Abnormal evoked potentials in Miller-Fisher syndrome: further evidence of combined central and peripheral demyelination

We read with interest the short report by Ferrer et al about a patient with Miller-Fisher syndrome in whom CNS demyelination was documented by MRI a few years later. We would like to direct attention to an additional case supporting the possibility of combined central and peripheral demyelination in this syndrome.

A 66-year-old previously healthy woman was referred for diplopia of 24 hours duration, an unsteady gait, and distal paraesthesiae. There were no upper limb symptoms. Examination showed bilateral ptosis, complete external ophthalmoplegia, and severe axial and limb areflexia. On the second day of admission to hospital, progressive proximal muscle weakness developed accompanied by generalised areflexia. The plantar response were flexor. All sensory modalities were intact. Routine laboratory examinations, CT of her brain, and EEG were normal. The sterile cerebrospinal fluid obtained on the fifth day of admission to hospital was clear and under normal opening pressure, with a protein concentration of 0.75 g/l and no oligocellular bands.

Electrodiagnostic studies disclosed prolonged distal motor latencies, slow conduction, and prolonged F-responses in the median, ulnar, common peroneal, and posterior tibial nerves. Needle electromyography indicated active symmetrical generalised denervation. The visual evoked potentials (VEPs) showed a prolonged wave pattern with prolongation of N100 peaks (right 126 ms; left 128 ms; normal 100-110 ms), which retained their normal amplitude. Brain stem auditory evoked potentials (BAEPs) showed an impaired wave pattern with prolongation of N130-N200 and brain stem conduction time.

Within one week and with only conservative treatment, spontaneous improvement was marked for cranial nerves, and vertical and horizontal eye movements, but there was still partial limitation of vertical gaze. Neurological examination after three months showed a full range of eye movements, normal muscle strength, and deep tendon reflexes. The VEPs and BAEPs had returned to normal.

The aetiology and pathogenesis of the Miller-Fisher syndrome are not well outlined, particularly the location of the patho­logical changes. Most workers favour a peripheral origin, although others suggest brain stem inflammatory lesions or a combination of peripheral and central demyelination. Abnormal BAEPs have been described in only a few cases. Results from MRI have usually been normal, except in the reports of Giroud et al and Ferrer et al. Abnormal BAEPs and VEPs, together with evidence of demyelination on brain MRI, support the hypothesis that in the Miller-Fisher syndrome there is a combination of peripheral and central demyelination. Considering the dis similarities between central and peripheral myelin, however, this conclusion leaves the immunological mechanisms by which this process is mediated unclear.
Transient sixth-nerve palsy as the first presentation of acute leukaemia.

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