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Electromyographic findings in sporadic inclusion body myositis

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ABSTRACT

Introduction: Clinically oriented diagnostic criteria can be as specific for diagnosis of sporadic inclusion body myositis (sIBM) as pathological criteria, especially at the time of presentation. EMG may provide an convincing proof that a muscle biopsy should be performed.
Aims: To compare the EMG results in patients with sIBM divided into subgroups based on the newest ENMC criteria for sIBM and to obtain the utility of EMG in the diagnostic process at the time of presentation.
Methods: We retrospectively analysed 16 patients with sIBM for motor unit action potential (MUAP) morphology as well as occurrence and distribution of abnormal spontaneous activity (SA) in muscles.
Results: Abnormal SA was recorded in 62.5% of sIBM patients. We found statistically significant differences between subgroups in the incidence of polyphasic MUAPs and high amplitude outliers which were more commonly seen in the "*clinico-pathologically defined sIBM*". Duration of MUAP in the tibialis anterior was significantly shorter in "*probable sIBM*".
Discussion: "Pseudo-neurogenic" MUAPs, mainly in lower limb muscles, are more commonly seen in "*clinico-pathologically defined sIBM*" while myopathic MUAPs with prominent abnormal SA are recorded in patients diagnosed with "*probable sIBM*". Both EMG patterns may be suggestive of sIBM and be an indication for further diagnosis.

1. Introduction

The term sporadic inclusion body myositis (sIBM) has usually been used to describe an inflammatory myopathy that develops in mid- or later life with a distinctive pattern of weakness (asymmetric involvement of the wrist flexors, finger flexors and quadriceps muscles) and distinctive pathologic features (inflammatory cells surrounding myofibers and rimmed vacuoles) (Greenberg, 2013). Although sIBM has been historically classified as an inflammatory myopathy, some of its features differentiate it from dermatomyositis (DM) or polymyositis (PM) and suggest degenerative pathophysiology (e.g., ineffectiveness of immunomodulatory and immunosuppressive treatment as well as a tendency for coexistence of both "neurogenic" and "myopathic" findings on EMG) (Barkhaus et al., 1999). In the initial IBM diagnostic criteria, EMG results were necessary to make the diagnosis of possible IBM (Griggs et al., 1995). As far as the histopathological criteria are concerned, concomitant presence of all cardinal changes (inflammatory, myodegenerative and mitochondrial) is highly specific for the diagnosis of IBM. However, it is uncommon to find them all in a biopsy, especially in early stages of the disease (Machado et al., 2014). Long-term clinical observations have shown that these pathological features may be absent, even on a repeat study, when patient follow-up confirms the correct clinical diagnosis (Suzuki et al., 2012; Tan et al., 2013). Although muscle biopsy remains the definitive diagnostic procedure, nowadays a shift from relaying mostly on histopathological criteria to an increasing importance of clinical features and clinico-pathological correlations can be observed (Chahin and Engel, 2008; Brady et al., 2013; Rose et al., 2013; Lloyd et al., 2014). It allows a diagnosis of IBM in the absence of what has previously been considered essential pathological features (Hilton-Jones and Brady, 2016).

Since 1995 when the Griggs criteria where established, there have been several proposals for revised diagnostic criteria, the most recent coming from the European Neuromuscular Centre (ENMC) Workshop in 2011 (Rose et al., 2013). In a recent review, Lloyd et al. (2014) analysed the sensitivity and specificity of the published criteria. They emphasised that all published criteria had high specificity (98–100%) but wide-ranging sensitivity (11–84%) and poor practical application of some of them could be attributed to relaying strongly on the pathological criteria which are highly specific but insensitive. Using features recorded at presentation, Hilton-Jones and Brady (2016) compared the

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Griggs band 2011 ENMC diagnostic criteria and concluded that clinically oriented diagnostic criteria can be as specific for the diagnosis of sIBM as conventional pathological criteria, and they show substantially greater sensitivity at the time of presentation.

It shows that EMG can still be a valuable procedure in the diagnostic process in sIBM patients, especially at the initial presentation. Previously published reports of the EMG findings were mostly based on studies in small- to medium-sized patient groups classified based on changing diagnostic criteria. Overall, the findings are thought to be nonspecific and the EMG examination has not been included in the latest criteria for sIBM (Rose et al., 2013). In clinical practice, however, EMG results still play an important role in the diagnostic work-up and selection of patients in whom a muscle biopsy should be performed (Needham and Mastaglia, 2016). In addition, Dalakas (2015) pointed out that EMG results may be useful for ruling out a neurogenic process and determining whether myopathy is active or chronic although they cannot be used for differentiating inflammatory myopathies from toxic and dystrophic myopathies.

In the present study, we analysed EMG results in patients with "clinico-pathologically defined" and "probable" sIBM diagnosed based on the latest ENMC diagnostic criteria for sIBM (Rose et al., 2013) to obtain the utility of EMG in the diagnostic work up at the time of presentation.

