disorders of connective tissues. Nasal CSF leakage is extremely rare.¹

Methods and results We present the case of a 40-year-old woman presented to hospital for few days history of postural headache associated with clear intermittent discharge from right nostril without any signs of meningism. There was no history of trauma. She has a background history of Marfan syndrome with associated complications of ASD repair at age 2, mechanical Aortic and Mitral valve replacement, aortic root repair, previous ST elevation MI with LV dysfunction, automated implantable cardioverter-defibrillator in situ, atrial fibrillation, and Hashimoto's thyroiditis. Her regular medications are warfarin, bisoprolol and thyroxine. The clear nasal discharge was positive for β -2 transferrin confirming cerebrospinal fluid. Her CT Brain did not reveal any clear site of CSF leak. She had a flexible nasendoscopy which showed normal nasal passageway, no defect in nasal mucosa and no active CSF leakage. She was managed conservatively with strict bed rest and advised to minimise strenuous activity and heavy lifting.

Conclusion Spontaneous cerebrospinal fluid leak is uncommon condition and frequently associated with hereditary disorders of connective tissues. Nasal CSF leakage is extremely rare.¹ Testing β -2 transferrin has high sensitivity and specificity.² Initial treatment may include bed rest, oral or intravenous hydration, oral caffeine or corticosteroids.^{3–4} If conservative therapy fails, surgical repair with nasal endoscopic approach is recommended.^{2–5}

REFERENCES

1. Ommaya A, Di Chiro G, Baldwin M, Pennybacker J. Non-traumatic cerebrospinal fluid rhinorrhea. *Journal of Neurology, Neurosurgery & Psychiatry* 1968;31(3):214–225.
2. Wang E, Vandergrift W, Schlosser R. Spontaneous CSF Leaks. *Otolaryngologic Clinics of North America* 2011;44(4):845–856.
3. Milledge J, Ades L, Cooper M, Jaumees A, Onikul E. Severe spontaneous intracranial hypotension and Marfan syndrome in an adolescent. *Journal of Paediatrics and Child Health* 2005;41(1–2):68–71.
4. Placantonakis D, Bassani L, Graffeo C, Behrooz N, Tyagi V, Wilson T, et al. Non-invasive diagnosis and management of spontaneous intracranial hypotension in patients with marfan syndrome: Case Report and Review of the Literature. *Surgical Neurology International* 2014;5(1):8.
5. Spontaneous cerebrospinal fluid rhinorrhea. *Medicine* 2018;97(7):e9954.

076

A RARE CASE OF ANTI-GLUTAMATE DECARBOXYLASE ANTIBODY SYNDROME PRESENTING WITH RECURRENT VOMITING

Shoaib Dal*, Bill O'Brien. Central Coast Local Health District, Gosford, NSW, Australia

10.1136/jnnp-2019-anzan.64

Introduction Anti-glutamate decarboxylase antibody (anti-GAD) has been linked with various neurological syndromes including stiff-person syndrome, limbic encephalopathy, cerebellar ataxia, eye movement disorders and epilepsy (collectively known as 'anti-GAD positive neurological syndromes').¹ We describe a very atypical phenotypic presentation of anti-GAD syndrome with unexplained vomiting and weight loss.

Case A 46 years old lady with no past medical or family history of note, presented with 6 months history of severe headaches and recurrent attacks of episodic vomiting (4–6 episodes of multiple vomiting daily) with no identified precipitant and complete normality in between the episodes with no other associated symptoms. She reported 15 kg of unintentional

weight loss. Neurological examination and investigations including MRI brain, CT angiogram and liver enzymes, immunoglobulins, thyroid function, vasculitic screen were normal. Upper GI endoscopy, gastric emptying studies, CT imaging of chest, abdomen and pelvis and whole body PET scan were unremarkable. Serum autoimmune antibody screen was positive with high titre of anti-GAD antibody (1200 kU/liter). The cerebrospinal fluid anti-GAD antibody titre was raised at 103.7 kU/liter with otherwise normal parameters including negative oligoclonal bands. The nerve conduction studies did not show continuous motor activity or spasmodic reflex myoclonus (seen in stiff-person syndrome).² A therapeutic trial of immunosuppression was introduced with moderate improvement in symptoms.

