2589 SUDDEN UNILATERAL AUDIOVESTIBULAR LOSS DUE TO ACUTE LABYRINTHINE HAEMORRHAGE CAN BE MISSED ON MRI BRAIN SEQUENCES

[1]Patrick Harrison*, [2]John Blazak, [3]Joshua Richmond, [4]Kristy Fraser-Kirk, [5]Aliese Hoffman, [6]Grant Collins, [1]Benjamin Tsang. *[1]Neurology Department, Sunshine Coast University Hospital, Sunshine Coast, QLD, Australia; [2]Radiology Department, Sunshine Coast University Hospital, Sunshine Coast, QLD, Australia; [3]Haematology Department, Sunshine Coast University Hospital, Sunshine Coast, QLD, Australia; [4]Ear, Nose, and Throat Department, Sunshine Coast University Hospital, Sunshine Coast, QLD, Australia; [5]Physiotherapy, Sunshine Coast University Hospital, Sunshine Coast, QLD, Australia; [6]Queensland Vestibular Cochlear Clinic, Sunshine Coast, QLD, Australia*

10.1136/bmjno-2023-ANZAN.31

A 45-year-old woman presented with sudden onset vertigo, left hypoacusis, and tinnitus, consistent with acute unilateral audiovestibular loss. Bedside examination revealed a left sided sensorineural hearing loss and leftward falling gait. Left peripheral vestibular hypofunction was confirmed acutely on video head impulse testing characterised by reduced vestibular-ocular reflex gains with overt catch-up saccades in the left horizontal and posterior semicircular canals. Acoustic reflexes were absent on the left side. Pure tone audiometry confirmed a profound left sensorineural hearing loss, with no measurable hearing threshold in the left ear. An MRI brain with diffusion weighted imaging performed after 24 hours was unremarkable. Repeat MRI of the internal acoustic canal (IAC) on day 7 demonstrated increased T1 and FLAIR signal throughout the left cochlear and semicircular canals, without contrast enhancement. A diagnosis of labyrinthine haemorrhage was made. Extensive testing for coagulopathies and vasculitides was negative. On long-term follow-up the patient remained profoundly deaf, however balance and vestibular symptoms improved with early vestibular physical rehabilitation. This case highlights the importance of an inner ear MRI in presentations of acute audiovestibular loss, as well as the utility of ancillary audiovestibular testing. Labyrinthine haemorrhage may be missed on stroke-specific sequences

2637 ELECTROENCEPHALOGRAPHIC CHANGES ASSOCIATED WITH PROPOFOL RELATED INFUSION SYNDROME

[1,2]Ali Buckland*, [3,4]Michael WK Fong, [7]Anna Tierney, [1,2,5]Soumya Ghosh, [1,6]Nicholas Lawn. *[1]Department of Neurology, Sir Charles Gairdner and Osborne Park Health Care Group, Perth, WA, Australia; [2]Perron Institute for Neurological and Translational Science, Perth, WA, Australia; [3]Westmead Comprehensive Epilepsy Unit, University of Sydney, Sydney, NSW, Australia; [4]Department of Neurology, Yale School of Medicine, New Haven, CT, USA; [5]Centre for Neuromuscular and Neurological Disorders, University of Western Australia, Perth, WA, Australia; [6]WA Adult Epilepsy Service, Perth, WA, Australia; [7]Sir Charles Gairdner Hospital, Perth, WA, Australia*

10.1136/bmjno-2023-ANZAN.32

Propofol Related Infusion Syndrome (PRIS) is a life-threatening complication of propofol use. EEG changes associated with PRIS have not been described.

A 21-year-old man with recent traumatic brain injury requiring craniectomy re-presented with recurrent tonic-clonic seizures requiring intubation. After convulsive seizures were terminated, with midazolam, levetiracetam, phenytoin and propofol; continuous EEG monitoring (cEEG) demonstrated focal non-convulsive status epilepticus. Propofol was increased with clonazepam and valproate added. On day 4 the patient developed acute cardiac dysfunction with metabolic acidosis, hyperkalaemia, rhabdomyolysis and hyperlipaemia (all consistent with PRIS). Propofol was ceased. The patient received haemofiltration and aggressive supportive care and survived.