2. Patients and methods

We evaluated retrospectively collected data of 16 patients (5 women and 11 men) aged 35 to –77 years with the diagnosis of sporadic IBM. The age at the disease onset varied from 21 to 74 years. All patients had a negative family history. When the criteria of Rose et al. were applied, "*clinico-pathologically defined sIBM*" was recognized in 10 cases (8 men and 2 women, mean age at the time of EMG examination 58.0 ± 14.0 years) and "*probable sIBM*" was recognized in 6 cases (3 men and 3 women, mean age at the time of EMG examination 60.6 ± 13.5 years). Clinical and morphological data of 7 patients were discussed in a previous report by Kierdaszuk et al. (2015). Both groups fulfil the clinical and laboratory features (duration of the disease, knee extension weakness > hip flexion weakness and/or finger flexion weakness > shoulder abduction weakness and serum CK level no greater than 15x UL). However only in "*clinico-pathologically defined sIBM*" subgroup all pathological features (endomysial inflammatory infiltrate, rimmed vacuoles and protein accumulation or 15–18 nm filaments) in muscles biopsies were found. Patients with "*probable sIBM*" did not fulfil the abovementioned pathological criteria or, in one case, presented mostly with proximal upper limb (pUL) muscle weakness. Pelvic girdle (PG) muscles were mostly affected but distal lower limb (LL) muscles were also involved in the majority of patients. Knee extensor and finger flexor weakness s was more pronounced than that of hip flexors and shoulder abductors, respectively. One patient used a wheelchair, three patients used a walking stick, while the remaining patients were mobile. Difficulties in swallowing were not reported. Serum creatine kinase (CK) activity ranged from 10 to 406 U/L (reference range 0–34 U/L), and thus it was within 15 times above the upper limit of normal. All patients were diagnosed and treated at the Department of Neurology, Medical University of Warsaw. Clinical characteristics of patients are presented in Table 1.

The study protocol was approved by the Bioethical Committee at the Medical University of Warsaw (No AKBE 122/2015). All the procedures were in accordance with the standards of the Committee on Human Experimentation at the Medical University of Warsaw and with the Helsinki Declaration of 1975.

Open muscle biopsies were performed under local anesthesia during routine diagnostic work-up. Twelve samples were obtained from the quadriceps and the remaining four from the biceps brachii muscle. The muscle specimens were processed for further analyses using routine histological, histochemical and ultrastructural methods (Dubowitz and Sewry, 2007). All biopsies were assessed using light microscopy (LM) and 11 of them also using electron JEM 1200 EX2 microscopy (EM). Morphological abnormalities including endomysial inflammatory infiltrates, rimmed vacuoles, β-amyloid or TDP-43 accumulation or inclusions composed of 15–18 nm tubulofilaments in EM were present in eleven cases. In the last five cases, assessment by EM was not performed. Together with the patient with predominant proximal upper limb weakness, these cases were categorized as "*probable sIBM*".

Table 1
Comparison of clinical and electrophysiological findings between the "*clinico-pathologically defined*" (n = 10) and "*probable*" (n = 6) sporadic inclusion body myositis (sIBM).

Characteristics	"*Clinico-pathologically defined*" sIBM	"*Probable*" sIBM	P
Total number of patients	10	6	
Gender (F:M)	2:8	3:3	NS
Mean age at the disease onset, yrs (range)	53.6 ± 13.6 (21–72)	49.2 ± 19.1 (25–74)	NS
Mean age at the time of examination, yrs (range)	58.0 ± 14.0 (22–76)	60.6 ± 13.5 (35–77)	NS
Limb onset			
Lower limb	10	4	NS
Upper limb	6	2	NS
Dysphagia	0	0	NS
Tested muscles (n)	53	23	
BB	16	8	NS
FID	7	2	NS
VL	15	8	NS
TA	14	5	NS
Number of muscles with myopathic MUAPs recorded (n; %)	20 (37.7)	15 (65.2)	
BB	7 (43.8)	5 (62.5)	NS
FID	0 (0)	0 (0)	NS
VL	6 (37.5)	5 (62.5)	NS
TA	7 (50)	5 (100)	NS
Number of muscles with "*pseudo-neurogenic*" MUAPs recorded (n; %)	16 (30.2)	1 (4.3)	
BB	1 (6.3)	1 (12.5)	NS
FID	3 (42.9)	0 (0)	NS
VL	6 (37.5)	0 (0)	0.0417
TA	6 (42.9)	0 (0)	0.0685
Number of muscles with mixed MUAPs recorded (n; %)	4 (7.5)	2 (8.7)	
BB	0 (0)	1 (12.5)	NS
FID	0 (0)	0 (0)	NS
VL	3 (18.8)	0 (0)	NS
TA	1 (7.1)	1 (20)	NS
Occurrence of fib./PSW (n; %)	11 (20.8)	7 (30.4)	
BB	2 (12.5)	2 (25)	NS
FID	0 (0)	0 (0)	NS
VL	4 (25)	2 (25)	NS
TA	5 (35.7)	3 (60)	NS
Occurrence of CRD (n; %)	3 (5.7)	3 (13)	
BB	1 (6.3)	1 (12.5)	NS
FID	0 (0)	0 (0)	NS
VL	1 (6.3)	2 (25)	NS
TA	1 (6.3)	0 (0)	NS
NCS results			
Normal	8	3	NS
Decreased CMAP amplitude	2	3	NS
Sensory neuropathy	0	0	NS

BB, biceps brachii; FID, first interosseous dorsalis; VL, vastus lateralis; TA, tibialis anterior; MUAP, motor unit action potential; NCS, nerve conduction study; CMAP, compound muscle action potential; NS, not significant; fib., fibrillation potentials; PSW, positive sharp waves; CRD, complex repetitive discharges.

2.1. EMG examinations

Electrophysiological studies were performed using Keypoint,