Conclusion Anti-GAD neurological syndromes are rare and this is a unique presentation of the same. It is not completely understood why the presence of one antibody causes varied syndromes. The hypothesis is that the recurrent vomiting is possibly due to diaphragmatic spasms.

REFERENCES

1. Saiz A, Blanco Y, Sabater L, et al. Spectrum of neurological syndromes associated with glutamic acid decarboxylase antibodies: diagnostic clues for this association. *Brain* 2008;131:2553–2563.
2. Buechner S, Florio I, Capone L. Stiff person syndrome: A rare neurological disorder, heterogeneous in clinical presentation and not easy to treat. *Case Rep Neurol Med* 2015; 2015:278065.

077

AN UNUSUAL PRESENTATION OF ANTIBODY NEGATIVE AUTOIMMUNE ENCEPHALITIS: A CASE REPORT

Viral Upadhyay*, Ashish Malkan, Hyunmin Park. Department of Neurology, Nepean Hospital, Sydney, NSW, Australia

10.1136/jnnp-2019-anzan.65

Introduction Immune-mediated encephalitis can occur in the absence of measurable pathogenic antibodies in CSF or serum.

Methods and results We present the case of a 29-year-old Sudanese woman presented with four days history of meningism. She is three months post-partum with her fifth child and does not have significant past medical history. She is a current smoker with history of methamphetamine and heroin use until 5 years ago. There were no focal neurological deficits on examination. CT brain was normal and her CSF had $420 \times 10^6/L$ leucocytes (Polymorphs 18, Mononuclears 402). Three days later, she deteriorated clinically with GCS 11 and developed left sided hemiplegia requiring intubation and ventilation in intensive care.

The CSF culture along with autoimmune encephalitis panel, anti-neuronal antibodies, HSV, VZV, Enterovirus, cryptococcal antigen, TB quantiferon, HIV, Malaria were negative. Her immunology tests were unremarkable including rheumatoid factor, complements, anti-nuclear antibodies, dsDNA, PR3, MPO, RNP, Smith, Ro, La, Scleroderma 70 and Jo-1 antibodies. The MRI Brain showed persistent changes in right hemispheric cortical and subcortical gyriiform FLAIR hyperintensities which were also present in DWI images. The EEG showed right temporo-occipital region 7–8 Hz rhythmic discharges with generalised diffuse slowing. She had right parietal lobe brain biopsy which shows perivascular and intramural inflammation by mononuclear cells almost comprised of CD3 T-cells consistent with encephalitis associated with secondary lymphocytic vasculitis. She was treated with intravenous pulsed methylprednisolone and IVIg

followed by plasma exchange. Then given high dose oral corticosteroids with slow tapering and intravenous Cyclophosphamide four weekly. Significant neurological improvement noted over next 8 weeks with patient being alert and participating in ongoing multidisciplinary rehabilitation for hemiplegia.

Conclusion A case of cerebral biopsy confirmed, CSF and serum antibody-negative encephalitis is presented.

078

SENSORY NERVE ABNORMALITIES IN MOTOR NEURON DISEASE

Allycia MacDonald*, Merrill Needham, Anthony Alvaro. *Department of Neurology, Fiona Stanley Hospital, Murdoch, WA, Australia*

10.1136/jnnp-2019-anzan.66

Introduction Electrodiagnostic evaluation is crucial in establishing the diagnosis of motor neuron disease (MND) and excluding other pathologies. It is recommended that sensory nerve conduction studies (NCS) include the ulnar and sural nerves, and generally accepted that sensory nerves are normal in MND. There are however previous reports in the literature documenting variable sensory abnormalities in patients with MND. We sought to determine the frequency of unexplained sensory abnormalities seen on NCS in patients with MND.

Methods Medical records of patients attending our tertiary MND clinic over a 2 year period were reviewed. We identified 92 patients with a clinical diagnosis of MND for whom electrodiagnostic studies were available to review. Sensory abnormalities in patients without a clear underlying aetiology (eg. compressive neuropathies, diabetes) were considered unexplained.

