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Supplementary Item 1:

Desmin Staining: For desmin staining of formalin-fixed sections, samples were deparaffinized, rehydrated through a graded alcohol series and antigens unmasked by heat-induced epitope retrieval using Decloaking Chamber (Biocare pressure cooker) and Target Retrieval Solution (Dako; catalog #: S1699). After rinsing in Tris Buffered Saline and Tween 20, endogenous peroxidases were blocked with hydrogen peroxide and then nonspecific binding was blocked with 10% normal goat serum. Sections were incubated with anti-desmin mouse monoclonal primary antibody (Dako; catalog # M0760; clone D33) at 1:200 dilution for 45 min. After rinsing the secondary linking antibody, EnVision+horse radish peroxidase (HRP) polymer conjugated goat anti-mouse immunoglobulins were applied (Dako; catalog #: K4001). Immunoreactivity was detected with AEC+ substrate chromogen (Dako; catalog #: K3469) and slides were counterstained with Mayer's hematoxylin. For frozen section IHC, cryostat cut sections were fixed in chilled acetone for 30 sec immediately after sectioning, air dried for 1hr, and then stored at -80c until staining. Sections were then post-fixed in 75% acetone/25% ethanol at 4°C for 10 min and air-dried before rinsing in Tris buffered saline with Tween. Nonspecific binding was blocked with 10% normal goat serum. Sections were incubated with the primary anti-desmin–mouse monoclonal antibody (Dako; catalog # M0760; clone D33; 1:100; 90 min). After rinsing, sections were blocked for endogenous peroxidase and incubated with a secondary linking antibody: EnVision+HRP polymer conjugate goat anti-mouse immunoglobulins (Dako; catalog #: K4001). Immunoreactivity was detected with AEC+ substrate chromogen (Dako; catalog #: K3469) and slides were counterstained with Mayer's hematoxylin.

Additional immunohistochemistry: The procedure was similar to that of frozen section desmin staining and used primary antibodies; anti-αβ-crystallin–rabbit polyclonal (Millipore; catalog #: ABN185 1:100; 90 min), anti-nebulin–rabbit polyclonal (Novus; catalog #: NBP1-87752 1:75; 90 min) and anti-dystrophin–rabbit polyclonal (abcam; catalog #: ab8502 1:1000; 60 min). Serial sections were also stained for myosin adenosine triphosphatase (ATPase) using pH 4.6

preincubation and in order to determine the fiber type specificity of protein aggregates. Nicatinamide adenine dinucleotide tetrazolium reductase (NADH) staining was performed to evaluate abnormalities in oxidative staining.

Electron microscopy: The samples were fixed with 2.5% glutaraldehyde overnight at 4C. After washing three times with 0.1 M sodium cacodylate, the samples were post-fixed with 1% OsO_4 for 40 min. Following three rinses, the samples were dehydrated in a grade series of ethanol. The samples were embedded in Epon 812 resin. Ultrathin sections (65 nm) were stained with uranyl acetate and lead citrate and then examined by JEOL 1200EX electron microscopy.