Case Report

Intensive exercise and a patient in acute phase of polymyositis

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Abstract. Background and objective: Polymyositis (PM) is an idiopathic inflammatory myopathy manifested by proximal limb muscles weakness, elevated creatinin kinase, electromyography changes, and muscle inflammation in biopsy. We report an instance of intensive rehabilitation therapy in a patient with clinically active polymyositis.

Case report: A 19-year-old female patient, diagnosed with ‘electromyography and biopsy proven’ polymyositis for 5 years, suffered from worsening limbs weakness and dysphagia. In her history, she had upper and lower limbs weakness accompanied by dysphagia which was further complicated by right bronchial aspiration 9 months ago. A four-week trial of intensive training and exercise rehabilitation, concurrently accompanied by medications was prescribed for this patient. At the end of therapy she achieved significant improvement in muscle strength, activities of daily living, and ambulation without any disease exacerbation.

Conclusion: We concluded that short-term intensive training and exercise may lead to improvements in patients with PM, without causing a progress in the disease. Due to the rarity of PM and difficulty of conducting well-controlled studies to examine the risks and benefits of exercise in these patients, further research is necessary to investigate benefits of exercise training in active phase of disease.

Keywords: Polymyositis, idiopathic inflammatory myopathy, electromyography, rehabilitation, creatinin kinase

1. Introduction

The inflammatory myopathies are a heterogeneous group of subacute, chronic or acute acquired diseases of skeletal muscle. Polymyositis (PM) is an idiopathic inflammatory myopathy manifested by proximal and neck flexor muscle weakness, elevated creatinin kinase(CK), myopathic and neuropathic changes including fibrillations and positive waves, as well as muscle biopsy evidence of inflammation [1]. The effects of the disease vary in severity, ranging from loss of fine hand movements, inability to perform gross movements such as walking to dysphagia and respiratory muscle weakness.

Long-term treatment by effective anti-inflammatory medication using glucocorticoids alone or combined with cytotoxic agents is specified in active phases. Until the recent decades, patients with inflammatory myopathies were advised to avoid active exercise training and physical activity as it was assumed that such activities would exacerbate muscle inflammation [2–4]. However, re-establishment of muscle strength or maintenance of what remains of it by active rehabilitation therapy in inactive phases of disease with or without continuation of medication is a keystone in the continuing treatment of PM [5]. This paper reports an in-
stance of intensive rehabilitation therapy in a patient with well-documented clinically active polymyositis.

2. Case report

The case was a 19-year-old female patient, diagnosed with ‘electromyography (EMG) and biopsy proven’ polymyositis in 2004 suffering from painless, bilateral upper limb weakness with predominantly proximal muscle involvement and serum CK level of 11911. The weakness and CK level gradually improved with maximal response to high-dose steroids and azathioprine. However, azathioprine medication had to be stopped due to impaired liver function. In February 2009, she developed upper and lower limbs weakness accompanied by dysphagia and further complicated by right bronchial aspiration. She was prescribed prednisolone 50 mg/daily plus metotrexate 7.5 mg/weekly, and several investigations were planned but she defaulted further regular following up and taking medication. Nine months later again, the patient reported worsening limbs weaknesses and dysphagia. At this time, she remained independent in some basic activities of daily living (ADL) such as feeding, brushing and grooming but had difficulty to perform overhead activities. She required aid to sit, stand, walk, and climb upstairs. The ADL difficulties happened insidiously and progressed slowly.

On examination, hypotonia was observed in the patient’s shoulder bilaterally with some grades of subluxation. Muscle wasting was noted over both shoulder girdle and limitation in range of movement of right elbow. The weakness and CK level gradually improved with maximal response to high-dose steroids and azathioprine. However, azathioprine medication had to be stopped due to impaired liver function. In February 2009, she developed upper and lower limbs weakness accompanied by dysphagia and further complicated by right bronchial aspiration. She was prescribed prednisolone 50 mg/daily plus metotrexate 7.5 mg/weekly, and several investigations were planned but she defaulted further regular following up and taking medication. Nine months later again, the patient reported worsening limbs weaknesses and dysphagia. At this time, she remained independent in some basic activities of daily living (ADL) such as feeding, brushing and grooming but had difficulty to perform overhead activities. She required aid to sit, stand, walk, and climb upstairs. The ADL difficulties happened insidiously and progressed slowly.

On examination, hypotonia was observed in the patient’s shoulder bilaterally with some grades of subluxation. Muscle wasting was noted over both shoulder girdle and limitation in range of movement of right elbow (flexion: 70°, extension: 150°) due to skin tissue contracture. The patient’s muscles power details are summarized in Table 1. Deep tendon reflexes were absent at the upper limbs except the left triceps and supinator, and symmetrically reduced in lower limbs. Plantar responses were downgoing bilaterally. Sensory systems were intact. There were hyperpigmented skin patches in the chest, right arm and back with a small erythematous maculo-papular lesion in the right elbow. There was no muscle tenderness. Functionally, she could turn in bed independently and her ambulation under supervision was safer. She had good static sitting balance, fair dynamic sitting and static standing balance and poor dynamic standing balance.

She started prednisolone 50 mg/daily plus metotrexate 7.5 mg/weekly. Her thyroid function test and serum calcium were normal, and thoraco-abdominal CT-scan did not have any evidence of malignancy. The patient underwent a special rehabilitation program including physical and occupational therapy for five days per week in a four-week period simultaneously. The functional score Modified Barthel Index (MBI) for the patient was 65/100, implying moderate dependency. Her adherence and compliance to the prescribed program was excellent, though a couple of weeks after launching she felt fatigue and dizziness for a few days.

