

Effectiveness and Safety of Physical Therapy Intervention in Adult Dermatomyositis: A Case Report

Isha S. Akulwar*

Department of Neuro Physiotherapy, K. J. Somaiya College of Physiotherapy, Mumbai, India.

*Corresponding author: Yohana Ayuba, Department of Neuro Physiotherapy, K. J. Somaiya College of Physiotherapy, Mumbai, India, Tel: 09769490996; E-mail: drishasa@yahoo.co.in

Received date: February 3, 2021; Accepted date: February 17, 2021; Published date: February 24, 2021

Citation: Akulwar IS (2021) Effectiveness and Safety of Physical Therapy Intervention in Adult Dermatomyositis: A Case Report. J Med Res Health Educ Vol. 5 No.2: 1.

Abstract

Background: The evidence for positive effects of exercise is insufficient in dermatomyositis. This article aims to outline the effects of physical exercise with regard to structure (disease activity, inflammation, metabolic milieu and muscle structure) as well as clinical outcome in terms of impairment, activity limitation, participation restriction according to the ICF; and QOL in an adult patient with dermatomyositis.

Case description: A 47 year old female diagnosed with definite DM since last 10 months was referred by a Rheumatologist for Physical therapy intervention.

Intervention: 12 weeks (three days a week) of Physiotherapy (moderate intensity, individualized, closely monitored) along with a structured home exercise program (low intensity, 5-6 days a week).

Outcome measures: Assessment of muscular pain, muscle strength, endurance, balance, functional evaluation, and QOL along with serological investigations.

Results: Effectiveness: Patient continued to show meaningful and clinically significant improvement on standardized outcome measures:

A major clinical response (total improvement score of 77) on IMAC S Core Set measures; Improvement across all domains of ICF viz. muscle strength, endurance; movement coordination; balance, aerobic endurance, confidence and performance in activities of daily living; participation in social activities; Probable disease-modifying effect: Progressively tapering dose of prescribed corticosteroids medication and improvement in disease-specific biochemical markers; Patient reported measures: Reduction in muscular pain, shortness of breath & fatigue; and improvement in Health-related QOL.

Conclusion: Moderate intensity of Physiotherapy intervention along with home exercise program is effective and safe in improving functional outcomes for dermatomyositis in a stable disease activity state. This case report enhances our understanding of the effects of physical exercise and may improve treatment and outcome in patients with dermatomyositis.

Keywords:

Dermatomyositis; Physical exercise; QOL

Abbreviations

IIM: Idiopathic Inflammatory Myopathies; DM: Dermatomyositis; PM: Polymyositis; WHO: World Health Organisation; ICF: International Classification of Functioning; QOL : Quality Of Life; MMT: Manual Muscle Testing

Introduction

The idiopathic inflammatory myopathies, collectively known as myositis are a heterogeneous group of systemic autoimmune diseases that primarily affect skeletal muscles. Traditionally, IIMs are classifiable in three subtypes, polymyositis, dermatomyositis, and inclusion body myositis, but recently, also other subgroups, such as necrotizing autoimmune myopathy and anti-synthetase syndrome have been identified. Dermatomyositis involves a characteristic skin rash and is characterized by reduced muscle strength, fatigue and myalgia. This is a rare condition with the estimated incidence of 1-9 cases per million people. Diagnosis is according to the EULAR/ACR criteria which combine clinical, laboratory, and pathologic features. It is coded in ICD-10 as M33.10 [1-5].

IIMs are considered as potentially treatable myopathies. Standard medical treatment consists of oral corticosteroids in combination with immunosuppressive treatment. Although responding to treatment, complete recovery of muscle function with pharmacological treatment does not always occur. Muscle strength, functional status and quality of life remain impaired and a majority of patients develop sustained disability, suggesting that physical inability is a great concern for these patients. In this context, Physical therapy is usually indicated to prevent muscle atrophy; and to regain muscle strength and range of motion. Over the last two decades studies evaluating active exercise in IIM support the notion of safety and benefits of exercise and indicate that active exercise, adapted to disease activity and disability should be included in the rehabilitation of patients in all stages of IIM. However, a Cochrane review in 2013 of five randomized controlled trials of exercise in muscle disease,

