associated with renal cell carcinoma and anti-NMDAR and EBV DNA detected in cerebrospinal fluid (CSF).

Methods This was a personal case of the authors, with documentation and investigations reviewed from patient medical records at Sir Charles Gairdner Hospital.

Results A 55 years old immunocompromised female patient presented with two weeks of increasing ataxia, oscillopsia and tremor, preceded by 3 weeks of headache, nausea and vomiting. This was on a background of three failed renal transplants for IgA focal segmental glomerulosclerosis and recent resection of stage I renal clear cell carcinoma. Examination was consistent with opsoclonus-myoclonus-ataxia syndrome. There were no features on encephalopathy apart from mild emotional lability. CSF results: lymphocytosis with an elevated protein, positive EBV using polymerase chain reaction and positive anti-NMDAR. Treatment: Plasma exchange, Rituximab, intravenous acyclovir for two weeks followed by oral acyclovir for 3 months. Improvement was marked, although with residual myoclonus on 2 month follow-up.

Conclusions This rare case of paraneoplastic double-positive EBV and anti-NMDAR opsoclonus-myoclonus syndrome in an immunocompromised patient demonstrates the broadening clinical phenotype of anti-NMDAR and highlights the contentious issue of EBV pathogenicity and treatment in an immunocompromised patient.

REFERENCES

- Garre, *et al.* Letter to the editor: EBV-NMDA double positive encephalitis in an immunocompromised patient. *Journal of the Neurological Sciences* 2019; **(396)**:76–77.
- Zhao CZ, Erickson J, Clinical Reasoning Dalmau J. Agitation and psychosis in a patient after renal transplantation. *Neurology* 2012; **79**:e41–e44.
- Derksen S, Goraj B, Molenaar J, van der Hoeven JJC. Severe anti NMDA encephalitis and EBV infection. *Netherlands J Critic Care* 2013; **17**:19–21.
- Cohen DA, Lopez-Chiriboga AS, Pittock SJ, Gadoth A, Zekeridou A, Boilson BA, *et al.* Posttransplant autoimmune encephalitis. *Neurol Neuroimmunol Neuroinflamm* 2018; **5**:e497.
- Armangué T, *et al.* Clinical and immunological features of opsoclonus-myoclonus syndrome in the era of neuronal cell surface antibodies. *JAMA Neurology* 2016; **73**(4):417–424.
- Klaas JP, *et al.* Adult-onset opsoclonus-myoclonus syndrome. *Archives of Neurology* 2012; **69**(12):1598–1607.

066

CONTACTIN-1-MEDIATED CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY (CIDP) PRESENTING AS AN ACUTE CASE OF GUILLAIN BARE SYNDROME (GBS)

Shane Cameron, Rami Haddad. *Orange Health Service, Orange, NSW, Australia*

10.1136/bmjno-2021-ANZAN.66

New and emerging evidence describing the role of autoantibodies to nodal and paranodal proteins in the pathogenesis of chronic inflammatory demyelinating polyneuropathy (CIDP) has arisen over the past decade, with significant diagnostic, therapeutic and prognostic implications. Although rare, CIDP with anti-contactin-1 (CNTN1) IgG4 antibodies has a distinct pathogenesis and clinical phenotype that differs from both typical CIDP and its other atypical variants. Here, we report the case of a young normally well 49-year-old man from rural Australia with severe refractory anti-CNTN1-mediated CIDP who presented like a case of Guillain-Barré syndrome (GBS) who improved dramatically after

chemoimmunotherapy with Rituximab and who also had an unexpected late response to subsequent treatments with intravenous immunoglobulin (IVIg). In reporting this case, we hope to highlight important considerations in the diagnosis and treatment of patients with severe refractory CIDP, and especially those patients with anti-CNTN1 seropositive disease.

REFERENCES

- Vural A, Doppler K, Meini E. Autoantibodies against the node of ranvier in seropositive chronic inflammatory demyelinating polyneuropathy: diagnostic, pathogenic, and therapeutic relevance. *Frontiers in Immunology* 2018; **9**:1–14.
- Miura Y, Devaux JJ, Fukami Y, Manso C, Belghazi M, Wong AHY, *et al.* Contactin 1 IgG4 associates to chronic inflammatory demyelinating polyneuropathy with sensory ataxia. *Brain* 2015; **138**:1484–1491.
- Bunschoten C, *et al.* Progress in diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy. *The Lancet Neurology* 2019; **18**(8):784–794.
- Querol L, Nogales-Gadea G, Rojas-García R, *et al.* Antibodies to contactin-1 in chronic inflammatory demyelinating polyneuropathy. *Annals of Neurology* 2013; **73**:370–380.
- Querol L, *et al.* Rituximab in treatment-resistant CIDP with antibodies against paranodal proteins. *Neurology, Neuroimmunology & Neuroinflammation* 2015; **2**(5):1–6.

