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Original Article

Deviation in the recovery of the lower limb and respiratory muscles of patients with polymyositis: a preliminary clinical study

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Abstract. [Purpose] The purposes of this study were to quantify the serial changes in lower limb and respiratory muscle strengths and to evaluate the acute effects of physiotherapy in polymyositis patients. [Subjects and Methods] Five patients (57.6 ± 9.0 years, 50 to 72; four females) received physiotherapy five days a week for four weeks. The lower limb and respiratory muscle strength, the % vital capacity, and the Barthel index were evaluated at baseline and after the intervention. [Results] The patient's symptoms and creatine kinase values did not change, and after four weeks, all of the patients exhibited significantly increased outcomes compared with the baseline. However, the inspiratory muscle strength of the patients presented smaller improvements than the expiratory muscle strength. [Conclusion] Differential changes in inspiratory and expiratory muscle strength were observed following physiotherapy, and an unbalanced muscle distribution may explain the pathological and therapeutic effects. **Key words:** Polymyositis, Respiratory muscle strength, Rehabilitation

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INTRODUCTION

Polymyositis (PM) is an idiopathic inflammatory myopathy with a prevalence of 10–13 cases per 100,000 people in Japan¹). In PM, muscle inflammation is frequently accompanied by structural changes and loss of muscle fibers. PM patients also exhibit weakness in their proximal muscles²). Respiratory symptoms, such as interstitial pneumonia, are frequently observed in patients with dermatomyositis³). Therefore, physiotherapy may improve the limb muscle strength and activities of daily living (ADL) of PM patients, and according to previous studies, physiotherapy improves the lower limb muscle strength and physical function of PM patients^{4–6}).

Patients with myositis who require percutaneous endoscopic gastrostomy have a high mortality rate⁷). Dysphagia is an important issue for PM patients, and the frequency of dysphagia is approximately 12–62% among PM patients^{8–10}). In addition, respiratory muscle weakness can cause dysphagia in PM patients. Thus, respiratory disorders may be associated with an increased risk of dysphagia in PM patients. Previous studies have not evaluated the respiratory muscle strength of PM patients^{4–6}). Therefore, the aims of this study were to quantify the serial changes in lower limb and respiratory muscle strengths, and to evaluate the effectiveness of physiotherapy accompanied by prednisolone treatment in PM patients.

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SUBJECTS AND METHODS

Five PM patients (57.6 ± 9.0 years of age, range 50 to 72 years, four females) who had initiated physiotherapy within two weeks of PM diagnosis were admitted to Hiroshima University Hospital for treatment between April 2010 and March 2014. PM was diagnosed according to the clinicopathological analyses established in a previous report²⁾. All subjects received physiotherapy combined with prednisolone treatment for four weeks. Table 1 shows the main features of the patients. This study was approved by Hiroshima University's Committee of Ethics in Research (No 1153), and all of the subjects signed an informed consent form prior to enrollment.

The knee extensor muscle strength was evaluated using a hand-held dynamometer (HHD; Anima Corp., Tokyo, Japan). The subjects were seated, and their hip and knee joints were flexed at 90 degrees. The HHD was then attached to the distal lower extremity. The subjects ramped up to maximum force for each of the five actions over a 3-second period, then maintained a static position for approximately 5 seconds, during which the maximum force was measured¹¹.

The respiratory function was evaluated as the maximal inspiratory mouth pressure (PImax), maximal expiratory mouth pressure (PEmax), and %VC using a spirometer (Autospiro, Minato Corp., Osaka, Japan). These parameters were measured according to the American Thoracic Society/European Respiratory Society statement on respiratory muscle testing¹²). The PImax and PEmax values were calculated as percentages of the predicted values according to gender, age, weight, and height.

Physiotherapy was performed five days per week for 40 minutes. The daily physical training was conducted by a physiotherapist. First, each patient was requested to continuously repeat simple isotonic movements (i.e., shoulder flexion, hip flexion, and knee extension) until they felt fatigued. The number of repetitions completed by each patient was counted weekly. During daily training, the patients reached 60–80% of their repetition maximum. Then, each patient received respiratory training. The patients placed a 500-g weight on their abdomen and performed abdominal breathing against the weight until they felt fatigued. Each exercise was performed without exceeding 17 on the 15-point Borg scale¹³.

Statistical analyses were performed using IBM SPSS version 22.0 (SPSS, Inc., Chicago, IL, USA). The data are presented as the means \pm standard deviation (SD) or medians (minimum-maximum). The paired t-test or the Mann-Whitney U test was performed to test the significance of differences between the values observed at baseline and those obtained after four weeks of treatment. HHD evaluations and spirometer analyses were performed five times at each time point. The mean values of the five measurements were used in the analyses. Significance was accepted for values of p<0.05.