including one study with patients with polymyositis or Dermatomyositis, demonstrated that the evidence for positive effects of exercise is insufficient due to rarity of this disorder, its complex clinical phenotypes, and small number of trials included, their risk of bias and lack of control group. Although effectiveness of exercise on muscle functions have been reported, its efficacy on disability and quality of life has not been proved and also its efficacy in terms of newly developed and validated outcome measures has not yet been established. Many clinical trials are currently registered and recruiting participants, however these have combined DM and PM. DM and PM are clinically, histologically and pathogenically distinct entities. Associated symptoms and physical findings may vary widely from case to case as patients may present differently. There are no standardized therapeutic guidelines for treatment of IIM and the therapeutic approach is mainly guided by expert opinion and case series. More studies are required to establish the safety and benefits of different types of exercise. The aim of this case report is to outline the effects of physical exercise with regard to structure (disease activity, inflammation, metabolic milieu and muscle structure) as well as clinical outcome in terms of impairment, activity limitation, participation restriction according to the ICF; and QOL in an adult patient with DM [7-9].

Case Presentation

A 47 year old female diagnosed with DM since last 10 months (June 2018) was referred by a Rheumatologist for Physical therapy intervention. She reported with muscle weakness in the trunk, around pelvic girdle & hip region (difficulty in walking, climbing stairs, getting up from squatting position and a seated position from ground); myalgia (especially in the neck and trunk region); dyspnea, fatigue; and marked skin changes over trunk and face [10].

Medical history, investigations and treatment history was noted. Patient was having multiple systemic and other unspecific symptoms since last 7 years however she was never suspected and thus was never investigated for DM and was undiagnosed for several years. In June 2018, patient developed acute skin changes characteristic of DM, muscular manifestations. Other symptoms were dysphagia, shortness of breath and dry cough. Investigations for initial diagnosis essentially showed signs of muscle inflammation with elevated levels of muscle enzymes and myositis-specific autoantibodies on serologic evaluation. EMG investigation was normal and muscle biopsy was not done. A diagnosis of definite DM with myopathic subtype was then established for her. Also she was clinically diagnosed with an overlap syndrome- Sjogren syndrome (though specific blood test done during acute flare of symptoms was negative). Patient was immediately started on high dose corticosteroid medication (oral and in divided doses) and corticosteroid-sparing agent. She underwent serological investigations periodically for monitoring her disease activity and response to immunosuppressive and immune-modulating agents.

Physical Examination

Her higher mental functions, cranial nerve testing and sensory examination was unremarkable, Patient was evaluated using the International Myositis Assessment Clinical Study Group (IMACS) core set of disease measures as mentioned. The use of this validated measure has been recommended to assist in standardizing outcome measurement and in optimizing therapeutic trials in myositis. It includes assessments of disease activity; muscle strength; physical function; serum activity of muscle enzymes; and an assessment tool to capture extra-skeletal muscle disease activity. Disease activity in myositis refers to the type, extent and severity of reversible manifestations due to myositis¹¹. Disease damage is defined as persistent changes in anatomy, physiology, pathology or function, which are present for at least 6 months. Damage may be the result of prior active disease (causing scarring, fibrosis and atrophy), complications of therapy, comorbid conditions, or other events. A portion of disease damage is disease chronicity. Definition of improvement is given as any 3 of 6 core set measures improved $\geq 20\%$ with no more than 2 (not including MMT) worse by $\geq 25\%$. Also, the degree of change in each core set measure that is clinically meaningful is defined. For the domains of muscle strength and physical function, a minimum of 15% improvement is clinically significant, whereas for the physician and patient global assessments, as well as the extra-muscular assessment, a minimum of 20% improvement is considered clinically meaningful, and for serum levels of muscle associated enzymes, at least 30% improvement is needed to be clinically important. $\geq 20\%$ improvement is consensus of clinically meaningful change. The Total Improvement Score is the sum of all 6 improvement scores associated with the change in each core set measure.