Initial cEEG demonstrated continuous 1-Hz left hemispheric lateralised periodic discharges (LPDs) with associated fast activity (LPDs+F) and recurrent left hemispheric electrographic seizures without clinical accompaniment. The right hemisphere demonstrated changes consistent with an anaesthetic effect. Electrographic seizures improved on Day 3 though LPDs+F persisted. On Day 4, prior to any overt clinical features of PRIS, there was progressive dysfunction initially of the right hemisphere, followed by prolonged periods of generalised suppression followed by burst suppression, with ongoing left LPDs+F. Following propofol cessation these changes persisted for 48 hours and by day 10 the right hemispheric changes had resolved with persistent focal slowing and frequent epileptiform discharges seen on the left.

The EEG changes seen, in the absence of hypotension or hypoxia, likely reflect PRIS-related cerebral mitochrondrial dysfunction. As cEEG is being increasingly utilised recognition of the spectrum of EEG changes of PRIS may allow early identification and immediate cessation of propofol.

2755 LIMBIC-PREDOMINANT AGE-RELATED TDP-43 ENCEPHALOPATHY – A MIMIC FOR AMYOTROPHIC LATERAL SCLEROSIS

[1,2]Jasmine F Ashhurst*, [1]Srestha Mazumder, [1,3]Matthew C Kiernan. *[1]Forefront Clinic, Brain and Mind Centre, Camperdown, NSW, Australia; [2]Department of Neurology, St George Hospital, Kogarah, NSW, Australia; [3]Department of Neurology, Royal Prince Alfred Hospital, Camperdown, NSW, Australia*

10.1136/bmjno-2023-ANZAN.33

Background Limbic-predominant age-related TDP-43 encephalopathy (LATE) is a TDP-43 proteinopathy, typically diagnosed in elderly patients. At present, a definitive diagnosis of LATE is made from neuropathological changes at autopsy. Neuropathological changes are characterised by the anatomical distribution of TDP-43 in the amygdala, hippocampus and middle frontal gyrus. Separately, LATE is most commonly associated with cognitive impairment, typically dementia of the Alzheimer's type.

In terms of causation, TDP-43 is a protein encoded by the TARDBP gene. Of relevance to motor function, mutations involving the gene encoding TARDBP are causative for amyotrophic lateral sclerosis (ALS) in <5% of familial cases of ALS. Separately, TDP-43 protein is the hallmark ALS pathology, present at autopsy in >90% of patients diagnosed with ALS.

Cases The present case series describes two patients managed for ALS, who were subsequently diagnosed with LATE following post-mortem examination.

Case 1 (76M) initially presented with upper limb wasting and predominantly lower motor neurone pattern weakness, progressing to generalised disease with weakness over a 12 year period. Case 2 (79F) presented with disease-onset in the distal left upper limb, progressing to a left sided spastic hemiparesis and spastic dysarthria. She was diagnosed with PLS (likely Mills' variant) and died 7 years after symptom onset.

Discussion Unifying phenotypic features of an ALS-presentation of LATE include limb-onset disease, slower disease progression

compared to typical ALS, without prominent cognitive features. Recognition of LATE and its mimics is of increasing relevance, particularly in an ageing population, with potential interventions directed against TDP43.

2764 A CASE OF SEMANTICS: HUNTINGTON DISEASE PRESENTING AS PRIMARY PROGRESSIVE APHASIA

[1,2]Antonia J Clarke*, [2]David Manser*, [3]Ronald Fleischer, [1,4,5]Michael J Fulham, [1,2]Rebekah M Ahmed. *[1]Faculty of Medicine and Health, University of Sydney, Camperdown, NSW, Australia; [2]Neurology, Royal Prince Alfred Hospital, Camperdown, NSW, Australia; [3]Genetics, Royal Prince Alfred Hospital, Camperdown, NSW, Australia; [4]Molecular Imaging, Royal Prince Alfred Hospital, Camperdown, NSW, Australia; [5]Engineering and Computer Science, University of Sydney, Camperdown, NSW, Australia*

10.1136/bmjno-2023-ANZAN.34

Objective We present a case of the semantic variant of primary progressive aphasia (PPA) as the initial presenting feature in a patient with Huntington disease (HD).

Case A 76-year-old Caucasian man was referred for evaluation of progressive language impairment, including impaired naming, object knowledge and single word comprehension consistent with the semantic variant of PPA. He subsequently developed behavioural changes and a hyperkinetic movement disorder, characterised by frank choreiform movements of the upper limbs, trunk, and lower limbs with oromandibular dyskinesia and motor impersistence of tongue protrusion. Magnetic resonance imaging (MRI) of the brain showed left anterior temporal lobe and hippocampal atrophy. A neurological FDG PET-CT showed reduced metabolism in the head of the left caudate nucleus. *HTT* gene testing revealed an expansion of 39 CAG repeats in one allele.