067

NEUROFILAMENT LIGHT CHAIN CONCENTRATION PREDICTS RISK OF RELAPSE IN PARTICIPANTS WITH RELAPSING MULTIPLE SCLEROSIS IN PHASE 3 OZANIMOD TRIALS

¹Sarah Harris, ²Giancarlo Comi, ³Bruce AC Cree, ⁴Lawrence Steinman, ¹James K Sheffield, ⁵Harry Southworth, ⁶Ludwig Kappos, ⁷Jeffrey A Cohen. ¹Bristol Myers Squibb, Princeton, New Jersey, USA; ²Department of Neurology, San Raffaele Scientific Institute, Vita-Salute San Raffaele University, Milan, Italy; ³Department of Neurology, Weill Institute for Neurosciences, University of California San Francisco, San Francisco, California, USA; ⁴Department of Neurology and Neurological Sciences, Beckman Center for Molecular Medicine, Stanford University Medical Center, Stanford, California, USA; ⁵Data Clarity Consulting Ltd, Stockport, UK; ⁶Neurologic Clinic and Policlinic, University Hospital and University of Basel, Basel, Switzerland; ⁷Department of Neurology, Mellen Center for MS Treatment and Research, Cleveland Clinic, Cleveland, Ohio, USA

10.1136/bmjno-2021-ANZAN.67

Objectives Plasma neurofilament light chain concentration (pNfL-c) is increased in patients with multiple sclerosis (MS) and may serve as a biomarker for neurologic damage and disease activity in relapsing MS. We analyzed changes in pNfL-c and on-treatment risk of relapse with ozanimod vs interferon β -1a (IFN).

Methods In this post hoc analysis of the phase 3 SUNBEAM (NCT02294058; ≥ 12 months) and RADIANCE (NCT02047734; 24 months) trials, pNfL-c was measured at baseline and after 12 and 24 months of treatment with oral ozanimod 0.46 or 0.92 mg/d or intramuscular IFN 30 μ g/wk. Poisson generalized linear models were used to fit the number of relapses as a function of baseline pNfL-c and treatment group with an offset for duration. Predictive modeling of expected annualized relapse rate (ARR) was calculated using median percentage change in pNfL-c from baseline.

Results At end of treatment, median pNfL-c was reduced from baseline by 20%–23% ($P < 0.01$) and 23%–27% ($P \leq 0.0001$) with ozanimod 0.46 and 0.92 mg, respectively, and by 13%–15% with IFN. Higher baseline pNfL-c was associated with more relapses ($P < 0.0001$), and greater median reductions in pNfL-c from baseline were associated with lower ARR.

Predictive modeling estimated that a 25% reduction in pNfL-c, similar to that observed with ozanimod 0.92 mg, predicts an ARR (standard error [SE]) of 0.18–0.23 (0.4), whereas a 13% reduction, similar to IFN, predicts an ARR (SE) of 0.29–0.37 (0.04).

Conclusion Our findings support pNfL-c as a biomarker for relapsing MS disease activity. Ozanimod caused greater dose-dependent reductions in pNfL-c and ARR than IFN.

068

BRADYCARDIA AS A RARE NEUROCARDIAC PRODROME TO LEUCINE-RICH GLIOMA INACTIVATED-1 ANTIBODY ENCEPHALITIS

Sai Nagaratnam, Yun T Hwang, Elizabeth Reyneke. *Neurology, Gosford Hospital, Gosford, NSW, Australia*

10.1136/bmjno-2021-ANZAN.68

Introduction Leucine-rich glioma inactivated-1 antibody encephalitis has been associated with bradycardia as a neurocardiac prodrome.¹ Concurrent occurrence of cardiac arrhythmia and faciobrachial dystonic seizures have not previously been reported.

Case A 73-year-old male presented with a 6 week history of frequent episodes of an unpleasant sensation associated with sinus bradycardia requiring pacemaker implantation. Episodes continued despite pacemaker. He was diagnosed with a seizure disorder and commenced on levetiracetam without response.

Subsequently, on video EEG, subtle facial grimace and upper limb tonicities were captured, in keeping with faciobrachial dystonic seizures without an EEG correlate. MRI Brain showed no radiological evidence of encephalitis. Serum limbic encephalitis panel confirmed LGI1 antibodies. Other autoimmune and paraneoplastic antibodies were negative. He was treated with a course of corticosteroids. Induction dose of intravenous immunoglobulin was prematurely terminated after one dose due to MRSA bacteraemia and tricuspid valve endocarditis, necessitating removal of the pacemaker with no recurrence of seizures or bradycardia at follow up without further treatment.