Muscle strength is central to the assessment of myositis patients. It was assessed clinically using MMT. Patient presented with marked muscle atrophy and weakness in the axial and proximal muscles symmetrically, notably in the hip flexors, extensors, abductors, neck flexors and shoulder abductors. She had considerable difficulty in recruiting abdominals with poor sustenance (less time and weak hold). Movement control and whole body coordination was impaired in presence of significant muscle weakness. She demonstrated weak hip and trunk postural control strategies. Her Berg balance scale score was 53 (moderate risk of falls). She had no history of fall, however reported 'fear of fall'. She had self-imposed limitations on physical activities due to lack of confidence during movement transitions. 'Shivering', 'wobbling' was the subjective feeling reported by her during walking. She reported pain in thoracolumbar region which was not related to any particular movement or activity and occasional soreness in leg muscles. This symptom often used to wax and wane for no apparent reason. Dyspnea as reported on a point scale was 8 (on activity, sometimes even at rest).

Functional Evaluation

Patient was independent in bed mobility, transfers and in sit to stand but with difficulty. She could walk on overhead ground,

however preferred assistance for walking indoor and outdoor. Her functional mobility as assessed on Timed up and Go test was 14.2 seconds. For the staircase she required mechanical support of railing as well as manual assistance. More than actual weakness, her fear of fall was predominantly contributed with unpredictable shivering, wobbling of the legs. Despite of having some weakness in her upper limb muscles, she was independent for all upper limb activities. Patient experienced significant fatigue and shortness of breath which interfered with her ADL.

Disability:

On Modified Rankin scale (a global assessment of disability) patient graded her disability as (described as 'unable to attend to own bodily needs without assistance, and unable to walk unassisted') which is considered as Moderately Severe Disability.

Health-related QOL:

It was measured with the self-administered questionnaire SF-36, which contains questions relating to self-experienced health in eight dimensions: physical functioning, physical role, bodily pain, general health, vitality, social functioning, emotional role and mental health. Patient reported poor perceived health across all dimensions.

Psychological status:

Patient reported episodes of anxiety, feelings of self-helplessness and disability. Her concerns were associated with prolonged (almost for 7 years) physical and psychological suffering before the diagnosis and treatment; uncertainty of the disease course; life-long need for medication; dependency in ADL and significant functional disability; impaired quality of life; dealing with menopause, etc. Her medical condition seemed to be stable. Supervised exercise program (to closely monitor response to exercises and prevent muscular injuries and to exercise at an optimal level) on regular basis was recommended.

Intervention:

Patient received 12 weeks (three days a week on non-consecutive days) of Physiotherapy (individualized, closely monitored) along with a structured home exercise program (low intensity, 5-6 days a week). Exercise program started on a lower intensity, gradually increasing to goal intensity during the first two weeks. Gradually the intensity and duration of exercise was increased to a rate of exertion that the patient found somewhat challenging but not overly difficult. Physiotherapist reviewed her expectations, prognosis, and progress on a regular basis and attempted to set realistic goals based on continued discussion.

The physical therapy plan of care focused on strengthening and stretching, with an emphasis on a home exercise program. Types of exercises included: Strengthening; flexibility exercises; movement control and coordination training; Postural control and balance training; Aerobic endurance training; Thoracic mobility and breathing exercises; Task-specific functional training; Ergonomic advice and activity pacing. Her physiotherapy protocol included: Each exercise session started with 10 min warm-up, stretching. Exercises to improve muscle

