2227 SPORADIC LATE-ONSET NEMALINE ROD MYOPATHY ASSOCIATED WITH MONOCLONAL GAMMOPATHY

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A 72-year-old man developed progressive camptocormia associated with head drop, dysphagia, proximal muscle weakness, numbness to the ankles, and 20kg weight loss over a 2 year period. Background history was significant for thyroid cancer treated with total thyroidectomy resulting in left accessory nerve injury, type 2 diabetes mellitus, hypertension, and gout. Examination revealed a man with reduced muscle bulk who walked with a flexed spine. He had weakness of neck flexion and proximal muscles with normal reflexes and reduced pinprick sensation to both feet. Nerve conduction studies showed length-dependent axonal sensorimotor neuropathy. Concentric needle electromyography showed widespread reduced recruitment of small polyphasic units consistent with a myopathic process. Creatine kinase level was normal. Myositis panel demonstrated low positive Mi2 of uncertain significance. An initial deltoid muscle biopsy did not show evidence of myopathy. Due to worsening of symptoms, a repeat muscle biopsy of vastus lateralis was performed, revealing atrophic myofibres with a patchy granular appearance. Electron microscopy identified nemaline rods. An IgG kappa paraprotein was identified. HIV serology was negative. A diagnosis of sporadic late-onset nemaline myopathy (SLONM) associated with monoclonal gammopathy was made. He was commenced on IVIG and referred for haematological evaluation. SLONM is a rare acquired adult-onset progressive myopathy associated with monoclonal gammopathy and HIV infection. It may present with head drop or dysphagia and is characterised electrophysiologically by a combination of axonal neuropathy and proximal myopathy. First-line treatment with intravenous immunoglobulin and second-line treatment with haematological/immunological therapies may result in significant improvement.

2228 CT PERFUSION IMAGING IN NON-CONVULSIVE STATUS EPILEPTICUS

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A 75-year-old previously well right-handed woman presented as a code stroke within 2 hours of sudden onset global aphasia. Examination revealed GCS 10 (E4, V1, M5), transient rightward gaze deviation, otherwise normal tracking eye movements, spontaneous movement of all limbs, and non-brisk reflexes with flexor plantar responses. CT stroke imaging revealed no established infarction or large vessel occlusion. CT perfusion (CTP) showed marked global hyperperfusion of bilateral cerebral hemispheres and subcortical structures. Non-convulsive status epilepticus (NCSE) was suspected and the patient was commenced on anti-epileptic and empirical antimicrobial therapy. Electroencephalography revealed left frontotemporal rhythmic delta activity with frequent ictal spikes consistent with focal NCSE. Initial CSF analysis was acellular with mildly raised protein 0.56g/L. Infectious, autoimmune, limbic, paraneoplastic and metabolic panels were negative in serum and CSF. Brain magnetic resonance imaging revealed T2 hyperintensity of the left hippocampus in keeping with ictal/post-ictal change without underlying structural abnormality. MR perfusion confirmed diffuse increase in regional cerebral blood volume of bilateral cortical and subcortical grey matter. The patient's language returned after 72 hours of seizure management in the intensive care unit. Progress CTP revealed resolution of perfusion abnormalities. Progress cerebrospinal analysis revealed mild monocytic pleiocytosis (8x10^6/L) with positive Herpes Simplex Virus 2 PCR. The patient completed 14 days of IV aciclovir. On discharge, the patient had new mild cognitive deficits but no further seizures. This case highlights the utility of CTP in the identification of stroke mimics such as NCSE, leading to prompt treatment and avoiding unnecessary and potentially harmful treatments.

2231 PARANEOPLASTIC GRANULOMATOUS ANGIITIS OF THE CENTRAL NERVOUS SYSTEM

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A 44-year-old right-handed male smoker presented with a 1 month history of bilateral throbbing headaches. Associated symptoms included mental fogginess, daytime hypersomnolence, 20kg weight loss, and subjective fevers and chills. Examination revealed no focal neurological deficits apart from sustained right-beating gaze-evoked nystagmus. He had neuropsychological deficits in executive functioning, working memory and verbal and non-verbal cognition. Routine blood tests and inflammatory markers were normal. CSF analysis revealed significantly raised protein (2.68g/L), low glucose (1.9mmol/L) and monocytic pleiocytosis (80x10^6/L). CSF flow cytometry and cytology showed no abnormal cells. Oligoclonal bands were not detected. Serum and CSF autoimmune/vasculitis, infectious, metabolic and limbic encephalitis panels were negative. Brain MRI revealed extensive signal change in bilateral cerebral white matter with punctate foci of perivascular enhancement, vascular congestion, and mild leptomeningeal enhancement. Brain PET was normal. Body PET revealed multiple glucose avid lymph nodes in right axillary and supra/infraclavicular lymph nodes. Lymph node biopsy showed Nodular Lymphocyte-Predominant Hodgkin's Lymphoma (HL). Brain biopsy revealed non-necrotising granulomatous vasculitis with reactive parenchymal change, consistent with a diagnosis of granulomatous angiitis of the central nervous system (GACNS). The patient was treated with high-dose pulse steroids and commenced R-CHOP chemotherapy. At 3 month follow-up (post-cycle 2 chemotherapy), he had significant improvement of neurological symptoms with metabolic remission of HL. GACNS is a rare paraneoplastic vasculitis associated with HL. Clinical and radiological features are diverse,