Conclusion This case illustrates a rare presentation of LGI-1 antibody encephalitis with complete remission following incomplete induction course of intravenous immunoglobulin and corticosteroids. Neurocardiac prodrome as episodic bradycardia or asystole may precede the onset of encephalitis by approximately 2 months.^{1 2} There is a good response to immunotherapy, however relapse is common.³ This case illustrates that clinically atypical presentations of cardiac arrhythmia may warrant neurological review and raises a possibility that early initiation of immunosuppressive therapy may significantly alter the disease course of LGI-1 antibody encephalitis.

REFERENCES

- Naasan G, Irani SR, Bettcher BM, Geschwind MD, Gelfand JM. Episodic bradycardia as neurocardiac prodrome to voltage-gated potassium channel complex/leucine-rich, glioma inactivated 1 antibody encephalitis. *JAMA Neurology* 2014;**71**(10):1300–4.
- Nilsson AC, Blaabjerg M. More evidence of a neurocardiac prodrome in anti-LGI1 encephalitis. *Journal of the Neurological Sciences* 2015;**357**(1):310–1.
- van Sonderen A, Thijs RD, Coenders EC, Jiskoot LC, Sanchez E, de Bruijn MAAM, et al. Anti-LGI1 encephalitis. *Clinical Syndrome and Long-Term Follow-up* 2016;**87**(14):1449–56.

069

A PUTATIVE MECHANISM FOR SUBCORTICAL APHASIA

^{1,2}Shadi El-Wahsh, ³David Greenup, ⁴Gemma White, ^{5,6}Michael J Fulham, ^{3,7}Arun Aggarwal, ¹G Michael Halmagyi. ¹Neurology, Royal Prince Alfred Hospital, Sydney, NSW, Australia; ²South Western Sydney Clinical School, University of New South Wales, Sydney, NSW, Australia; ³Department of Rehabilitation Medicine, Balmain Hospital, Sydney, NSW, Australia; ⁴Speech pathology department, Balmain Hospital, Sydney, NSW, Australia; ⁵Department of Molecular Imaging (PET and Nuclear Medicine), Royal Prince Alfred Hospital, Sydney, NSW, Australia; ⁶Sydney Medical School, University of Sydney, Sydney, NSW, Australia; ⁷Neurology, Concord Repatriation General Hospital, Sydney, NSW, Australia

10.1136/bmjno-2021-ANZAN.69

Objectives The role of subcortical structures in language function are still poorly understood. We aim to provide a putative mechanism for subcortical aphasia through a structural and functional imaging-based case discussion.

Methods We present a case of subcortical aphasia due to basal ganglia hypertensive haemorrhage and discuss serial MRI and PET imaging findings to elucidate the mechanism of profound language impairment in acute subcortical pathology.

Results A 71-year-old right-handed architect presented with acute onset global aphasia and right-sided hemiparesis. CT imaging showed a flame-shaped left-sided basal ganglia haemorrhage. MRI brain showed a left basal ganglia haemorrhage without ischaemic or haemorrhagic damage to the overlying fronto-parietal cortex. FDG-PET imaging showed profound left fronto-parietal cortex hypometabolism, as well as ipsilateral caudate, putamen, thalamic and pontine hypometabolism. MR tractography identified truncation of the arcuate fasciculus around the left angular gyrus as well as disconnection of the left fronto-parietal association fibres. Over 12 weeks of rehabilitation, the patient began to generate verbal output and was discharged home with ongoing word finding difficulties, nominal aphasia, and semantic paraphasias. Progress PET imaging revealed persistent hypometabolism in the aforementioned regions.

Conclusion We believe this is an important educational case for neurologists regarding the presentation of aphasia due to isolated subcortical lesions and raises some interesting hypotheses regarding a putative mechanism for subcortical aphasia due to dominant hemisphere cortical inactivation.

070

HEMI-CORD INFARCTION FOLLOWING VERTEBRAL ARTERY DISSECTION IN A PATIENT WITH CONGENITAL HYPOPLASTIC VERTEBRAL ARTERY: A CASE REPORT

¹Alanna Rottler, ^{1,2}Yew Li (Michelle) Dang, ^{1,3}Wai Foong Hooi, ⁴David A Burrows, ^{4,5}Hong Kuan Kok, ^{1,6}Douglas E Crompton. ¹Department of Neurology, Northern Health, Epping, VIC, Australia; ²Department of Neurology, Eastern Health, Box Hill, VIC, Australia; ³Department of Neurology, Austin Health, Heidelberg, VIC, Australia; ⁴Department of Radiology, Northern Health, Epping, VIC, Australia; ⁵School of Medicine, Faculty of Health, Deakin University, Burwood, VIC, Australia; ⁶Department of Medicine, Melbourne University, Northern Health, Epping, VIC, Australia

10.1136/bmjno-2021-ANZAN.70

Background Whilst often causing posterior circulation strokes, vertebral artery dissections may also, more rarely, cause spinal cord infarction.¹ This is the case report of a 39-year-old female with a right-sided high cervical hemi-cord infarction caused by vertebral artery dissection of a hypoplastic right vertebral artery.