The role of quantitative electromyography in inclusion body myositis

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Abstract

Objective and methods—Inclusion body myositis is said to have both myopathic and neurogenic features on electrophysiological tests. Twenty one studies from 20 patients with biopsy defined inclusion body myositis, 13 of whom had quantitative electromyography (qEMG), were reviewed to determine if this technique added diagnostic specificity (one patient had both needle EMG and a later study with qEMG before muscle biopsy).

Results—Excessive numbers of polyphasic motor unit potentials (MUPs) (>12% per muscle) were seen in 11 of the 13 patients. In 10 of 13 patients, mean MUP duration was abnormally reduced (26% to 48%). In three patients, mean MUP duration was abnormally reduced only after polyphasic MUPs were excluded. In all 13 patients, the simple MUP duration was reduced. Myopathy was unequivocally diagnosed in all 13 studies that included qEMG; of the remaining eight patients, the conclusions of the electrophysiological studies without qEMG was myopathy (one), neurogenic (four) or non-diagnostic (three).

Conclusions—There is no evidence of a neurogenic component in inclusion body myositis if qEMG is used. Quantitative EMG is often necessary to make an electrophysiological diagnosis of a myogenic disorder in patients with inclusion body myositis.

Keywords: inclusion body myositis; quantitative electromyography; motor neuron disease; fasciculations; motor unit potentials; myopathy

Inclusion body myositis is a chronic inflammatory myopathy, clinically characterised by both proximal and distal limb weakness and a poor response to steroid treatment. Pathologically, the condition is defined by the presence of rimmed vacuoles, filamentous inclusions, and intracellular amyloid deposits. Electrophysiological studies show features of a myopathy, but several investigators have reported neurogenic changes, including reduced recruitment and "neurogenic motor unit potentials." Quantitative EMG (qEMG) separates neurogenic from myogenic disease at least as well as muscle biopsy. To determine if the use of qEMG enhances the specificity of the physiological findings we reviewed the electrophysiological features of patients with inclusion body myositis, with and without qEMG to determine if there were findings suggestive of a neurogenic component.

Methods

We reviewed the neurophysiological studies performed at Columbia-Presbyterian Medical Center of 20 consecutive patients with muscle biopsies diagnosed as inclusion body myositis by one of us (APH). Biopsies had features of a myopathy with rimmed vacuoles and inflammatory cells. In selected cases, electron microscopy was performed to identify the characteristic 15-18 nm filaments. The neurophysiological tests preceded the biopsy in all cases, except for patient 15 as noted in table 2.

Quantitative EMG was done as previously described. Twenty motor unit potentials (MUPs) were collected with a concentric needle. Band path was 2 Hz-20 kHz. Duration was measured between the initial deflection of the MUP from the baseline and the terminal return to baseline at a sensitivity of 100 µV/division. Mean duration was calculated for the total number of measured MUPs. Mean duration of simple MUPs (those with fewer than five phases) was calculated separately. Amplitude was measured from peak to peak. Values were considered abnormal when the mean duration deviated by more than 20% beyond the normal mean duration of the specific muscle, matched for the age of the patient.

All qEMG studies were done by physicians trained and experienced in the technique. The experience of electromyographers performing only routine EMG was equivalent.

Results

Twenty patients with symptoms for one to 13 years were identified with clinical and electrophysiological features as summarised in tables 1 and 2. Some patients were referred with the diagnosis of ALS and although they did not have definite upper motor neuron signs such as a Babinski’s sign or clonus, reflexes were preserved in weak and wasted limbs. All biopsies showed rimmed vacuoles and endomysial fibrosis. Inflammatory cells and groups of atrophic fibres were seen in most cases. Target fibres or fibre type grouping were not seen in any biopsy.

Sensory and motor conduction velocities were normal in all but two patients. Quantitative EMG (qEMG) was performed in 13 patients. Excessive numbers of polyphasic MUPs were seen in 11 patients (15% to 60%). In 10 of these patients, the mean duration of all
TABLE 2 Electrodiagnostic results from 20 patients with inclusion body myositis

