their sedating adverse effects waned. The patient started to spontaneously open his eyes; improved awareness was noted, in form of slow and irregular voluntary gaze deviation towards the examiner and grimacing and vocalizations on cutaneous stimulation. After ten months of KD, the clinical amelioration remained stable. Longitudinal brain magnetic resonance imaging showed gradual reduction of areas of diffusion restriction and appearance of diffuse atrophy. No significant adverse effects were noted.

Discussion: KD improved the control of hyperkinetic movement disorders in our case of CJD. The efficacy of KD in controlling myoclonus and other hyperkinetic movement disorders is well described in glucose transporter 1 deficiency syndrome [2]. We previously examined the efficacy of KD in the management of myoclonus and other symptoms of subacute sclerosing panencephalitis [3]. However, its efficacy in other hyperkinetic movement disorders remains to be tested. The observed improvement allowed a reduction in pharmacologic load, that was accompanied by a sustained improvement in consciousness. Overall, the clinical course appeared to be milder than usual; further studies on KD in CJD are warranted to examine the complete spectrum of effects.

Conclusion: KD may represent a complementary treatment for CJD-associated hyperkinetic movement disorders such as dystonia and myoclonus, with an excellent safety profile.

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PARAINFECTIVE AND POST-VACCINE SARS-COV-2-RELATED MYELITIS: A CASE-BASED COMPARISON

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Introduction and objectives: Myelitis is a known parainfective complication of Covid-19 infection and can follow SARS-CoV-2 vaccination. Here, we present two of such cases comparing clinical, laboratory and radiological features.

Case 1: A 40-year-old male attended our Emergency Department (ED) with Covid-19 (day-9) infection with new onset urinary retention lower limb sensory-motor weakness and a T10 sensory level. Spinal MRI confirmed subacute thoracic myelitis (T3-T8) with heterogeneous contrast uptake for which high-dose steroids were started. Cerebrospinal fluid (CSF) on day 2 showed 16 leucocytes/microL (mostly polymorphonuclear). PCR for SARS-CoV-2 was still positive on serum but negative on CSF. Clinical deterioration continued to severe upper limbs weakness, paraplegia, a T3 sensory level and a weak cough reflex despite a 5-day course of immunoglobulins. Considering the progressive course and a negative COVID swab (day 5), 6 plasma exchange sessions were arranged. On day 27, CSF showed 1 leucocyte/microL with intrathecal IgG synthesis and no blood brain barrier (BBB) dysfunction and a repeat MRI confirmed progression of myelitis with involvement of cervical segments without contrast enhancement. Eventually, clinical stability was reached after infusion of cyclophosphamide on day 31.

Case 2: An 83-year-old man presented to the ED with new-onset left leg sensory-motor deficits associated with haematuria whilst on oral anticoagulants for atrial fibrillation (AF). He had received the 3rd vaccination against COVID-19 (mRNA-based) 3 days prior. Following initial discharge, he experienced urinary retention and progressive ascending weakness. Neurological examination after 3 weeks showed paraplegia, lower limbs areflexia, an extensor plantar response on the left, anesthesia below T5 and upper limbs weakness. MRI of the spine without contrast revealed altered signal in the cord from the bulbar to the thoracic levels and around the conus medullaris, compatible with diffuse myelitis. CSF showed a mild protein increase (67 mg/dL), type 2 oligoclonal bands and BBB dysfunction. A 5-day course of high dose IV methylprednisolone (1g) followed by a tapering regimen led to a good clinical response. Spinal MRI with contrast after 20 days confirmed improvements with intramedullary enhancement suggesting persistent barrier damage. The patient was eventually discharged after 32 days with residual paraparesis. Discussion and conclusions: Our patient with parainfective myelitis showed no evidence of BBB dysfunction, little contrast uptake on MRI and a more severe clinical course refractory to several therapies. Conversely, the post-vaccination case exhibited a more inflammatory and benign phenotype suggesting specific disease-related mechanisms and pathogenesis could account for these differences.

ATYPICAL POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME WITH HEMISPHERIC, BRAINSTEM AND SPINAL CORD INVOLVEMENT IN A PATIENT WITH ACUTE KIDNEY INJURY – A CASE REPORT

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Objectives: Posterior reversible encephalopathy syndrome (PRES) is a disorder characterized by variable clinical presentation and MRI features consisting of vasogenic oedema in parieto-occipital regions [1]. The description of cases of atypical PRES has led to reconsidering such syndrome as a complex and multifaced clinical entity [2]. Herein, we provide clinical and radiological description of a patient affected by atypical PRES.

Materials: We describe a case of a 42-year-old man, smoker, with negative clinical history, except for sporadic headaches treated with NSAIDs. Due to an acute episode of headache, vertigo, and subsequent loss of consciousness, he presented to the emergency department where high blood pressure values where reported (240/110 mmhg). He was then admitted to our neurology ward.

Methods: The patient was alert and keenly responsive, with mild deficit of left abducens and facial nerves and a remarkable tendency to retropulsion during upright stance. Laboratory tests showed renal failure (Creatinine 3.8 mg/dL, Azotemia 101 mg/dL, Na 131 meq/l, K 2.8 meq/l) and low platelet count (110.000 mm3). Brain MRI showed remarkable hyperintensity in the T2/FLAIR sequences, likely corresponding to vasogenic edema, bilaterally involving centrum semiovale, periventricular white matter, internal and external capsules, thalami, optic chiasm, both vermis and cerebellar hemispheres, the whole brainstem and the spinal cord until C6 level. Hypointensities located in the basal ganglia and cerebellum were observed on susceptibility-weighted imaging, likely corresponding to microhemorrhages.

