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Chronic Disease Course and IV Ig-Dependence in Long-term Follow-up of Anti-HMGCR Immune-Mediated Necrotizing Myopathy.

Objectives: Anti-HMGCR antibodies have been associated with a severe form of immune-mediated necrotizing myopathy (IMNM) with a poor muscle strength recovery and early muscle damage. These patients tend to require aggressive immunosuppressive therapy and present relapsing disease course. Our objective was to evaluate real-life treatment strategies in anti-HMGCR IMNM patients.
Methods: This single centre study included all patients with anti-HMGCR IMNM with at least 12 months of follow-up. Medical records were retrospectively reviewed to assess clinical features at diagnosis, HLA typing, treatment strategies over the follow-up period (including corticosteroid (CS) use, number of immunosuppressive (IS) agent and intravenous immunoglobulin (IV Ig) duration), disease course, and clinical status and therapeutic profile at last follow-up. Remission was defined as the presence of CK level ≤ 2 times the upper limit of normal associated with a stable manual muscle testing for ≥3 months. Quantitative variables are reported as median [IQR1-IQR3].
Results: Thirty-five patients were included. Age at diagnosis was 47.1 [26.1-60.2] years, 74% of patients were female, 29% were statin-exposed, all patients presented with muscle weakness (deltoid and psoas MRC-5 was 4.0 [2-4] and 4.0 [3-4], respectively) and highest CK level was 8146 [5000-12090] IU/L. Time from symptoms onset to treatment initiation was 0.8 [0.3-4.7] years. During the follow-up period, 91% of patients were treated with CS in combination with an IS agent, the majority of patients received IV Ig (91%) and the number of treatment intensification was 2 [1-4]. 40% of patients also received plasma exchanges as part of induction therapy.
All patients demonstrated a chronic disease course and no patients were in treatment-free remission at last follow-up after 4.9 [3.1-8.9] years. At last follow-up, 60% of patients were in remission - most of which with IV Ig (71%) -, 57% of patients were still receiving CS and CS dose was 8 [5-10] mg/day and 54% of patients had an IS agent. At last follow-up, only 40% of patients had a normal muscle strength, deltoid and psoas MRC-5 was 5 [4-5] and 4 [3-5], respectively, and CK level was 299 [200-559] IU/L. No predictors of remission or IV Ig use at last follow-up were identified, including demographic features, HLA-DRB1*11:01 and HLA-DRB1*07:01 status, muscle disease severity at onset and statin use. Therapy-related side effects were reported in 26% of patients.
Conclusions: In our population, anti-HMGCR IMNM was associated with a chronic disease course associated with IV Ig-dependence.