Results Unexplained sensory abnormalities were detected in at least one nerve in 18/92 (20%) patients. In 17 of those 18 patients, the ulnar sensory response was abnormal. 12 of 18 patients demonstrated abnormalities in 2 or more sensory nerves. Sensory abnormalities were present in 4 of 37 (10.8%) patients with bulbar onset MND and 14 of 55 (25.4%) patients with limb onset MND. Sensory symptoms were infrequently reported and did not correlate with abnormalities found on NCS.

Conclusions Unexplained sensory nerve action potential abnormalities are not uncommon in MND, with ulnar sensory responses the most frequently affected. These findings raise the possibility of sensory nerve pathology in patients with MND and suggest that the presence of unexplained sensory abnormalities should not exclude a diagnosis of MND.

079

GLIAL FIBRILLARY ACIDIC PROTEIN (GFAP) ASTROCYTOPATHY ASSOCIATED WITH CEREBRAL MICRO-INFARCTION AND POOR THERAPEUTIC RESPONSE

¹Allycia MacDonald*, ¹James Triplett, ¹Srimathy Vijayan, ²Michael Bynevelt, ³Rahul Lakshmanan, ¹Thomas Chemmanam. ¹Department of Neurology, Sir Charles Gairdner Hospital, Nedlands, WA, Australia; ²Neurological Intervention and Imaging Service of Western Australia, Sir Charles Gairdner Hospital, Nedlands, WA, Australia; ³Department of Neuroradiology, Perth Children's Hospital, Nedlands, WA, Australia

10.1136/jnnp-2019-anzan.67

Introduction Glial fibrillary acidic protein (GFAP) astrocytopathy is a lesser recognised immune-mediated meningo-encephalomyelitis, which is steroid responsive in the majority of cases. Neuroimaging is unique with a distinctive symmetric white matter perivascular linear and punctate enhancement pattern. We present a case with classical phenotype but delayed clinical response, and highlight the importance of early recognition and treatment.

Case A 59-year-old Caucasian female presented with a two month history of headache, gait disturbance, insomnia, agitation, disorientation and reduced oral intake. Examination revealed a high frequency upper limb tremor, hypertonicity and pathologically brisk reflexes with impaired cognitive function. MRI brain and spinal cord demonstrated high T2 signal and striking perivascular and punctate enhancement in supratentorial white matter, cervical and upper thoracic cord. CSF examination revealed lymphocytic pleocytosis and elevated protein. Brain biopsy demonstrated reduced GFAP expression, perivascular T-lymphocytic infiltrate, and recent white matter microinfarction. CSF and serum GFAP antibodies were positive.

Motor deterioration accompanied progression to a stuporous state. High dose corticosteroids were commenced, followed by intravenous immunoglobulin and mycophenolate. While there was marked improvement of perivascular contrast enhancement on imaging, the patient continued to demonstrate prominent tremor, gait disturbance and behavioural issues 9 months following symptom onset.

Conclusions The persistence of disability in this case is likely the result of axonal loss from the initial insult, reflected by the biopsy evidence of microinfarction. Awareness of the unique pattern on MRI and the clinical phenotype will aid in early recognition and prompt treatment of this condition, thus preventing the potential long term morbidity.

080

THE INCIDENCE, DIAGNOSIS AND OUTCOMES OF IDIOPATHIC INTRACRANIAL HYPERTENSION IN THE SOUTHERN TASMANIAN CATCHMENT

Natasha Krishnadas*, Bruce Taylor. *Neurology, Royal Hobart Hospital, Hobart, TAS, Australia*

10.1136/jnnp-2019-anzan.68

Introduction This study aimed to identify the incidence of idiopathic intracranial hypertension (IIH) in Southern Tasmania, Australia. Secondary aims were to elucidate demographics, current approaches to investigation, treatment and outcomes. To our knowledge, similar regional studies have not been performed.

Methods The study was approved by the University of Tasmania Human Ethics Committee. Patients presented between June 2016-June 2018 to Royal Hobart Hospital, the single tertiary Neurology service in Tasmania. Cases were identified by screening lumbar punctures (LP) performed by Neurology services (inpatient, outpatient, Radiologically-assisted) and by surveying all regional Neurologists. Medical records were used to corroborate LP results and determine whether patients met Modified Dandy Criteria (MDC) (used to define IIH in current literature). Regional population statistics were obtained from the Australian Bureau of Statistics (ABS). Duplicate records were excluded. Exclusion criteria included age <18 at