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.1

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 paresthesias. Examination showed bilateral ptosis, complete external ophthalmoplegia, and severe axial and limb ataxia. On the second day of admission to hospital, progressive proximal muscle weakness developed accompanied by generalised areflexia. The plantar responses were flexor. All sensory modalities were intact. Routine laboratory examinations, CT of her brain, and EEG were normal. The sterile acellular 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 oligoclonal 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) were markable for prolongation of the P100 peaks (right 126 ms; left 128 ms; normal 100–110 ms), which retained their normal amplitudes. Brain stem auditory evoked potentials (BAEPs) showed an impaired wave pattern with prolongation of N1, N2, and brain stem conduction time.

Within one week and with only conserva- tive treatment, spontaneous improvement was noted in the axial and horizontal eye movements, but there was still partial limitation of vertical gaze.

Neurological examination after three months showed a full range of eye move- ments, 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 patho- logical changes. Most workers favour a peripheral origin, although others suggest brain stem inflammatory lesions or a combination of both.4 Abnormal BAEPs have been described in only a few cases.5 Results from MRI have usually been normal,4 except in the reports of Giroud et al.6 and Ferrer et al.6

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 dissimilarities between central and peripheral myelin, however, this conclusion leaves the immunological mechanism by which this process is mediated unclear.

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Transient sixth-nerve palsy as the first presentation of acute leukaemia

It is not unusual for the cranial nerve to be affected early in the course of acute lymphoblastic leukaemia (ALL) and this is considered to be the result of meningeal infiltration.1,2 Unless specifically treated, the neurological deficit is usually progressive. We report here the case of a patient with transient sixth-nerve palsy as a presentation of ALL.

A 43-year-old man was admitted with an acute onset of diplopia. He had a three month history of night sweats and weight loss. He had previously had a systemic evaluation, which was normal except for a Westergren erythrocyte sedimentation rate of 40 mm/h, a transient disturbance in liver function tests (aspartate transaminase, alanine transaminase) and a constant increase in lactate dehydrogenase. Serological tests for hepatitis were negative.

On examination a complete left sixth- nerve palsy was found. The rest of the general and neurological examination was normal. A complete blood count and film, blood glucose concentration, thyroid function tests, antinuclear factor, serum immunoglobulins, immunoelectrophoresis and antibodies to acetylcholine receptor were all within normal limits. Lumbar puncture yielded a clear acellular fluid with 530 mg/l protein and 3.6 mmol/l glucose.

Computed tomography showed no leptomeningeal changes. A brain scan showed homogenous replacement of the skull bone marrow by an enhancing material with extension to the left cerebral hemisphere. A bone marrow specimen was therefore obtained and showed extensive infiltration with immature lymphoid cells, compatible with ALL.

A week after admission, there was spontaneous improvement in eye abduction. The patient was treated with a standard chemotherapy protocol for adult ALL. An Ommaya reservoir was inserted and intrathecal methotrexate and cytarabine were administered with cranial radiotherapy. The patient is currently in complete remission 12 months after the diagnosis of ALL.

Cranial nerve palsies as the first presentation of lymphoproliferative disorders, although uncommon, have already been reported.1,3 There is a single case report of transient sixth-nerve palsy in acute non-lymphocytic leukaemia.4 We believe that this is the first description of a patient with a systemic lymphoproliferative disorder, namely ALL, presenting with a spontane- ous resolving ocular motor deficit. The cause of the left sixth-nerve palsy in our patient was most probably leukemic involvement of the left cavernous sinus, as was shown by MRI. The transitory nature of the cranial nerve dysfunction may be due to transient pressure by leukemic cells on the cranial nerve itself or on the vasculature of the cavernous sinus, with subsequent ischaemia. The exact mechanism of transience remains obscure, however. To conclude, cranial nerve palsy led to the diagnosis of ALL with minimal systemic clues. Spontaneous resolution of a neurological deficit should not lead one to conclude that an underlying malignancy can be ruled out.

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