strength and function targeted the trunk, pelvic girdle, hip and knee muscles and included active and resistance exercises adapted as per the degree of muscle weakness. These included progressive resistance training (moderate intensity) with therabands and also functional exercises (sit-to-stand training, reaching for weights on multi-level shelving). A detailed strength examination at every visit was done for assessing treatment response and strengthening program was adapted accordingly. Flexibility exercises included muscle lengthening, range of motion, and stretching with relaxation and breathing incorporated. In movement control and coordination training, large segment stability and mobility muscles were targeted. Postural control and balance training was given in various functional upright positions to improve postural awareness and postural strategies (both anticipatory and reactive). Task-specific functional exercises included sit to stand training and gait training on level surfaces and also stair- case training was started later. In view of negative effects of corticosteroid medication on bone health, low-impact weight-bearing exercises were incorporated. Thoracic mobility and Breathing exercises were practiced intermittently in upright positions with various upper limb movements. Aerobic endurance conditioning exercises started with Initial three weeks of lower extremity stationary cycling on a bike (on day one- terminated only after 1.11 seconds due to shortness of breath, later progressed to 10 min of continued cycling). Treadmill training was started after three weeks and at a minimum speed of 0.8 km/hour for 5 min, progressively 2-5 min were added subsequently for every session and progressed to 20 min of continued walking at a speed of 4 km/hour. Each therapy session ended with stretching of trained muscle groups and rating of overall exertion during the exercise session on the Borg RPE-scale. Owing to her limited endurance for exercises- multiple goals were incorporated into the same exercise regime viz. strengthening in functional patterns involving multiple segments and in upright positions thus simultaneously improving movement coordination and balance.

A home exercise program was designed by incorporating similar principles as that of the supervised therapy program. The patient performed exercises at home on an easy-to moderate intensity adapted to initial muscle weakness, five days a week for 12 weeks. The program consisted of stretching exercises; breathing exercises; strengthening exercises for the trunk and pelvic girdle muscles; seated dynamic knee-extensor exercise; biceps and deltoid curls, seated rowing, heel and toe lifts, whole-body rising from sitting to standing; kneel walking, etc. The program was divided into two/more sessions with adequate rest in-between and took about 30 minutes to perform. She filled out an exercise diary commenting on loads and number of repetitions for each task. Progressively during the course of therapy patient was advised for outdoor walks of 20 minutes.

Statistical Analysis

Findings, tolerance and response to Physiotherapy was monitored on daily basis and changes were made accordingly. Before exercise session patient used to rate self-reported pain and fatigue on a visual analogue scale and describe in her own

words information about possible changes in medication dose, any side effects from medication as well as possible negative and positive effects from previous exercise sessions, as well as physical activities performed since the previous exercise session. She filled out an exercise diary for HEP and her physical activity log. Borg 6-20 RPE scale was used every day to monitor the intensity of exercise program. Thus, exercise was individually adapted to disease activity, medication dose and especially to the level of pain and fatigue. Patient showed good compliance to supervised therapy program and HEP. Ergonomic advice was given for ADLs and advice on activity pacing to reduce fatigue and improve endurance. She was advised for some environmental modifications at home for reducing functional limitations such as use of grab bars for the bath, shower, and raised toilet seats.

Patient was instructed that during acute flare up, exercises to be discontinued or reduced to gentle isometrics and ROM exercises. Avoid additional exercises and physical activity was restricted to daily functional mobility and walking. Also, for HEP she was told not to exercise on any given day if experiencing fatigue or pain from the previous day. Advice was given on dietary consultation.

Results

Patient continued to show meaningful and clinically significant improvement on standardized outcome measures over a period of 12 weeks.

Effects of Codeine on Body and Ovarian Weight and Ovary to Body Weight Ratio

The changes in the mean body weights of animals treated with codeine and their control are presented in Table 1. There was an increase of 27.80g in mean body weight of rats of the control group. In the group administered 30mg/kg of codeine there was an increase of 27.00g. In the group administered 60mg/kg the mean body weight increased by 20.20g while in the group administered 90mg/kg codeine the mean body weight increased by 23.60g. The increased in weight reduced in the treated groups when compared to the control group.

The result was not significant ($p < 0.05$) in the group treated groups. Ovarian to body weight ratio was significant in the groups that received 60mg/kg and 90mg/kg codeine when compared to the control group at $p < 0.05$.

ICF Domain	Parameter	Outcome
Body structure and Function	Muscle strength	Improved grade on MMT
	Muscle endurance	Increased sustenance-(time and strength of hold)
	Motor coordination	Better initiation and sustenance of movements and better multi-segment control
	Postural control and	Improved reactive strategies and BBS

	balance	score reached 56 (maximum score)
	Aerobic capacity	Increased cycling and treadmill time and other intensity related parameters
Activity level	Functional abilities	Ease, quick, less physical assistance, better confidence to move and without provoking pain or fatigue after the activity
	Functional mobility	TUGT score 60 % improvement
Participation level	Social participation	started going out of the home to visit community places (10 times in 12 weeks as compared to 7 months of complete home-bound isolation), every time with less direct manual assistance, could confidently face crowded places, and it did not provoke any of the disease symptoms.
		For her ritual prayers she could not sit down in kneeling position. After intervention she could sit more comfortably and ease of getting after wards made her concentrate more peacefully during prayers.

