Electrophysiological and Clinical Examination of Polymyositis: A Retrospective Analysis

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Abstract: *Background:* We retrospectively analyzed electromyogram (EMG), laboratory and clinical data of 93 patients with polymyositis (PM) to help improve early diagnostic accuracy. *Methods:* Data were collected from hospitalized patients who were diagnosed with PM between January 1, 2006 and April 1, 2010 at Chinese People's Liberation Army General Hospital. *Results:* Eighty-six (92.47%) patients exhibited an isolated myopathic EMG pattern, 3 (3.23%) showed myopathy combined with neuropathy and 1 (1.08%) had diabetes mellitus. Among the 4 without myopathic EMG, 1 had inflammatory myopathy, and 3 had prior corticosteroid treatment with clinical improvement before EMG examination. Muscle biopsy, performed in 40 cases, inclusive of the 7 cases that did not show an isolated myopathic EMG patterns, revealed that 80% exhibited typical pathological features of PM, 17.5% showed nonspecific muscle fiber necrosis and 2.5% were normal. EMG showed that tibialis anterior and biceps brachii exhibited more abnormal EMG results than abductor pollicis brevis. Patients with disease duration >3 months had significantly higher alanine aminotransferase and reduced motor unit potential (MUP) amplitudes compared to patients with shorter disease duration. Statistical analysis revealed a significant association between disease duration and MUP amplitude reduction. Patients without interstitial lung disease showed significantly longer disease duration and higher creatine kinase and lactate dehydrogenase levels than patients with interstitial lung disease. *Conclusions:* Our results show that MUP is a sensitive yardstick for diagnosis of PM and is inversely related to disease duration. Our study also highlights that selecting specific muscles for EMG may improve diagnostic accuracy in PM.

Key Indexing Terms: Polymyositis; Electromyogram; Retrospective analysis; Interstitial lung disease; Creatine kinase. **[Am J Med Sci 2014;348(2):162–166.]**

Polymyositis (PM) is an idiopathic inflammatory myopathy (IIM) characterized by progressive muscle weakness, increased levels of serum muscle enzymes and abnormal electromyography (EMG) findings.[1] The European Neuromuscular Center divided IIM into 5 subtypes: PM, dermatomyositis (DM), sporadic inclusion body myositis (sIBM), nonspecific myositis and immune-mediated necrotizing myositis.[2] The prevalence of PM within IIMs varies widely among different studies. An epidemiological survey of a population in Minnesota showed that the incident rate of sIBM and PM was 0.79% and 0.41%, respectively,[3] whereas a New Zealand study carried out between 1989 and 2001 reported that 41%, 39% and 14% of IIM patients had DM, PM and sIBM, respectively.[4] PM is unique in that it is often associated with disorders in other systems and its onset usually involves acute proximal muscle weakness accompanied by an increase in serum creatine kinase (CK) levels.[1] The pathological entity of PM is the T cell, especially those that are CD8 positive. CD8-positive cells surround and invade muscle fibers expressing the major-histocompatibility class-I antigen.[5]

PM is a rare disease and reduces patient movement and causes disability, both of which can affect their quality of life or even endanger it.[1] The 5-year mortality rate can be as high as 1 in 5 patients and is significantly influenced by the time of diagnosis and complications, with an early diagnosis having higher chance to ensure a better prognosis.[6,7] Corticosteroid therapy can relieve symptoms, reduce impairment and improve quality of life, especially when administered in the early stages of disease.[8,9] Therefore, early diagnosis is critical for maximizing good prognosis.

EMG is an effective and safe tool for PM diagnosis and disease monitoring.[6,7] Abnormal EMG results in patients with PM include abnormal insert potential, spontaneous fibrillation potential, reduced motor unit potential (MUP) duration and amplitude, increased polyphasic MUP and myopathic interference patterns. In severe cases with extensive muscle fiber necrosis, EMG often shows neuropathy or mixed pathology.[10] Some studies have reported different EMG patterns in different muscles.[11] Among the different parameters that can be assessed with EMG, decreased MUP duration is a sensitive outcome of disease progress.[11]

In this study, we retrospectively analyzed and compared EMG and clinical data collected from 93 patients with PM between 2006 and 2010 to understand the EMG pattern and clinical outcomes in patients with PM. Our findings will be useful for clinicians when interpreting test results and deciding whether to diagnose a patient with PM especially when the muscle biopsy is not available.

METHODS

Patients, Inclusion and Exclusion Criteria

Data were collected from hospitalized patients who were diagnosed with PM between January 1, 2006 and April 1, 2010 at Chinese PLA General Hospital. Due to technical limitations, a combination of lack of international standard technical equipment, reagents and other conditions in most community hospitals across China (which are the single largest health care provider in China) and patient notion that muscle biopsy is an invasive examination and ensuing aversion, muscle biopsy is not the method of choice for diagnosis of PM in China. Therefore, PM diagnosis is still based on the Bohan-Peter criteria[1]: (1) Symmetric proximal muscle weakness determined by physical examination; (2) elevation of serum skeletal muscle enzymes, particularly CK; (3) EMG results indicating myopathy and (4) muscle biopsy indicating inflammatory myopathy.

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To be included in the study, patients had to meet the criteria for definite PM (meet all 4 criteria) or probable PM (meet any 3 criteria) and respond to corticosteroid therapy. Probable PM patients with nonspecific muscle fiber necrosis, concomitant cancers or those currently using methotrexate or azathioprine were excluded.

