there was severe weakness of right elbow, wrist and finger extension with an anaesthetic patch over the anatomical snuff box. Ultrasonography showed fascicular oedema of the radial nerve in the upper arm. Nerve conduction studies confirmed an acute axonotmetic radial neuropathy at the spiral groove. The patient was referred for hand therapy and at 4 months regained most of the function in her hand, with some mild persistent sensory impairment.

Conclusions Cryolipolysis is a cosmetic treatment that aims to locally reduce subcutaneous fat. The procedure is performed using a vacuum applicator to cool the selected area to temperatures as low as -9 degrees Celsius. Peripheral neuropathies following the procedure have been rarely described¹ but, to our knowledge, this is the first report of an acute neuropathy developing during the procedure. The causative mechanisms of cryolipolysis-induced nerve injury in this case were likely due to nerve compression related to local oedema and thermal effect on the radial nerve.

REFERENCE

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093 CJD AND MOTOR NEURON DISEASE: A GROWING ASSOCIATION

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Introduction Amyotrophy in Creutzfeldt-Jakob Disease (CJD) is rarely a conspicuous clinical finding. The overlap of CJD (a prionopathy) and motor neuron disease is reported in the literature but it remains to be established whether the neuronopathy is an integral part of CJD presentation, or, whether this represents a distinct variety. Our case describes a male patient with clincopathological diagnosis of sporadic CJD along with evidence of motor neuronopathy on nerve conduction studies.

Case Summary At presentation to our neurology service, the patient was a 72-year-old male, living at home with his wife. He was initially referred for progressive short-term memory loss, personality change, and gait disturbance. On review, it was noted that in addition to gait and limb ataxia, and cognitive impairment, he demonstrated prominent generalised fasciculation. Nerve conduction and electromyography studies showed normal nerve conduction but fasciculations in proximal and distal muscle groups of the left upper and lower limbs, in keeping with a motor neuronopathy. CSF 14-3-3 and EEG provided little bearing. MRI demonstrated progressive T2-hyperintense, diffusionrestricted lesions in the bilateral basal ganglia, thalami and medial frontal cortices consistent with CJD. Post-mortem examination demonstrated spongiform encephalopathy and immunohistological staining (12F10) in-keeping with diagnosis of CJD.

Conclusion In our patient, the combination of clinical and neurophysiologic features of motor neuron disease and a confirmed diagnosis of Creutzfeldt-Jakob Disease raises the vexed question of whether this represents a distinct overlap syndrome or an infrequent manifestation of the same pathology. Further research is required to establish this.

094 NOVEL NOTCH1 VARIANT IN A PATIENT WITH SPONTANEOUS INTERNAL CAROTID ARTERY DISSECTION

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Introduction We report a case of spontaneous left internal carotid artery (ICA) dissection associated with a novel *NOTCH1* variant.

Case A 43-year-old lady presented with a 3-day history of severe headache and transient expressive dysphasia. There was no history of preceding trauma. CT brain (CTB) and carotid angiography (CTA) demonstrated small areas of established subcortical infarction with occlusion of the M1-segment of the left middle cerebral artery (LMCA). CT cerebral perfusion (CTP) displayed a region of increased mean transit time and cerebral blood volume consistent with a large ischaemic penumbra.

Digital subtraction angiography confirmed an occlusion of the LMCA with luminal irregularity of the supraclinoid ICA suggestive of arterial dissection. An intracranial stent was deployed from the superior M2-division of the LMCA to the cavernous ICA. Progress CTA and CTP demonstrated reperfusion of the LMCA territory.

Six-months later, she remains well with no recurrence of symptoms or detectable neurological signs. Targeted gene panel demonstrated a novel heterozygous missense variant (c.56C>T;p.Ala19Val) in exon-1 of the NOTCH1 gene. Segregation testing demonstrated an identical variant in her mother. Conclusion Spontaneous intracranial ICA dissection is a rare condition described mostly in single case reports. Mortality rates have been reported of up to 75%. The NOTCH-signal-ling pathway is involved in the embryonic development of arterial endothelium. NOTCH1 variants have been associated with autosomal dominant bicuspid aortic valve aortopathy, and rarely in extracranial arterial dissection. To our knowledge; this is the first reported case of intracranial dissection where a previously undescribed NOTCH1 variant is identified.

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'NO END IN SIGHT', MANAGEMENT DILEMMA OF REFRACTORY MOG ANTIBODY POSITIVE OPTIC NEURITIS

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Background The patient is a 38-year-old lady who presented with impaired visual acuity in her right eye which was accompanied with pain on extra-ocular movements. Her symptoms initially resolved with high dose steroid therapy. This is on a background of eosinophilic asthma which is refractory to maximal inhaler therapy and IL-5 monoclonal antibody therapy.

Methods/Results The patient had unremarkable blood results and inflammatory makers and a normal CSF study. She was subsequently found to be myelin oligodendrocyte glycoprotein