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.

results the patient was considered unsuitable for stenting.

Her symptoms improved for 2 weeks after CSF withdrawal but subsequently regressed. A ventriculoperitoneal shunt with an adjustable flow control valve was inserted 2 months later. She remained asymptomatic for 2 weeks after shunt insertion but then relapsed with headaches. The performance level of the valve was reduced from 2.0 to 1.5, and at the most recent follow-up, 1 year post-shunting, she was asymptomatic with normal visual acuity and no papilloedema.

Discussion

Although many patients with IIH have transverse sinus stenoses, there has been debate over whether these are the cause or effect of raised intracranial pressure (ICP), and if they are an effect, whether they are an epiphenomenon or an exacerbating factor. Patients with IIH may constitute a heterogeneous group with respect to this issue. Increases in ICP can compress the venous sinuses, and secondary compression of the venous sinuses has been demonstrated in a number of patients with IIH. King *et al* showed that the pressure gradients across transverse sinus stenoses in IIH resolved after CSF drainage.¹ Subsequently, investigators using magnetic resonance and CT venography have shown that transverse sinus stenoses in IIH can resolve after CSF diversion procedures. The case we describe implies that in some patients with IIH the calibre of the transverse sinuses responds immediately to changes in ICP.

Unlike CSF diversion, non-invasive reduction of CSF pressure does not seem to effect transverse sinus stenoses. A series of 14 patients with IIH and transverse sinus stenoses were treated with acetazolamide 250 mg twice daily. This normalised CSF pressure in nine patients but in all cases the transverse sinus stenoses persisted unchanged on MR venography.⁴ The authors suggested that CSF hypovolaemia induced by CSF diversion procedures causes the stenoses to resolve rather than a direct relationship between sinus calibre and CSF pressure alone. Our patient had a closing pressure of 2 cm H₂O after 45 ml of CSF was withdrawn, which supports this view. This phenomenon might be expected from the Monro-Kellie doctrine whereby a decrease in the volume of CSF within the rigid space of the skull is compensated for by an increase in the volume of blood in the venous sinuses.

Stenting of the transverse sinuses has been proposed as an alternative to CSF diversion procedures in patients with IIH on the assumption that transverse sinus stenoses play some part in causing symptoms.⁵ Intuitively, however, patients whose sinuses dilate in response to CSF withdrawal would seem to be inappropriate candidates for stenting. This, in turn, invites questions about what proportion of patients with IIH have sinuses that respond in this way and what would be the role of stenting in these cases. Stenting seems to have been successful in some patients but not in others.⁵ It is not known whether the sinuses responded differently to changes in CSF pressure in patients who did well with stenting compared with those who did not because the CSF withdrawal test was not a part of their diagnostic workup. For the same reason it is also not known whether, in patients in whom stenting was successful, the stent addressed

the primary cause of raised ICP or simply mitigated an exacerbating factor.

The rapidity with which the calibre of the venous sinuses in these patients can respond to changes in ICP means that imaging should be interpreted with caution. It is now recognised that most patients with IIH have narrowing of the venous sinuses even to the point where those patients with IIH who do not have venous narrowing require some explanation. Perhaps in some of these patients ICP was not raised at the time of the investigation—for example, because of recent LP and CSF drainage. The appearances of the venous sinuses in patients with IIH should be interpreted with reference to the CSF pressure at the time of the examination.

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BOOK REVIEW

Neuromuscular disease. Evidence and analysis in clinical neurology

Michael Benatar. New Jersey: Humana Press Inc, 2006, US\$145.00, pp 462. ISBN 92-4-159392

This is a relevant, well written and fascinating book that will be of interest to general neurologists as well as those specialising in nerve and muscle. Michael Benatar attempts to emulate the style of a "Socratic dialogue" with a question-answer type format to analyse the complex literature surrounding neuromuscular disease. He frequently cites original studies and small series as well as systematic reviews, and is sceptical of claims made without evidence. It is

well laid out, easy to read and with tables adjacent to relevant text. Of course, in any text that reviews current literature, this will with time become dated, but it is a good starting point.

Part 1 is generic, applying to any clinical study, discussing basic epidemiology and statistical analysis, treatment trials and the analysis of prognosis, with clear discussion of logistic regression and survival curves. The rest is in logical anatomical groups.

The usefulness of electromyography (EMG) in the diagnosis of amyotrophic lateral sclerosis, for instance, reminds us that abnormal paraspinal EMG is present in 19% of patients with other neurological disorders, and that EMG of the sternocleidomastoid has a higher sensitivity than that of the tongue, in practice probably performed less often. The evidence for benefit from use of non-invasive positive pressure ventilation, percutaneous endoscopic gastrostomy or riluzole are perhaps less straightforward than many of us assume.

The discussion of chronic inflammatory demyelinating polyneuropathy (CIDP) highlights the eight papers with different electrodiagnostic criteria for this, often prepared for trials, but important to recognise. Fortunately, new criteria for defining CIDP are currently being designed from first principles. Perhaps the limits of available evidence become clear with the reported study that suggests that nerve biopsy may not improve diagnostic performance. In practice, supportive biopsy evidence may be invaluable for the uncertain physician actually starting the immunosuppression. The explosion of knowledge in the field of inherited neuropathies in recent years has left many neurologists confused, and this chapter provides a useful and detailed summary of the main findings in this ever changing world to date. There are extensive tables detailing symptoms, signs and EMG criteria in the main classes, although these mostly consist of heterogeneous molecular groups, limiting their usefulness.

There is a useful chapter on carpal tunnel syndrome. Although neurologists may be surprised to hear that meta-analysis showed that reporting nocturnal paraesthesia was not useful diagnostically, they probably will not be surprised that Tinel's and Phalen's tests were hopeless.

Dr Benatar clarifies that in the world of neuromuscular disease, the randomised controlled trial is often lacking, and treatments are frequently used on the basis of extensive uncontrolled literature. This should prove a useful reference book for any clinical neurologist, highlights areas needing further investigation for aspiring researchers and hopefully is the first in a series analysing the evidence in a range of subspecialties.

Carolyn Gabriel

NOTICE

J L Miller, G A James, A P Goldstone, *et al*. Enhanced activation of reward mediating prefrontal regions in response to food stimuli in Prader-Willi syndrome. *J Neurol Neurosurg Psychiatry* 2007;**78**:615–19. The online version of this paper has been replaced with a corrected version containing figures 1–3 in colour.