2960 TWO CASES OF MILLER-FISHER SYNDROME AND COVID-19 WITHOUT DETECTABLE GQ1B

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10.1136/bmjno-2024-ANZAN.14

Background Miller Fisher syndrome (MFS) is an uncommon variant of Guillain-Barré syndrome (GBS) characterised by ophthalmoplegia, ataxia, and depressed or absent muscle stretch reflexes. GQ1b ganglioside is a cell surface component present in the third, fourth, and sixth cranial nerves. IgG antibodies against GQ1b can develop following some infections due to molecular mimicry, and are an important mediator of the ophthalmoplegia seen in MFS.
Cases Case 1 was a 35 year old woman presenting at 9 weeks gestation with acute diplopia, having tested positive on respiratory NAAT to COVID-19 the preceding day. Initial examination revealed an isolated near complete right abducens palsy, but over two days she developed a bilateral complex ophthalmoplegia with non-fatiguable ptosis, bilateral ataxia, and diffusely depressed or absent muscle stretch reflexes. Case two was a 42 year old man presenting with acute diplopia, having tested positive on respiratory NAAT to COVID-19 two days prior. Initial examination demonstrated right eye exophora and hypophoria ('down and out') consistent with a pupil-sparing partial oculomotor nerve palsy, and a fatiguable right eyelid ptosis. Upper limb muscle stretch reflexes were depressed, but there was no ataxia. Both patients tested negative for GQ1b, AChR and MuSK antibodies and received intravenous immunoglobulin, making prompt and complete recoveries.
Conclusions We systematically reviewed similar reported cases (37 total), and anti-GQ1b antibodies were only detected in 28% of 25 cases tested, compared to 83–85% in typical MFS. It is proposed that COVID-19-associated MFS may result from direct viral neurotropism, rather than being mediated by anti-ganglioside antibodies.

2961 TWO CASES OF 4-AMINOPYRIDINE-ASSOCIATED NEW ONSET STATUS EPILEPTICUS

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10.1136/bmjno-2024-ANZAN.15

Background The potassium channel blocker 4-aminopyridine (4-AP) has long been used as symptomatic therapy for multiple sclerosis (MS) to treat spasticity and improve ambulatory speed, though evidence remains lacking. 4-AP selectively blocks fast voltage-gated potassium channels and increases calcium influx at nerve terminals, enhancing neuromuscular transmission. 4-AP is also used in animal models of epileptogenicity via this mechanism, and its therapeutic use or overdose in MS has been associated with several cases of new-onset seizures, which is otherwise a rare symptom in MS.
Cases Case one was a 66 year old woman with primary progressive MS presenting with two days of impaired gait and hyperkinetic movements leading to convulsive status epileptics (CSE). Case two was a 53 year old man with relapsing-remitting MS presenting just two days later with a day of hyperkinetic movements and agitation leading to CSE. Both cases were taking 4-AP from the same compounding pharmacy, having recently been dispensed the same batch, and both required intravenous sedation and intubation in an Intensive Care Unit. In neither case was an alternative pathology identified to account for new-onset CSE, but both patients subsequently made a good recovery and remained seizure free.
Conclusions We systematically reviewed similar reported cases (16 in total) of seizures associated with 4-AP, and found at least half were attributable to either intentional or unintentional overdose or drug compounding error. The majority developed status epilepticus requiring intravenous sedation and intubation, yet nearly all made complete recoveries and remained seizure free after cessation of 4-AP.

2962 PARANEOPLASTIC PROGRESSIVE ENCEPHALOMYELITIS WITH RIGIDITY AND MYOCLONUS ASSOCIATED WITH MONOCLONAL B-CELL LYMPHOCYTOSIS IN THE SETTING OF LONGSTANDING METHOTREXATE USE

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10.1136/bmjno-2024-ANZAN.16

Background Progressive encephalomyelitis with rigidity and myoclonus (PERM) is a rare but debilitating disease within the stiff-person syndrome (SPS) spectrum and is characterised by muscle rigidity, spasms, myoclonus, dysautonomia and brainstem dysfunction. The exact pathogenetic mechanism is unclear, although there is an association with the presence of glycine receptor antibodies in serum and cerebrospinal fluid, and some cases are paraneoplastic.
Results Here we report a complex case of paraneoplastic PERM associated with an otherwise subclinical monoclonal B-cell lymphocytosis (MBL) of the non-chronic lymphocytic leukaemia (CLL) phenotype, which may be in turn likely secondary to long-term methotrexate use (i.e. methotrexate-associated lymphoproliferative disorder, MTX-LPD) or underlying autoimmune disease.
Conclusions PERM often has a paraneoplastic aetiology and likely exerts antitumour response against malignancies, thus potentially rendering the primary malignancies indolent or atypical in presentation. This is, to our knowledge, the first reported association between PERM and MBL, or between PERM and MTX-LPD.

2963 A TALE OF THE IGG4 - MUSK MYASTHENIA GRAVIS IN A PATIENT WITH IGG4 RELATED DISEASE

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10.1136/bmjno-2024-ANZAN.17

Case A 73-year-old oriental female was admitted with 4 years of fluctuating dysarthria and dysphagia. She had breast cancer in remission and stable IgG4 disease with prior intermittent low dose prednisolone and rituximab therapy.

Following COVID-19 infection, she developed progressive dysphagia with weight loss of 13kg, limb weakness and blurred vision.

On initial neurological exam, she was poorly muscled, without fasciculations. She had bifacial weakness, complex ophthalmoplegia, lingual and palatal weakness associated with