow-up, the patient

Figure 1 (A) Low rate (3 Hz) repetitive stimulation test; recordings at the left hand over the abductor digiti minimi. A 40% decrement is recorded. Note the continuous decrease in M response from the 10th stimulus, which is not the usual myasthenic pattern but is suggestive of Lambert–Eaton myasthenic syndrome, and the amplitude of the initial M response within the normal range (8.6 mV). (B) High rate (50 Hz) repetitive stimulation test; recordings at the left hand over the abductor digiti minimi. A mild 80% increment is recorded. (C) High rate (50 Hz) repetitive stimulation test; recordings at the left foot abductor hallucis brevis. A marked 750% increment is recorded. Note that amplitude of the initial M response is markedly reduced (0.3 mV). (D) Stimulated (10 Hz) single fibre electromyography performed in the left extensor digitorum communis. The jitter of the marked muscle fibre potential is strongly increased at 213 μV , and there is a 51% rate of blocking.