RESULTS

All five patients completed the exercise program and underwent prednisolone treatment. The patients' laboratory results (i.e., creatine kinase values) did not change during the intervention (Fig. 1). After four weeks of intervention, all patients exhibited significantly increased knee extensor muscle strength and PImax, PEmax, %VC, and Barthel index values compared

Case	Gender	Age	BMI	Medication
		(years)	(kg/m^2)	
1	Female	55	19.4	Prednisolone 0.9 mg/kg/day
2	Female	61	23.5	Prednisolone 0.49 mg/kg/day
3	Female	60	16.4	Prednisolone 0.26 mg/kg/day
4	Male	72	24.6	Prednisolone 0.4 mg/kg/day
5	Female	51	28.1	Prednisolone 0.6 mg/kg/day

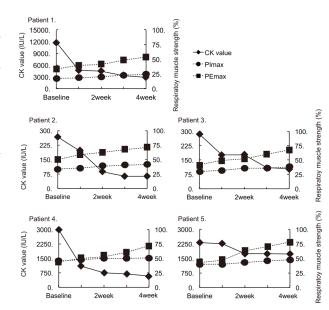


Fig. 1. Clinical course of each patient

Creatine kinase levels and respiratory muscle strengths of the five patients during the observation period.

Table 1. Patient characteristics and medications

with the baseline (p<0.001, Table 2). Differences in recovery with respect to improvements in expiratory and inspiratory muscle strength were also found (p<0.001, Table 2). The inspiratory muscle strength of the patients showed little improvement compared with their expiratory muscle strengths.

DISCUSSION

In the present study, the serial changes in the knee extensor muscle strength, respiratory function, and ADL of PM patients were evaluated. Physiotherapy accompanied by prednisolone treatment for four weeks did not exacerbate the patients' symptoms. The treatment also showed different recovery ratios between the expiratory and inspiratory muscles.

Performing intensive physiotherapy during the acute disease phase was previously thought to lead to further functional losses¹⁴). However, none of the patients included in the present study experienced exacerbated symptoms, and their creatine kinase values remained the same during the intervention period. This result supports previous findings regarding the safety of acute-phase rehabilitation^{5, 15}). The present study showed that PM symptoms are not exacerbated by physiotherapy when it is performed cautiously, based on the patient's degree of subjective fatigue, muscle pain, and creatine kinase values.

The knee extensor muscle strength, PEmax, PImax, %VC, and Barthel index significantly increased (Table 1). However, the %VC and PImax values did not improve satisfactorily compared to the knee extensor muscle strength and PEmax. Previous studies have reported improvements in knee extensor muscle strength and physical function in PM patients after physiotherapy, as evaluated via a functional index for myositis^{4–6, 16)}. However, these studies did not evaluate the respiratory muscles. Notably, previous studies showed that the respiratory muscle strength of patients with chronic obstructive pulmonary disease was improved by physiotherapy, and that these improvements in inspiratory and expiratory muscle strength did not differ significantly^{17, 18)}. Our results indicate there is a difference between inspiratory and expiratory muscle strength improvements among PM patients. The recovery of inspiratory muscle strength was lower than that of expiratory muscle strength. The present results may explain some of the characteristics of muscle dysfunction experienced by PM patients. PM patients show diaphragmatic degeneration and atrophy^{19, 20)}. Teixeira et al. reported a more than 75% prevalence of diaphragm dysfunction in patients with PM, dermatomyositis, and inclusion body myositis²⁰⁾. Therefore, myositis of the diaphragm may reduce the recovery potential of inspiratory muscle strength.

Our study had some limitations. First, this study was performed at a single center with a small sample. The prevalence of polymyositis is rare. A multi-center trial using standardized physiotherapy may be warranted. Second, our physiotherapy intervention was performed for only four weeks (five days per week). It is thus necessary to evaluate the chronic effects of physiotherapy in PM patients. Third, this study did not include a control group without prednisolone treatment or physiotherapy to determine the treatment-dependent effects. However, the inclusion of a control group may be unethical.

In conclusion, the present study demonstrated the safety of physiotherapy for PM patients. Different levels of improvement were observed between inspiratory and expiratory muscle strengths following physiotherapy. This difference may be due to the pathology of PM. Larger studies and mechanistic studies are imperative to determine the related pathology and therapeutic effects.

	Baseline (n=5)	After intervention (n=5)	Change (%)
Respiratory function			
PEmax (%)	42.5 ± 6.0	$68.6\pm9.0*$	$61.9\pm14.0\dagger$
PImax (%)	33.1 ± 10.5	$40.3\pm10.4*$	23.9 ± 9.5
% VC (%)	58.7 ± 7.0	$62.6\pm5.4*$	6.9 ± 4.0
Lower limb muscle strength			
Right knee extension (N)	75.9 ± 12.8	$149.6 \pm 37.5*$	97.3 ± 37.5
Left knee extension (N)	82.8 ± 14.9	$159.8\pm24.0\texttt{*}$	94.2 ± 13.4
Barthel Index score	75 (50-80)	95 (85-100)*	33.3 (25-70)
CK value (IU/L)	$3,735.2 \pm 3,106.1$	$1,254 \pm 1,104.0$	-60.8 ± 18.4

Table 2. Mean value of respiratory function,	muscle strength,	Barthel Index,	and CK	value in pat	ients with PM
baseline and after intervention					

The data are presented as the means \pm standard deviations or medians (minimum-maximum) for each outcome. Change (%) = change in mean values.

*p<0.001 compared with baseline

†p<0.001 compared with PImax

PEmax: maximal expiratory mouth pressures; PImax: maximal inspiratory mouth pressures; %VC: % vital capacity; CK: creatine kinase

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