Data Collection

A thorough medical evaluation was performed at admission to collect demographic data, medical history, laboratory, clinical and electrophysiological data. Disease duration was calculated from the onset of the muscle weakness to the EMG. Muscle enzyme data were obtained from the most recent test before the EMG. Motor function data were obtained from the most recent examination of biceps brachii and quadriceps femoris before the EMG. Interstitial lung disease (ILD) was diagnosed with x-ray or computed tomography plus a pulmonary function test. Considering that corticosteroid therapy may affect the results of EMG, the patients were divided into groups receiving hormone therapy (≥30 mg/d corticosteroids for more than 7 days within 3 months before EMG, resulting in no significant improvement of the symptoms) and the group not receiving hormone therapy. After admission, once the patients were diagnosed with PM, all 93 were given standard corticosteroid therapy. Biceps brachii, abductor pollicis brevis, quadriceps femoris and tibialis anterior muscles were used for EMG data collection. Insert potential, spontaneous potential and MUP were measured. Nerve conduction velocity was also assessed when a patient's EMG showed neurogenic impairment.

Statistical Analysis

Qualitative data were analyzed using Pearson's χ^2 tests, and quantitative data were analyzed with Student's t tests. Linear regression was used to analyze associations between disease duration and test values. Differences were considered statistically significant at $P < 0.05$.

RESULTS

Demographic Results

A total of 93 patients (24 male, 69 female) met the inclusion criteria, including 32 with definite PM and 61 with probable PM. The average age at admission was 43.19 ± 15.64 years, and disease duration was 19.16 ± 33.12 months with a median of 6 months.

Overall, 36 (38.7%) received corticosteroid therapy before admission, 11 of which were definite PM and 25 probable PM. With regard to comorbidities, 18 (19.4%) patients had ILD, 17 (18.3%) patients had connective tissue disease (CTD) and 1 (1.1%) patient had non-Hodgkin's lymphoma. Among 17 CTD cases, 5 (29.4%) patients had rheumatoid arthritis, 3 (17.6%) patients had systemic sclerosis, 3 (17.6%) patients had Sjögren's syndrome, 2 (11.8%) patients had Behcet's syndrome and 1 (5.9%) patient had psoriasis. Only 40 (43.0%) patients submitted to muscle biopsy, of which 32 patients showed evidence of typical PM, 7 patients exhibited nonspecific muscle fiber necrosis and 1 patient was normal.

Electrophysiological and Clinical Test Results

The EMG data obtained from biceps brachii, abductor pollicis brevis, quadriceps femoris and tibialis anterior of 93 patients are summarized in Table 1. Decreased MUP duration >20%, fibrillation potential and positive sharp waves were found in most muscles (79.94%, 68.80% and 66.85%, respectively). Lower distal muscle (tibialis anterior muscle) exhibited significantly more abnormal EMGs than the upper distal muscle (abductor pollicis brevis), and the upper proximal muscle (biceps brachii) had significantly more abnormal EMGs than the upper distal muscle (abductor pollicis brevis). There was no difference in abnormal EMG findings between biceps brachii and quadriceps femoris, biceps brachii and tibialis anterior, abductor pollicis brevis and quadriceps femoris or biceps brachii and tibialis anterior.

All patients had CK measured, whereas alanine aminotransferase (ALT), aspartate aminotransferase (AST), lactate dehydrogenase (LDH) and erythrocyte sedimentation rate (ESR) were measured in 67, 67, 78 and 40 patients, respectively. Among them, 83.87% had increased CK, 88.06% had increased ALT, 85.07% had increased AST, 97.33% had increased LDH and 65% had increased ESR.

We next determined whether disease duration affected any of the assessed parameters. We found that patients who had experienced PM symptoms for longer than 3 months had significantly decreased MUP amplitudes in the biceps brachii (399.73 ± 81.71) than patients with shorter duration (403.80 ± 36.59) ($P = 0.025$). These patients also had significantly higher ALT levels than those with shorter PM duration (Table 2). Linear regression revealed a positive association between disease duration and MUP amplitude reduction of the biceps brachii (Table 3). Among 93 patients, 74 had ILD, with an average age of 49.16 ± 15.10 and PM duration of 9.02 ± 2.85 months. Non-ILD patients had an average age of 41.67 ± 15.51 years and PM duration of 21.77 ± 4.21 months, which was significantly longer than that of ILD patients ($P < 0.05$). Non-ILD patients also exhibited significantly higher CK and LDH levels compared with ILD patients (Table 4). In addition, non-ILD patients showed more abnormal EMG results than ILD patients (Figure 1 and Table 5).

Seventeen patients (18.28%) had definite CTDs, 1 had cancer, 3 had suspected CTD and 72 (77.42%) had PM without concomitant disease. There were no significant differences in muscle enzymes, ESR or EMG results between definite CTD

TABLE 1. Limb muscle EMG results of patients with PM, n(%)

EMG parameter	Biceps brachii (n = 93)	Abductor pollicis brevis (n = 80)	Quadriceps femoris (n = 93)	Tibialis anterior (n = 93)
Prolonged insert potential	19 (20.43)	4 (5.00)	16 (17.20)	18 (18.35)
Fibrillation potential	66 (70.97)	46 (57.50)	59 (63.44)	76 (81.72)
Positive sharp wave	66 (70.97)	39 (48.75)	59 (63.44)	69 (74.19)
MUP short duration	85 (91.40)	43 (53.75)	81 (87.10)	78 (83.87)
Polyphasic MUPs increase	6 (6.45)	15 (18.75)	18 (19.35)	13 (13.98)
Myopathic interference pattern	7 (7.53)	11 (13.75)	10 (10.75)	7 (7.53)

EMG, electromyogram; PM, polymyositis; MUP, motor unit potential.