

Macrophage rich myositis with calpain-3 mutation

Figure legends

Figure 1.

Muscle biopsy from left quadriceps demonstrated regional clusters of necrotic and regenerating fibres surrounded by an inflammatory infiltrate (a). In some areas the process appears to respect fascicular boundaries. The inflammatory infiltrate reveals moderate numbers T-cells combined with numerous macrophages upon CD3 (b) and CD68 (c) immunohistochemical studies (same area as a). Clusters differed in regard to the presence of small basophilic fibres (d-f), suggesting various stages of regeneration. MHC Class I upregulation was restricted to regenerating fibres and was not present on neighboring normal appearing fibres (g). Regenerating fibres also expressed membranous utrophin (h) and neonatal myosin (i).

Figure 2.

Axial T1-weighted MRI of thighs and calves. There was fatty replacement in adductor magnus (black arrows), soleus, and medial gastrocnemius (black arrowheads) bilaterally. STIR sequence showed no evidence of water accumulation (not shown). The patient was on immunosuppressive treatment at the time of examination.

Figure 3.

Western blot analysis. Probing with antibodies against exon 1 of calpain did not detect bands for the full size protein or 30kDa fragment. Exon 8 antibodies showed a barely visible band for the full sized protein, much reduced compared to control.

Figure 1

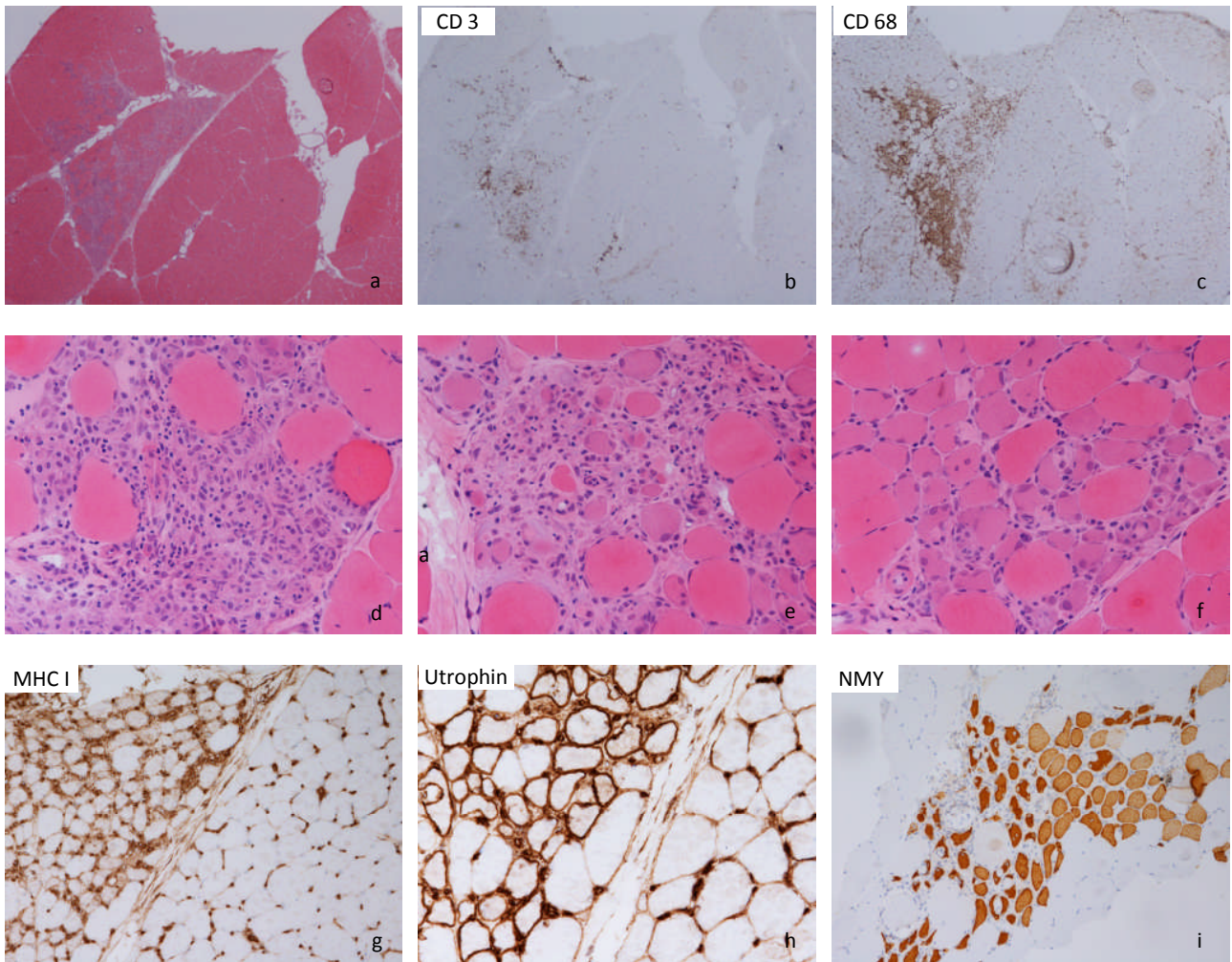


Figure 2

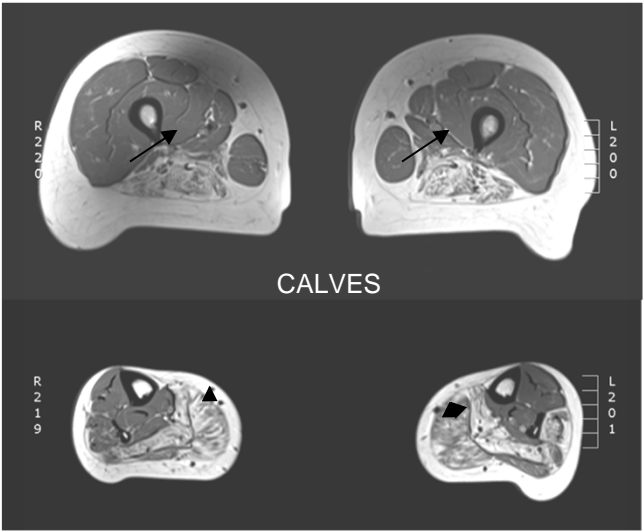


Figure 3