The physiotherapist (PT) set up a daily lower limb strengthening exercise, suspension sling exercise for bilateral hip flexors and abductors as well as gait training. A week later, she went through stepping exercise with low stool and ambulation training with an obstacle. The therapy continued with 15–20 minutes of daily cycling. The patient’s muscle strength improved (Table 1), and she was able to cover an unaided ambulation distance of 20 minutes after 2 weeks of therapy.

The occupational therapist (OT) initiated personal ADL retraining specially on personal hygiene, bathing self, upper and lower garment dressing. We then continued with upper limb functional exercise by strengthening and electrical stimulation of both shoulders. The patient was able to do a trial of ‘lift up seat’ for standing from a sitting position with minimal assistance. Finally, she had an MBI score of 82/100 with mild dependency and was able to do tasks independently.

In the end, she was discharged home with improvement in proximal muscle weakness and obvious decline in CK, erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) (Table 2). As a long-term treatment program, she was asked to cooperate with the rehabilitation team (PT/OT) as an outpatient for achieving further rehabilitation aims including stairs climbing, as well as personal and community ADL retraining.

3. Discussion

There is no single effective treatment for patients with PM. Strategies aimed to enhance muscle adaptations are particularly relevant to diseases characterized by skeletal muscle disorders. In this context, the treatment of PM seems to be increasingly dependent on a skilled multidisciplinary team.

Previously, it was believed performing exercise in the acute phase of the disease can lead to its deterioration. However, research findings indicate positive effects of active physical exercise on patients with inflammatory myopathies. Exercise training in rehabilitation of patients with active inflammatory stage on top of chronic phase would pose unique challenges [2]. A correlation
Table 1
Muscle strength according to Medical Research Council pre and post therapy

<table>
<thead>
<tr>
<th>Side</th>
<th>Shoulder</th>
<th>Elbow</th>
<th>Wrist</th>
<th>Fingers</th>
<th>Hip</th>
<th>Knee</th>
<th>Ankle</th>
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<td>G F E AB AD G F E AB AD G F E AB AD G F E AB AD</td>
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<td>Post</td>
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</table>

F = Flexion, E = Extension, AB = Abduction, AD = Adduction, G = Gripe.

Table 2
Laboratory results

<table>
<thead>
<tr>
<th></th>
<th>ESR</th>
<th>CRP</th>
<th>CK</th>
<th>AST</th>
<th>ALT</th>
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<tbody>
<tr>
<td>Pre therapy</td>
<td>24</td>
<td>0.7</td>
<td>11249</td>
<td>248</td>
<td>452</td>
</tr>
<tr>
<td>Post therapy</td>
<td>&lt; 0.4</td>
<td>1857</td>
<td>181</td>
<td>323</td>
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</tr>
</tbody>
</table>


between muscle strength and functional ability in patients with myositis has previously been demonstrated [6]. It was shown that early rehabilitation therapy with medication resulted in better outcomes [7]. Recent studies illustrated significant improvement in muscle function and strength in upper and lower limbs, walking distance, oxygen uptake relative to body weight, fitness and general health in patients with inflammatory myopathy as a result of physical training without any increase in disease activity [7,8].

The current report had novelty because of its intensive physical and occupational training program for a patient in the acute relapsing phase in combination with anti-inflammatory medications. In previous research such treatment was done in chronic and early recovery stages (2 to 3 weeks after the end of the acute symptoms) or in the stable and inactive phase of PM [5,7–9]. Kilmer et al. believed that use of resistance exercises in active inflammatory myopathy should be avoided until muscle enzymes and motor strength have stabilized [10]. However, our trial results after a short period of 4 weeks were comparable to those of recent controlled studies [7,9]. According to her weekly neuromuscular strength chart, the patient showed some improvement in proximal muscle power of upper and lower limbs; meanwhile, her distal muscles that are less seriously affected may have become significantly stronger, contributing to an overall improvement in ADL and ambulation.

Whereas her MBI upon admission indicated moderate dependency, she needed mild aid after discharge. The laboratory parameters (ESR, CRP and CK) considerably had declined; however, these parameters are not always related to disease activity or functional changes, necessitating quantitative assessment of muscle strength [5]. As no exacerbation was observed in the patient, it was concluded that no major risk was taken by starting physiotherapy in the acute relapse phase of PM. Training, however, must be carried out under medical supervision and must be adjusted to fit the needs of the patients [7].

In general, a short period of four-week therapy, fatigability and lack of exercises for strengthening of truncal muscles in the treatment program should be considered as some limitations for achieving our rehabilitation goals in this patient. However, related studies have been done in a 6-week randomized controlled program combining bicycle exercise and step aerobics [7]. Similarly, benefits were reported with a 6-month training program of the same regimen and a 30-minute home program 5 days per week for a 3-month period [11,12].

4. Conclusion

This study shows that short-term intensive training and exercise, applied as an individual tailored program, improved function in a patient with PM, without causing an increase in the disease activity. Because of the rarity of PM, it is hard to perform well-controlled studies to examine the risks and benefits of exercise in these patients. Further studies should search for other benefits (i.e., physiological, psychological and social) of exercise training in the active phase of the disease in these cases as well as the mechanisms underlying these adaptations.

References