Conclusion The case outlines the substantial overlap between the clinical presentation of HD and frontotemporal lobar degeneration (FTLD) syndromes. Genetic testing in this case supported the diagnosis of atypical HD, presenting with semantic variant PPA.

2823 MADD MUSCLES: A CASE REPORT OF VERY-LATE ONSET MULTIPLE ACYL-COA DEHYDROGENASE DEFICIENCY (MADD)

[1,2]Matthew Linger*, [3]Catriona McLean, [1]Edrich Rodrigues, [1]Belinda Cruse*. *[1]Neuromuscular Service, Neurology Department, The Royal Melbourne Hospital, Parkville, VIC, Australia; [2]Neurology Department, Grampians Health, Ballarat Hospital, Ballarat, VIC, Australia; [3]Victorian Neuromuscular Laboratory Service, Anatomical Pathology, Alfred Health, Melbourne, VIC Australia*

10.1136/bmjno-2023-ANZAN.35

Introduction We present a case of a 79-year-old female with very-late onset Multiple Acyl-CoA Dehydrogenase Deficiency (MADD).

Case The patient presented with progressive proximal weakness and difficulty walking over twelve months. Past medical history was significant for a multi-nodular goitre on carbimazole 5mg daily, hypercholesterolaemia on simvastatin 10mg daily, and non-melanomatous early-stage skin cancers (all fully excised). Examination revealed moderate-severe proximal limb and axial weakness. She was unable to independently transfer or walk. Creatinine kinase levels were normal. Nerve conduction studies were normal and needle electromyography of weak muscles showed reduced recruitment of long-duration polyphasic motor units with no spontaneous activity. FDG-PET scan demonstrated intense, symmetrical uptake within multiple proximal muscles and reduced cardiac uptake. On a left biceps muscle biopsy, a lipid myopathy was diagnosed with moderate accumulation of lipid in type 1 fibres raising the possibility of MADD on morphologic grounds. Metabolic profile testing revealed low serum carnitine, elevated concentrations all length acyl-carnitines and elevated urine organic acids consistent with a diagnosis of MADD. There was significant functional improvement within 3 months of oral Riboflavin and Coenzyme Q10 administration; the patient returned to walking independently with a four wheel frame.

Conclusion This case highlights a rare, but effectively treatable diagnosis of a lipid storage myopathy suspected to be very-late onset MADD.

2786 CASE REPORT: SNAP, CRACKLE, AND POP! UNUSUAL SYMPTOMS OF MOTOR DYSFUNCTION OF THE TRIGEMINAL NERVE PRECEDING TRIGEMINAL NEURALGIA

Daniel Guilfoyle*, Kaitlyn Parratt. *Neurology, Royal Prince Alfred Hospital, Camperdown, NSW, Australia*

10.1136/bmjno-2023-ANZAN.36

Case A 53-year-old woman presented with paroxysmal ear-popping followed by paroxysmal facial pain. Her background included hypertension, prior smoking, hysterectomy, and menopausal symptoms on oestrogen replacement.

She initially noticed intermittent right ear-popping & crackling, triggered by turning her head whilst supine, and increasing in frequency over days. Attacks subsequently began occurring when upright also. This was followed a month later by paroxysmal severe, shooting pain in the right mandibular & maxillary regions, associated with pallor, and lasting seconds to minutes. There were no other neurological or systemic symptoms, nor headache or sensory symptoms between attacks. Her cranial nerve examination was unremarkable including trigeminal motor/sensory function, hearing, and otoscopy. An MRI brain with gadolinium contrast was normal with no structural pathology of the right trigeminal nerve or brainstem found.

She was treated with carbamazepine for suspected trigeminal neuralgia. This led to a complete resolution of both paroxysmal pain and ear-popping, with no reported recurrence after 3 months. Further clinical and radiological follow-up is planned.

Conclusion Trigeminal neuralgia manifests painful sensory paroxysms, though motor symptoms, such as hemi-masticatory spasm,[1] are rarely described. Via innervation of tensor veli palatini (TVP), the trigeminal nerve is required for normal Eustachian tube function. Dysfunction of the trigeminal motor branch causing intermittent TVP contraction was theorised to cause the ear-popping experienced by our patient. The resolution with carbamazepine supported this explanation. When faced with noncanonical but focal neurological symptoms,