Results: The patient was treated with intravenous nitroglycerin infusion until blood pressure normalization. He underwent haemodyalitic treatment due to worsening of his kidney failure. Serologic level of ADAMTS-13 was tested negative. Abdomen ultrasonography showed symmetric, volumetric reduction of both kidneys. The brain MRI, executed at the normalization of kidney laboratory tests and pressure levels showed remarkable reduction of the T2-FLAIR hyperintensities with total resolution in the spinal cord together with millimetric areas of diffusion

restriction likely corresponding to cytotoxic oedema. After the placement of an artero-venous fistula the patient was discharged home in good clinical conditions.

Discussion: the patient was diagnosed with atypical PRES due to hypertension in the setting of acute kidney injury and improved both clinically and neuroradiologically after the removal of the offending cause.

Conclusions: While PRES is promptly recognized due to its typical imaging features, atypical and complicated PRES are uncommon and may constitute a diagnostic dilemma, especially when clinical history and examination are poorly suggestive [3]. Patients with atypical PRES, characterized by focal hemorrhages and diffusion restriction may have a poorer prognosis and should be readily recognized.

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DO NOT FORGET CLINICAL EXAMINATION! A CASE OF OPMD WITH NEUROGENIC PATTERN AT EMG MISDIAGNOSED FOR MND

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Background: Oculopharyngeal Muscular Dystrophy (OPMD) is a late-onset myopathy caused by the abnormal expansion of GCN repeats in exon 1 of the poly (A) binding protein, nuclear 1 (PABPN1) gene. As per other poliglutamine-expansions-related diseases, the length of GNC repeats influences the disease severity, the age of disease onset and the involvement of respiratory muscles. The main symptoms at disease onset are ptosis and ocular motor abnormalities, followed by the occurrence of dysphagia and lower proximal limb weakness, with only slightly increased creatine kinase levels. Electromyogram usually shows myopatic pattern, though a coexistence with peripheral neuropathy and the presence of neurogenic pattern at electromyogram are often reported. In the latter case, mainly when symptoms are incomplete or isolated, differential diagnosis with motor neuron diseases is more difficult. Here we describe a case of a patient with dysphagia and neurogenic pattern at electromyogram addressed to our Centre with the suspicion of a motor neuron disease.

Case Reports: The patient was a 70 years old female patient who reported difficult swallowing started a few years prior and progressively worsened in time. She also noticed the occurrence of cramps and a progressive hyposthenia in lower limbs that led to gait difficulties. Her mother suffered from a similar form of dysphagia and lower limbs hyposthenia as well, never better investigated. In time, she underwent several exams, including a fiber-optic endoscopic evaluation of swallowing showing a slowness in tongue mobility and an increased amount of food aspiration, a cerebral and spine MRI resulted normal, blood test with creatine kinase dosage resulted slightly increased (196U/l). An electromyogram showed a diffuse neurogenic pattern with no acute denervation. In the suspicion of a motor neuron disease, she was addressed to our Centre for Motor Neuron Diseases in Santa Chiara Hospital, Pisa, Italy. The neurological examination revealed nasal voice, an hyposthenia in orbicularis oris, slight hyposthenia in corrugator, hyposthenia in iliopsoas, no signs of upper motor neurons involvement excluding jerk and glabellar positivity. Interestingly, she did not present ptosis.

Considering the clinical picture and the family history, we performed a DNA test for oculopharyngeal muscular dystrophy. The exam showed heterozygous GNC triplets expansion, pointing to an autosomal dominant form of oculopharyngeal muscular dystrophy.

Discussion and conclusion: Our case underline the importance clinical evaluation, that remain the pivotal element in patient evaluation. When coupled with anamnesis, clinical evaluation can even outperform instrumental exams.

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WHY ALZHEIMER PROGRESSES SO SLOWLY IN SOME PATIENTS?

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Background: Alzheimer's disease (AD) progression is heterogeneous for various reasons including resilience, cognitive reserve and genetic characteristics, still partially understood. AD is often complicated by cerebral amyloid angiopathy (CAA) that adds another variance source. We describe the astonishing case of a man who became aware of his cognitive deficit 18 years ago and who is still in its mild cognitive impairment (MCI) stage.

Case report: An 82-year-old man with 13 education years and dementia familiarity came to first visit in 2004 reporting anomia and memory problems. ApoE genetic test was ε3/ε4, and all investigations, including brain perfusion SPECT, were normal. In 2012 brain MRI showed left hippocampal atrophy and FDG-PET highlighted moderate hypometabolism in left posterior cingulate and precuneus. CSF biomarkers (December 2012) and amyloid PET (December 2013) were consistent for AD. EEG was normal in 2004 but showed moderate theta activity in anterior regions. Neuropsychological tests were normal until 2019, when mild impairment of verbal memory was highlighted. In 2020 performance decreased in attention tests, selection and attentional shifting tasks and visuospatial research tasks (TMTA, TMTB, Stroop). The Grober-Buschke test was impaired in immediate free recall, delayed recall and recognition with poor cueing effect while logical memory test was borderline. Serial brain brain MRIs between September 2020 and May 2022 showed 4 new asymptomatic microbleeds and superficial siderosis in the left parietal area, standing for probable CAA. His last MMSE score (April 2022) was 26/30.

Discussion: SCI should not be underestimated, since can represent a prodromal disease phase that we are still unable to detect with currently available neuropsychological tests. Patients with SCI at higher risk to develop neurodegenerative diseases include those with subjective decrement in memory irrespective of function in other cognitive domains, onset of SCI within the past 5 years or at 60 years and older, concern associated with SCI, persistence of SCI over time, those seek of medical help and those show cognitive decline confirmed by an observer.

Conclusions: SCI with normal assessments requires adequate investigations and follow up to identify the underlying potential cause and