57th ANNUAL DIAGNOSTIC SLIDE SESSION 2016.

CASE 2016-7

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Clinical History: The patient is a 78 year old woman who presented with approximately 10 years of progressive lower extremity weakness causing difficulty with ambulation requiring her to use a cane or walker and eventually a wheelchair. She has also had progressive weakness in her arms more recently over the last couple years. She has been evaluated at multiple institutions throughout her disease course, and her serum CK level has never been elevated. She has had multiple electromyography and nerve conduction studies in the past that were reportedly without abnormalities. Acetylcholine receptor autoantibody studies were negative. She has not responded to trials of prednisone, mycophenolate, azathioprine, or pyridostigmine. Physical exam at current presentation revealed normal muscle tone and bulk in all extremities. However, she had diminished strength in her bilateral hip flexors (3/5), hip abductors (4/5), hamstrings (4/5), anterior tibialis (4/5), and extensor halluces longus (4/5). She had decreased vibratory, pin pick, and light touch sensation bilaterally in the lower extremities up to her shins. She demonstrated a kyphotic posture and antalgic gait. Repeat electromyography and nerve conduction studies demonstrated myopathic motor units without evidence of muscle membrane instability in multiple muscle groups consistent with a myopathic process. A biopsy of the biceps muscle was performed.

Material submitted: H&E, Gomori trichrome, NADH, ATPase at pH 9.2, fast myosin, and slow myosin stained frozen sections from biceps muscle biopsy. Also toluidine blue stained sections from a portion of the biopsy that was glutaraldehyde-fixed and Epon embedded for ultrastructural analysis.

Points for discussion: 1. Diagnosis
2. Pathogenesis