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

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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)

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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

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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,