<table>
<thead>
<tr>
<th>Patient No</th>
<th>Referring diagnosis</th>
<th>EMG diagnosis</th>
<th>MNC</th>
<th>SNC</th>
<th>Muscle</th>
<th>MUPs (n)</th>
<th>Poly</th>
<th>Duration of simple MUPs (µV)</th>
<th>Amplitude (µV)</th>
<th>Spontaneous activity</th>
<th>Recruitment</th>
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<tr>
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<td>N</td>
<td>Tib ant</td>
<td>20</td>
<td>15</td>
<td>10.88 (68)</td>
<td>9.9 (67)</td>
<td>330</td>
<td>1-4 fbs</td>
</tr>
<tr>
<td>2</td>
<td>ALS</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Tib ant</td>
<td>20</td>
<td>30</td>
<td>9.5 (62)</td>
<td>8.8 (57)</td>
<td>496</td>
<td>1-3 fbs/</td>
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<tr>
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<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>24</td>
<td>0</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>147</td>
<td>1-2 fbs</td>
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<td>4</td>
<td>ALS v N</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Tib ant</td>
<td>28</td>
<td>21</td>
<td>11.3 (73)</td>
<td>10.6 (69)</td>
<td>329</td>
<td>1-2 fbs</td>
</tr>
<tr>
<td>5</td>
<td>PM</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>25</td>
<td>8</td>
<td>11.9 (96)</td>
<td>11.4 (93)</td>
<td>215</td>
<td>1-2 fbs</td>
</tr>
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<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>20</td>
<td>10</td>
<td>8.4 (74)</td>
<td>8.5 (75)</td>
<td>514</td>
<td>1-2 fbs</td>
</tr>
<tr>
<td>7</td>
<td>Myopathy</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>20</td>
<td>50</td>
<td>8.4 (84)</td>
<td>8.4 (84)</td>
<td>227</td>
<td>1-2 fbs</td>
</tr>
<tr>
<td>8</td>
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<td>N</td>
<td>Biceps</td>
<td>20</td>
<td>25</td>
<td>8.1 (65)</td>
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<td>N</td>
<td>Biceps</td>
<td>20</td>
<td>50</td>
<td>8.4 (84)</td>
<td>8.4 (84)</td>
<td>227</td>
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<td>Myopathy</td>
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<td>N</td>
<td>Tib ant</td>
<td>20</td>
<td>15</td>
<td>10.88 (68)</td>
<td>9.9 (67)</td>
<td>330</td>
<td>1-4 fbs</td>
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<tr>
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<td>Myopathy</td>
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<td>N</td>
<td>Biceps</td>
<td>28</td>
<td>21</td>
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<td>10.6 (69)</td>
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<td>1-2 fbs</td>
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<tr>
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<td>Myopathy</td>
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<td>N</td>
<td>Biceps</td>
<td>25</td>
<td>8</td>
<td>11.9 (96)</td>
<td>11.4 (93)</td>
<td>215</td>
<td>1-2 fbs</td>
</tr>
<tr>
<td>13</td>
<td>ALS v N</td>
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<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>20</td>
<td>10</td>
<td>8.4 (74)</td>
<td>8.5 (75)</td>
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<td>1-2 fbs</td>
</tr>
<tr>
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<td>MND</td>
<td>N</td>
<td>N</td>
<td>P</td>
<td>Long</td>
<td>21</td>
<td>8.4 (68)</td>
<td>6.7 (54)</td>
<td>193</td>
<td>1-2 fbs</td>
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<tr>
<td>15</td>
<td>ALS</td>
<td>SMN</td>
<td>N</td>
<td>N</td>
<td>P</td>
<td>Long</td>
<td>21</td>
<td>8.4 (68)</td>
<td>6.7 (54)</td>
<td>193</td>
<td>1-2 fbs</td>
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<tr>
<td>16</td>
<td>ALS v N</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>P</td>
<td>R</td>
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<td>6.7 (54)</td>
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<td>N</td>
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<td>N</td>
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</table>

MUPs was abnormally reduced by 26% to 48%. In three patients, the mean duration of all MUPs was normal, but it was abnormally reduced if only simple MUPs were considered (34% to 46% below the normal mean; table 3).
myopathy and fasciculations are also occasionally seen in various muscle diseases. The presence of long duration polyphasic MUPs in myopathies correlate with regenerating fibres. Patients with neuropathic, myopathic, or normal EMG may have individual MUPs that could be considered “neuropathic” (>16 ms) or “myopathic” (<6 ms). Only the mean duration of a sufficient number of simple MUPs correlates with the presence of a neuropathy or myopathy. In one report of eight patients with inclusion body myositis, no patients showed neurophysiologic changes using quantitative EMG. Studies using single fibre EMG and macro-EMG in patients with inclusion body myositis also found no evidence of a neuropathic process or reinnervation as a cause of the long duration polyphasic motor units seen.

Quantitative EMG is necessary to accurately characterise the myogenic motor unit morphology in some cases of inclusion body myositis. Without qEMG, in the presence of spontaneous activity (fibrillations, positive sharp waves, and fasciculations), long duration polyphasic MUPs, and reduced recruitment, a pure motor neurogenic disorder, such as motor neuron disease, may be erroneously suspected.

We thank Dr Lewis P Rowland for helpful comments on the manuscript. Presented in part in abstract form at the meeting of the American Neurology Association, Washington, DC, 22-25 October 1995.

Reduced recruitment is a qualitative measurement, but is also non-specific because it may be seen in myopathies when loss of fibres is so extensive that whole motor units drop out.

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