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 myosis, 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 myosis if qEMG is used. Quantitative EMG is often necessary to make an electrophysiological diagnosis of a myogenic disorder in patients with inclusion body myosis.**

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

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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.[1 2] Electrophysiological studies show features of a myopathy,[3] but several investigators have reported neurogenic changes, including reduced recruitment and "neurogenic motor unit potentials."[4–10]

Quantitative EMG (qEMG) separates neurogenic from myogenic disease at least as well as muscle biopsy.[11] To determine if the use of qEMG enhances the specificity of the physiological findings we reviewed the electrophysiological features of patients with inclusion body myosis, 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 myosis 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.[12 13] 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.[14]

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 1 Clinical characteristics

Sex (M:F)	14:6	
Mean age at onset (range)(y)	62	(44–73)
Duration of symptoms (mean (range)y)	6	(1–13)
Patients given diagnosis of ALS	7	
Cramps	0 of 10	
Symptoms at onset (n (%)):		
Legs	13 of 20	65
Arms	6 of 20	30
Dysphagia	2 of 20	10
Asymmetry at onset (n (%))	5 of 20	25
Finger flexor weakness or atrophy (n (%))	13 of 20	65
Quadriceps weakness or atrophy (n (%))	17 of 20	85
Neck weakness (n (%))	3 of 11	27
Bulbar symptoms (n (%))	6 of 19	32
Facial weakness (n (%))	7 of 18	39
Mean serum CK (X upper limit of normal)	4.1	0.7–12.9
Response to IVIG (minimal)	4 of 7	
Response to prednisone	2 of 9	
Response to azathioprine (imuran)	0 of 3	

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).

Therefore a myogenic disorder was diagnosed in all 13 studies in which qEMG was done. Of the remaining eight studies, when formal analysis of MUP form was not undertaken the findings were interpreted as neurogenic (four), including three as motor neuron disease or non-diagnostic (three). In only one of eight patients was a myogenic disorder correctly diagnosed (table 3). In one patient (10), the EMG was interpreted as showing motor neuron disease, without using quantitative EMG; when repeated five months later, qEMG disclosed short duration MUPs compatible with myopathy.

Eleven of the 20 patients with inclusion body myosis were referred to us with the diagnosis of motor neuron disease or neuropathy. A myogenic disorder was diagnosed in all five in whom qEMG was done. In only one of the six patients tested without qEMG was the myogenic nature of the illness identified using physiological methods.

Two patients (6 and 13), with EMG evidence of fasciculations, had biopsies of the motor nerve to the gracilis muscle to consider the possible coexistence of a motor neuropathy.

Table 2 Electrodiagnostic results from 20 patients with inclusion body myositis

Patient No	*Referring diagnosis*	*EMG diagnosis*	*MNC*	*SNC*	*Muscle*	*MUPs (n)*	*Polys (%)*	*Duration (ms) (% normal)*	*Duration of simple MUPs (% of normal)*	*Amplitude (μV)*	*Spontaneous activity*	*Recruitment*
1	IBM	Myopathy	N	N	Tib ant	20	15	10 (68)	9.9 (67)	330	1+fibs	4, 6
2	ALS	Myopathy	↓a (p)	N	Ttib ant	20	30	9.5 (62)	8.8 (57)	496	1-3+fibs/ 1+fascics	2, 5, 6
3	MND	Myopathy	N	N	Vas lat	28	21	11.3 (73)	10.6 (69)	329	1+fibs	1, 4
					Biceps	24	0	10.2 (84)	10.2 (84)	147		
					Deltoid	25	8	11.9 (96)	11.4 (93)	215		
4	ALS *v* N	Myopathy	N	N	Vas lat	17	12	17.6 (114)	17.7 (115)	910	2+fibs/ 1+fascics	1, 3, 4, 5
					Tib ant	15	15	7.8 (52)	6.4 (43)	641		
5	PM	Myopathy	N	N	Biceps	20	10	7.7 (60)	7.9 (62)	419	2+fibs	4
6	Myopathy	Myopathy	N	N	Biceps	20	10	8.4 (74)	8.5 (75)		1-2+fibs/ fascics/myo-kymia/CRDS	5
7	Myopathy	Myopathy	N	N	Biceps	20	50	8.4 (68)	8.4 (68)	227	1-2+fibs	4, 6
8	PM	Myopathy	N	N	Biceps	20	25	8.1 (65)	7.4 (60)	514	fibs	4
9	Myopathy	Myopathy	<a (t)	N	Vas lat	18	44	9.6 (62)	9.4 (60)	749	1+fibs/ 2+fascics/CRDS	1, 2, 4, 6
10	SMA *v* myopathy	MND	↓a(u, p, t) ↓cv(t)	↓CV(m, u, s)			P	Long			1-2+fibs/ 1-2+fascics/ CRDS	2, 3
	SMA	Myopathy	Not done	Not done	Biceps	21	24	8.4 (68)	6.7 (54)	193	rare fibs/CRDS	2, 4
					Vas lat	18	39	11.3 (72)	11.6 (74)	374		
11	Myopathy	Myopathy	↓a(p)	N	Tib ant	20	30	20 (136)	8 (54)	455	2+fibs	1, 2, 5
12	Myopathy	Myopathy	N	N	Vas lat	28	29	14.9 (102)	13.9 (95)	729	1+fibs	1, 2
					Tib ant	22	50	12.7 (89)	9.4 (66)	358		
13	ALS	Myopathy	N	N	Biceps	20	60	11.3 (98)	6.9 (60)	589	1-2+fibs/ fascics/CR DS	5
14	ALS	MND	N	N		—	P	Long		I	1-2+fibs/ 1-3+fascics	2, 3
15	ALS	SMN	↓a(u, p)	↓CV(m, u, s), <a(m, u)		—	P			I	1-2+fibs/fascics	2, 3
After bx	ALS *v* N	SMN	↓CV(m, p), <a(p)	↓cv,a(m,u), ab (s)		—	P			I	1-2+fibs/ 1+fascics	2, 3
	Myopathy	Myo/ SMN	↓a(p, t)	↓CV, a(m), ab(s)		—	P	R		R,I	1-2+fibs	2, 4
	Myopathy	Myo/ SMN	↓CV(p, t), ↓a(p, t)	↓CV, a(m, u), ab(s)		—						
16	IBM *v* MND	Myopathy	N	N			N	R		R	1+fibs	2, 4
17	PM *v* PN	Non-diag	N	N		—	P	N		N	None	2
18	MND	MND	N	ab(s)		—	P			I	1-2+fibs	1, 2, 4, 6
19	Myopathy	Normal	N	N		—	N	N		N	None	1
20	Myositis	Normal	N	Ab(s)		—	N	N		N	None	1, 6

