EP.17

The impact of HTLV-I infection on clinical features of inclusion body myositis

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Inclusion body myositis (IBM) is a slowly progressive myopathy with unique clinical and pathological features. So far, there are several case reports of patients with IBM and HTLV-I infection. However, there is no study that investigated clinical features of IBM associated with HTLV-I infection. We investigated the clinical differences between the IBM patients with and without anti-HTLV-I antibodies. In 402 patients enrolled into the study, 250 patients fulfilled the ENMC2011 criteria for diagnoses of IBM. Among them, 12, 171, and 67 patients were positive, negative, and unexamined for anti-HTLV-I antibodies, respectively. The patients with the antibodies significantly started with difficulty in ambulation, and tended to exhibit a male predominance, preserved respiratory function, and effectiveness of steroid therapy. The IBM patients with anti-HTLV-I antibodies showed similar clinical features as those without the antibodies, except a frequent onset of difficulty in ambulation, at the instant of diagnoses. However, longitudinal analyses would be necessary to understand the effect of HTLV-I infection on the clinical course of IBM.

http://dx.doi.org/10.1016/j.nmd.2021.07.042

EP.18

Oculopharyngeal Muscular dystrophy patients with treatment-responsive, statin-associated autoimmune necrotic myopathy

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Our objective is to describe two genetically confirmed Oculopharyngeal Muscular Dystrophy (OPMD) patients with HMGCoARdase-antibody positive autoimmune necrotic myopathy (AINM) responsive to immunosuppressive treatment. Although clinicians have suspected muscular dystrophy patients are at increased risk of statin side-effects, to date no patients with genetically confirmed muscular dystrophy have been reported with HMGCoRdase-antibody positive AINM. Here we describe the clinical presentation, serology, muscle pathology and response to treatment in two patients with genetically confirmed OPMD who developed statin-associated AINM. A 46-year-old male with previously known OPMD and only mild ptosis presented 2 years after discontinuing atorvastatin with severe proximal weakness and CK up to 10000 U/L. He became wheelchair-bound and developed progressive orthopnea with BiPAP dependence. Muscle biopsy demonstrated rimmed vacuoles, necrotic and regenerating fibers and endomysial inflammatory cells (CD8+ > CD4+). MHC was upregulated in scattered non-necrotic muscle fibers. The second patient was a 71-year-old male who presented (while on statin) with a one-year history of proximal arm and leg weakness, myalgias, and gait deterioration. In addition, he had longstanding ptosis and dysphagia. OPMD was confirmed with genetic testing. CK was measured at 5880 U/L. Both patients had positive HMGCoRdase antibodies. Both patients were treated with steroids and IVIG (patient 2 was additionally treated with methotrexate). Both patients had dramatic improvements in their strength and CKs normalized. Clinicians should be vigilant for the co-occurrence of statin-associated AINM in patients with OPMD or other dystrophies. If statins are recommended, CK and symptoms should be closely monitored and any abrupt increase in CK and weakness should prompt discontinuation of the statin, HMGCoRdase antibody testing and muscle biopsy. Muscle necrosis seen in muscular dystrophy patients may predispose them to statin-associated AINM due to necrosis-induced liberation of the intracellular HMGCoARdase protein thus permitting antibody formation.

http://dx.doi.org/10.1016/j.nmd.2021.07.043

COVID-19 AND NEUROMUSCULAR DISEASES

EP.19

Facial Onset Acute Inflammatory Demyelinating Polyneuropathy Related to SARS-CoV-2

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Guillain–Barré is a disorder of the peripheral nervous system characterized by acute onset ascendance paresis. We present a case diagnosed with facial onset acute inflammatory demyelinating polyneuropathy after being infected with SARS-CoV-2. A 51-year-old man presented with facial diplegia to the emergency room. Then he developed bilateral ascending paralysis. He noticed that for one month he had smell and taste disturbances. SARS-CoV-2 infection was suspected. Nasopharyngeal swab polymerase chain reaction test was negative but anti-SARS-CoV-2 antibody found to be positive. Nerve conduction study showed prolonged motor distal and F wave latencies with decreased motor and sensory CMAP amplitudes. Lumbar puncture revealed albumino-cytologic dissociation. According to neurological examination and laboratory findings, the case diagnosed as acute inflammatory demyelinating polyneuropathy. Axonal excitability study revealed fanning in pattern with prolonged refractoriness which indicates nodal sodium channel disturbances. Even if facial onset SARS-CoV-2 related Guillain–Barré cases have been rarely reported, facial involvement seems to be one of the features of the neurological findings.

http://dx.doi.org/10.1016/j.nmd.2021.07.044

EP.20

The change in weight gain during the coronavirus pandemic in children with Duchenne muscular dystrophy

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Assessment of the change in the trend of weight gain in children affected by DMD during the first twelve months of the coronavirus pandemic in one paediatric neuromuscular centre. We noted that a number of children affected by Duchenne muscular dystrophy appeared to have had significantly accelerated weight gain during the first year of the pandemic; potentially greater than weight gain in previous years and that expected for age. To evaluate this, the body mass index BMI change and centiles during this year will be compared to that in the two years preceding the pandemic for each patient. In addition, the BMI of those children seen in clinic will be compared to patients same age and ambulatory status seen in the six months prior to March 2020 when national lockdown started due to the pandemic. The findings of those two comparisons of weight gain in ambulant and non-ambulant patients with DMD will help in planning the service for this group of patients in the coming period.

http://dx.doi.org/10.1016/j.nmd.2021.07.045

EP.21

COVID-19 pandemia: Physical, psychological and social impact of lockdown on neuromuscular patients.

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The unexpected Coronavirus disease 2019 (Covid 19) worldwide crisis induced a lockdown of the whole population in Belgium from 18th March