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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10.1136/bmjno-2022-ANZAN.127

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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10.1136/bmjno-2022-ANZAN.128

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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10.1136/bmjno-2022-ANZAN.129

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.

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2376 IMMUNOADSORPTION, THERAPEUTIC PLASMA EXCHANGE AND PREGNANCY IN WOMEN WITH MULTIPLE SCLEROSIS – A CASE SERIES FROM GERMANY

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10.1136/bmjno-2022-ANZAN.130

Objectives The aim of this study was to asses pregnancy outcomes and disease activity after apheresis in women with multiple sclerosis (MS).

Methods Women who relapsed during pregnancy and received apheresis, documented in the German Multiple Sclerosis and Pregnancy Registry (DMSKW) are presented. Pregnancy outcomes and disease course were captured with a standardized questionnaire in regular telephone interviews during pregnancy and post-partum. Relapse, treatment data and EDSS-values were verified by treating physicians.

Results In 28 pregnancies 15(53.6%) women received immunadsorption, 8(28.6%) received therapeutic plasma exchange and 5(18%) received both on separate occassions due to severe relapses, with a median(range) EDSS of 4(1.5–9.5) during relapse. The median (range) number of apheresis cycles was 5(2–8). Most women (92.86%) received corticosteroids prior to apheresis with a mean(SD) of 1950mg(375mg). 28 live-births with 2(7.4%) congenital malformations were reported, transposition of the great arteries and hip dysplasia. 3(10.7%) SGA births were reported and 6(21.4%) babies were born prematurely. The mean(SD) birthweight was 2688g (792.2g) and excluding pre-term births, 3006g(514.1g).

Conclusion Women with severe relapses receiving a combination of corticosteroids and apheresis during pregnancy have a higher risk for preterm birth and reduced birthweight. Our data add useful information on pregnancy outcomes after treatment with apheresis during pregnancy, however is limited by the small sample size. Also, concurrent corticosteroid exposure is a possible confouding factor, which will be investigated in further analysis. Women with highly active MS should be counselled and plan their pregnancy beforehand.

2377 SPECIALIST COGNITIVE MANAGEMENT IN A MULTIPLE SCLEROSIS AND NEUROIMMUNOLOGY CLINIC

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10.1136/bmjno-2022-ANZAN.131

Objectives The Royal Melbourne Hospital newly hosts a dedicated cognition clinic for patients with multiple sclerosis (MS) and other neuroimmunological disorders. An audit of the cognitive unit was conducted following the first eighteen months of service operation to describe the approach and practice of cognitive diagnosis and to characterize the first cohort of patients to receive care.

Methods Medical records of all patients referred to the unit were reviewed for patient diagnosis, basis of cognitive referral, psychometric profile, and management outcomes. Cognitive dysfunction was determined using the Brief International Cognitive Assessment for MS (BICAMS) with scores less than 1.5SD below the mean considered impaired.

Results The clinic serviced patients across cognitive, neurological, psychological and functional referral points. Of 127 patients seen, 104 had MS. 37%, 11% and 2% of MS patients were impaired on at least 1, 2 or 3 subtests of BICAMS, respectively. Recommendations for patient management frequently included referral for clinical psychology (38%), further neuropsychological management (13%) or psychiatric opinion (7%), as well as other interventional strategies for fatigue and pain.

Conclusion Consistent with the MS scientific literature, cognitive impairment was heterogenous in the cohort. The rate of diagnosed cognitive impairment, however, diverges from published prevalence estimates. This unique sample of patients referred to the Royal Melbourne Hospital Multiple Sclerosis Centre Cognitive Unit provides a new window to the MS patient, outside parameters set for recruited research samples. The high service demand from both patients and clinicians alike demonstrates the need for specialist MS and neuroimmunological cognitive opinion.

2378 GNE MYOPATHY WITH A NOVEL LIKELY PATHOGENIC GENETIC VARIANT

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10.1136/bmjno-2022-ANZAN.132

Background GNE myopathy is a rare autosomal recessive disease caused by mutation in the GNE gene, also known as quadriceps sparing myopathy or hereditary inclusion body myopathy. The estimated worldwide prevalence is 1/1000,000. There are over 150 mutations known to be causative for GNE myopathy. We describe a case of GNE myopathy with compound heterozygosity for one pathogenic and one novel, likely pathogenic variant in the GNE gene.

Case Report A 32-year-old previously healthy woman of Indian descent was referred to our clinic with progressively worsening waddling gait and weakness in her hands. She was uncertain about where her weakness started first and she had no significant family history. On examination, she had weakness of finger flexion, thumb abduction, hip flexion, knee flexion and ankle dorsiflexion with decreased reflexes and normal sensation. Muscle biopsy suggested hereditary inclusion body myopathy and it was confirmed by neuromuscular panel showing heterozygous C.2179G>A (12p.val727Met); C.1368C>A (7p.Ser456Arg) in the GNE gene. She was referred to orthopaedics for foot splints, physiotherapy for muscle strengthening and occupational therapy for impaired hand function. Her condition remained stable at present.

Conclusion GNE myopathy is a rare distal myopathy due to variants in GNE gene. While C.2179G>A being a common variant in Indian population, C.1368C>A is a novel variant likely causative for the disease. Currently there is no disease modifying therapy and management is centred around