a=amplitude; ab=absent; ALS=amyotrophic lateral sclerosis; CRDS=complex repetitive discharges; fascics=fasciculations; fibs=fibrillations and positive sharp waves; CV=conduction velocity; I=increased; IBM=inclusion body myositis; m=median nerve; MND=motor neuron disease; N=normal; np=neuropathy; p=peroneal; PM=polymyositis; R=reduced; s=sural; SMA=spinal muscular atrophy; SMN=sensorimotor neuropathy; t=tibial nerve; u=ulnar nerve; 1=full recruitment on maximal effort; 2=reduced recruitment on maximal effort; 3=discrete recruitment on maximal effort; 4=full recruitment on submaximal effort; 5=full recruitment on maximal effort in a weak muscle; 6=reduced recruitment on submaximal effort.

Table 3 Electrophysiological findings

Findings	*n (%)*	
Nerve conduction:		
Reduced CMAP	5 (25%)	
Slow motor and sensory conduction velocity	2 (10%)	
EMG:		
Fasciculations	8 (40%)	
Fibrillations/Positive sharp waves	17 (85%)	
Myokymia	1 (5%)	
Complex repetitive discharges	4 (20%)	
Quantitative EMG in 13 patients:		
Increased polyphasics (15-60%)	11 (85%)	
Short mean duration - all MUPs (26-48%)	10 (77%)	
Short mean duration - simple MUPs (34-46%)	13 (100%)	
Quantitative EMG and diagnosis of myopathy:	*qEMG done*	*qEMG not done*
Diagnosis of myopathy	13	1
No diagnosis of myopathy	0	7

The motor nerve was morphologically normal in both and immunohistochemistry for nerve growth factor receptor was absent in one (6) and in the other there was only trace expression (13).

Discussion

Claims of a neuropathic component in inclusion body myosis have been based on features from muscle biopsy and electrophysiological testing. Group atrophy and angular fibres are seen which are suggestive although not specific for a neurogenic disorder. Fibre type grouping, which is specific for a neurogenic disorder, is generally not seen[4–7 9 10] unless there is a concomitant neuropathy as occurred in three of 48 patients in one series.[3] Long duration or "neurogenic" MUPs on EMG are also cited as evidence of a neurogenic component in inclusion body myosis.[4–9] In fact, long duration polyphasic MUPs are characteristically found in chronic myogenic disease.[15 16] Fibrillations and positive sharp waves are often seen in myopathy[17] and fasciculations are also occasionally seen in various muscle diseases.[18] One report found fasciculations in 10% of patients with inclusion body myosis.[3] Our findings, therefore, do not support the presence of a neurogenic component in inclusion body myosis if qEMG is employed.

Nerve conduction studies were normal except for mildly decreased evoked response amplitude in five of our patients, compatible with muscle atrophy, which may result from a myopathy. Sensory and motor conduction velocities were normal in all but two. One patient (15) had a coexisting neuropathy with sensory loss, absent ankle reflexes, and a raised CSF protein content. Nerve biopsy disclosed a severe neuropathy with rare onion bulbs. The second patient (10), a 68 year old woman with no sensory complaints, had mild slowing of the median, ulnar, and sural sensory nerves and the tibial motor nerve. The amplitude of the sensory responses was normal however, and low skin temperature was the implicated cause. The abnormalities on conduction studies in these two patients, therefore, can be explained by factors other than implicating neurogenic involvement of inclusion body myosis.

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.[19]

The presence of long duration polyphasic MUPs seen in myopathies correlate with regenerating fibres.[16] Patients with neuropathic, myopathic, or normal EMG may have individual MUPs that could be considered "neuropathic" (>16 ms) or "myopathic" (<6 ms).[13 20] Only the mean duration of a sufficient number of simple MUPs correlates with the presence of a neuropathy or myopathy.[11] In one report of eight patients with inclusion body myosis, no patients showed neuropathic changes using quantitative EMG.[21] Studies using single fibre EMG[22] and macro-EMG[23] in patients with inclusion body myosis 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 myosis. 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.

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