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Severe Axial and Pelvi-femoral Muscle Damage in Immune-Mediated Necrotizing Myopathy evaluated by Whole-body MRI.

Objectives: Immune-mediated necrotizing myopathy (IMNM) is associated with severe muscle involvement and early thigh muscle damage on MRI. Pattern and severity of muscle damage in other muscle groups and its relationship with clinical and serological features remain to be clarified.

Methods: IMNM patients with a whole-body MRI (WB-MRI) (n=42) were included as well as 60 IBM patients as controls. Each muscle groups (n=55) were evaluated and fat replacement was estimated in T1 sequence using the Mercuri score (1= normal; 2= mild, fat replacement < 30%, 3=moderate, 30-60%; 4=severe, > 60%). Overall lesion load was defined as the sum of all abnormal Mercuri scores (percentage) and lesion load quotient was defined as the overall lesion load divided by disease duration (years). Multidimensional analysis was performed to define homogenous groups of patients and to assess linear relationships between variables.

Results: IMNM patients (anti-HMGCR, n=25; anti-SRP, n=12 and sero-negative, n=5) at WB-MRI were aged 48.1±15.8 years and had a disease duration of 9.8 ±8.1 years. Most severely affected muscle groups were located in the pelvi-femoral (gluteus minimus: 2.71±1.15, gluteus medius: 2.6±1.17, great adductors: 2.55±1.27 and perineal: 2.43±1.42) and lumbar (lumbar extensors: 2.79±1.12) region.

Unsupervised analysis showed two subgroups of patients: one with a mild lesion load (15±10%, n=32/42) and another with a severe lesion load (60±10%, n=10/42: $p<0.001$). In the first group, the mean disease duration before WB-MRI was 6.8 ±6.0 years compare to 19.5±5.7 years in the second ($p<0.0001$). Correlational studies demonstrated that disease duration was the most important predictor of muscle damage (lumbar $r=0.76$, and pelvi-femoral muscle groups r ranging from 0.55-0.73; $p<0.01$).

Nonetheless, multivariate analyses – adjusted for disease duration, age at first symptoms and treatment initiation delay - demonstrated a more severe involvement of the great adductor ($p=0.02$) and the vastus lateralis ($p=0.02$) in female patients.

No difference was found between overall lesion load quotient in IMNM compared to IBM (14±14.9 vs 9.4±8, $p=0.3$), but muscle involvement pattern was different. Muscle damage progression over time was more severe in the trapezius ($p=0.04$), infraspinatus ($p=0.03$), psoas ($p < 0.001$), iliac ($p < 0.001$), longus adductor ($p=0.01$) and pectinus ($p < 0.001$) in IMNM.

Conclusions: IMNM is associated with a severe axial and pelvi-femoral muscle damage. Disease duration and gender status are important predictors of muscle damage. IMNM and IBM patients have a comparable overall lesion load, but progression was more severe in some muscle groups.