2371 NOT ALL NMDA ANTIBODIES ARE EQUAL: TWO CASES OF GLIOBLASTOMA MULTIFORME PRESENTING WITH SUBACUTE NEUROLOGICAL SYNDROMES AND POSITIVE NMDA ANTIBODIES

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While the specificity of anti-NMDAR-IgG in serum and particularly in CSF is high, false positive results do occur and may lead to an erroneous diagnosis of autoimmune encephalitis. We describe two cases of glioblastoma multiforme (GBM) presenting with neurological syndromes and positive anti-NMDA antibodies. Initial immunotherapy was given based on autoimmune findings, while clinical/radiological red flags in both cases lead to the final diagnosis of GBM. Case 1: A 64-year-old male presented following seizures on a background of subtle mood changes. MRI brain demonstrated increased T2/FLAIR signal of the left temporal lobe, and he returned positive serum and CSF anti-NMDAR antibodies. Intravenous immunoglobulin (IVIg) was administered with initial improvement however due to atypical imaging findings he underwent a brain biopsy confirming grade 4 GBM. Case 2: A 38-year-old male presented with refractory status-epilepticus following subacute symptoms of neuropsychiatric changes. MRI brain again showed increased T2/FLAIR signal affecting the left temporal lobe. He returned positive serum NMDAR antibodies that were negative in CSF (delayed lumbar puncture due to initial concerns of raised intracranial pressure). He received plasmapheresis, methylprednisolone, IVIg and rituximab, with prompt clinical and initial radiographic improvement. Interval MRIs however demonstrated atypical progressive imaging changes and a brain biopsy was performed, also confirming a grade 4 GBM.

Conclusions Clinicians should have a heightened awareness of the possibility of intrinsic cerebral tumours presenting as a mimic of NMDAR-mediated encephalitis. Clinical and/or radiological progression despite immunotherapy and atypical features at presentation can alert clinicians to this entity.

2372 EFFECT OF DELAY TO FIRST-LINE IMMUNOTHERAPY ON OUTCOMES IN AUTOIMMUNE ENCEPHALITIS

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Objectives Autoimmune encephalitis (AE) encompasses a spectrum of clinical syndromes that can be associated with diagnostic and therapeutic delays. Our aim was to determine whether delay to immunotherapeutic intervention in patients with AE leads to worse outcomes, as measured by the Clinical Assessment Scale in Autoimmune Encephalitis (CASE) score.[1]

Methods This retrospective cohort study examined medical records of all patients with AE[2] treated with immunotherapy at two tertiary epilepsy centers. Early treatment was defined as immunotherapy within 90 days of symptom onset. CASE scores were determined at initiation of immunotherapy, and at 6 months and 12 months post treatment. Covariates included age, sex, AE type, exposure to second line treatment, and CASE score at start of immunotherapy.

Results Thirty patients met study inclusion criteria. Mean CASE score at start of immunotherapy was higher in patients treated early (10.7) compared to late (5.1, $p=0.0059$). However there was no difference between the two groups at 6 months (4 vs 2.7, $p=0.06$) and 12 months (3.6 vs 2.3, $p=0.06$). Patients treated early had a greater improvement in CASE score at 6 months (reduction of 6.7 vs 2, $p=0.005$) and 12 months (reduction of 7.7 vs 3.3, $p=0.03$). Regression modelling showed the only significant covariate associated with a greater change in score at 6 months was higher CASE score at start of treatment ($p<0.001$).

Conclusion Delayed immunotherapy in AE patients still provides clinical benefit. AE patients with a delayed diagnosis or subacute course should be considered for induction immunotherapy.

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2373 ANTI-LGI1 ASSOCIATED MYOPATHY IN SETTING OF NEUROMUSCULAR HYPEREXCITABILITY SYNDROME

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Background Anti-leucine-rich glioma inactivated 1 (LGI1) antibodies are most commonly associated with autoimmune encephalitis, however the clinical spectrum is evolving, and peripheral neuropathy and neuromuscular hyperexcitability syndrome have recently been recognised.[1] [2] We describe a case of anti-LGI1 associated hyperexcitability syndrome associated with myopathy, which has not been described previously.

Case Description A 56-year-old man presented with three months of myalgia, decreased exercise tolerance, dysaesthesias, hyperhidrosis, insomnia and diarrhoea. Examination revealed myokymia in orbicularis oris and mentalis, calf fasciculations, and absent ankle reflexes. Blood tests showed an elevated Creatine Kinase (CK). Electromyography confirmed fasciculations in right gastrocnemius and myokymia in mentalis. Magnetic Resonance Imaging (MRI) of the lower limb demonstrated hyperintensity of the mid left gastrocnemius with a biopsy of this site showing myopathic/dystrophic changes without inflammation. Further serum testing revealed positive anti-LGI1 antibodies.

He was diagnosed with anti-LGI1 associated neuromuscular hyperexcitability and myopathy. Administration of corticosteroids and azathioprine resulted in complete clinical remission and normalisation of CK.

Conclusion Myopathy with raised CK has not previously been reported as part of the anti-LGI1 clinical spectrum. Neurologists should consider CK testing in patients with anti-LGI1 antibodies and peripheral neuromuscular symptomatology, as it may assist with diagnosis of associated myopathy and provide an object marker for disease monitoring.