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.

was clinically stable with persistence of fluctuant weakness that was sometimes exacerbated and sometimes ameliorated by exercise. The biological, oncological and immunological screenings remained unchanged.

Discussion

LEMS is a rare disorder that can be paraneoplastic (60%) or non-paraneoplastic (40%). In both forms autoantibodies to VGCC are implicated.¹ In our patient, LEMS was established by clinical features (weakness sometimes ameliorated by exercise, autonomic symptoms, weak tendon jerks) and suggestive electromyography findings associated with positive anti-VGCC antibodies, known to be disease specific.²

Apart from HIV infection and HIV related treatments, we did not find any other trigger for the appearance of LEMS. The oncological screening was normal and no other concomitant autoimmune markers were detected. Interestingly, the spectrum of reported autoimmunity in HIV patients is increasing.³ In addition, HIV infection may lead to molecular mimicry, known to be a common mechanism of autoimmunity. Furthermore, HIV infection has been associated with B cell stimulation and many autoantibodies are reported in HIV patients.³ Sustained immune restoration secondary to the initiation of HAART has been associated with the appearance of autoimmune diseases, but neither significant immunosuppression nor high viraemia, or opportunistic infection, has been detected. In addition, no other neurological complication of HIV secondary to immunodeficiency or direct cytotoxicity of the virus was observed.⁴ Neuromuscular junction autoimmune disorders such as myasthenia gravis have been reported previously in HIV patients,⁵ whereas LEMS has never been described in association with HIV.

In this case report, the appearance of LEMS may have been triggered by modulation of the immune system secondary to HIV infection, although a fortuitous association cannot be excluded. Nevertheless, the mild clinical expression persisting after 1 year of follow-up, despite the absence of treatment, remains unusual and may suggest a direct influence of the virus on autoimmunity. Interestingly, other examples of a milder phenotype of the disorders already exist in HIV infected patients, such as progressive multifocal leukoencephalopathy. In addition, improvement in myasthenia gravis symptoms in HIV infection turning into AIDS has been described.

In conclusion, this report underscores (i) the possible link between autoimmune disorders and HIV and (ii) that investigation of a HIV patient with unexplained fluctuant weakness should include exploration of LEMS.

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Resolution of transverse sinus stenoses immediately after CSF withdrawal in idiopathic intracranial hypertension

The cause of idiopathic intracranial hypertension (IIH) remains unknown but catheter venography has shown that many patients have intracranial venous hypertension proximal to transverse sinus stenoses. These stenoses have subsequently been demonstrated on magnetic resonance and CT venography and it has been proposed that by reducing the passive resorption of CSF, intracranial venous hypertension due to these stenoses might be the cause of IIH. However, CSF withdrawal reduces venous sinus pressures, implying that venous hypertension is a secondary phenomenon.¹ Moreover, resolution of transverse sinus stenoses has been reported in three patients with IIH treated by CSF diversion procedures.^{2,3}

We report a patient with IIH in whom catheter and CT venography showed transverse sinus stenoses which resolved immediately after CSF withdrawal by lumbar puncture (LP).

Case report

A 35 year old woman presented with a 2 year history of headache and transient visual obscurations. She had papilloedema and constricted visual fields but no focal neurology. MRI of the brain was normal. At LP, the opening pressure was 35 cm H₂O with normal CSF constituents. IIH was diagnosed and treatment started with acetazolamide 250 mg three times daily.

One year later the patient was referred to our institution with persistent headaches and papilloedema for consideration of a CSF diversion procedure. Magnetic resonance venography at this time suggested stenoses in the anterior part of both transverse sinuses and she was further investigated to establish the degree and reversibility of these stenoses with reference to possible stenting.

Direct retrograde cerebral venography and CT venography were performed before and after drainage of 45 ml of CSF by LP. The opening pressure was 26 cm H₂O and closing pressure 2 cm H₂O. The baseline catheter venogram recorded a pressure of 30 mm Hg in the superior sagittal sinus and gradients of 19 mm Hg across stenoses at the anterior ends of both transverse sinuses (fig 1A). The right transverse sinus was dominant. Immediately after CSF withdrawal, sagittal sinus pressure fell to 7 mm Hg, the right transverse sinus stenosis resolved (fig 1B) and the pressure gradient resolved. The left transverse sinus was not re-examined. CT venography confirmed the morphological changes, showing expansion of both transverse sinuses (figs 1C, D). With these

Figure 1 (A) Cerebral venogram, tilted lateral view, shows a stenosis in the anterior part of the right transverse sinus (arrow). Numbers denote pressure (mm Hg) at each asterisk. (B) CT venogram axial source image shows a generally narrow right transverse sinus with a stenosis anteriorly. (C) Cerebral venogram after lumbar puncture (LP) shows resolution of the right transverse sinus stenosis and no pressure gradient. (D) CT venogram 1 h after LP confirms the right transverse sinus stenosis has resolved, with a general increase in sinus calibre. The left transverse sinus is not in the plane of section but responded similarly.