Table 1: Patient showed improvement across all ICF domains

Patient-reported outcomes:

Pain in thoraco-lumbar and pelvic region subsided completely.

Improvement across all components of Health-related Quality Of Life(SF -36). Sleep patterns and quality of sleep improved. Commented that I feel now I am living in my body. Effect of Codeine on some selected reproductive hormones in female rats. Estrogen, progesterone, follicle stimulating hormone and luteinizing hormone blood levels in the female treated groups did not show much variation when compared to control.

Discussion

Dermatomyositis is an inflammatory disabling neuromuscular disorder. The main goals when treating patients with DM are to improve function and prevent disability. This manuscript reviews the effectiveness and safety of an active rehabilitation program on all measures of disease activity, damage and patient-reported outcomes in a patient with adult dermatomyositis. Use of core set measures has been documented in recent studies^{15, 16}. It needs close collaboration between the physician, Physical therapist and the patient in disease assessment. Patient was a responder to medical and Physical therapy intervention according to the International Myositis Assessment and Clinical Studies Group definition of improvement. Patient met the

IMACS improvement criteria for the entire core set measures and showed a major clinical response with 12 weeks of Physiotherapy intervention. Regarding effectiveness, muscle strength and MD Global activity are considered as top measures of improvement. It is important to mention that patient showed considerable improvement even in the more affected muscle groups. Resistive exercises given in this study were of moderate intensity and repetition for both supervised therapy program and HEP. Our findings on improvement in muscle function are in line with previous studies demonstrating effectiveness of resistive muscular training program in patients with chronic DM/PM

Conclusion

Based on the previous research and from the results of this case presented here, it can be concluded that physical exercise is safe and effective in adult dermatomyositis. We suggest that physical exercise, adapted to disease activity and disability should be included in the rehabilitation of in all stages of dermatomyositis.

References

1. Semet M, Paci M, Magnan JS, Metzler-Guillemain C, Boissier R, et al. (2007) The impact of drug abuse on male fertility: a review. *Andrology* 5: 640-663.
2. Abs R, Verhelst J, Maeyaert J, Van Buyten JP, Opsomer F, et al. (2000) Endocrine consequences of long-term intrathecal administration of opioids. *J Clin Endocrinol Metab* 85: 2215-2222.
3. Aurilio C, Bachiocco V, Biasi G, Fiorenzani P, Pace MC, et al. (2009) Endocrine consequences of opioid therapy. *Psychoneuroendocrinology* 34: 162-168.
4. He Z, Qiu J, Li J, Zhao D, Chen G, et al. (2013) Long-term effects of conversion from cyclosporine to rapamycin on testicular function and morphology in a rat transplantation model. *Transplant Proc* 45: 763-769.
5. Derry S, Karlin SM, Moore RA (2015) Single dose oral ibuprofen plus codeine for acute postoperative pain in adults. *Cochrane Database Syst Rev*.
6. Hout MCV (2014) Doctor shopping and pharmacy hopping: practice innovations relating to codeine. *Drugs Alcohol Today* 14: 219-234.
7. Nielsen S, Hout MCV (2016) Over-the-counter codeine from therapeutic use to dependence, and the grey areas in between. *Curr Top Behav Neurosci* 34: 59-75.
8. Robinson G, Robinson S, McCarthy P, Cameron C (2010) Misuse of over-the-counter codeine-containing analgesics: dependence and other adverse effects. *N Z Med J* 123: 59-64.
9. Nigeria's deadly codeine cough syrup epidemic. BBC Africa's new investigations unit, Africa Eye (2018). British Broadcasting Commission.
10. Nigeria bans codeine cough syrup to stop an addiction epidemic. But something worse could take its place (2018). Washington post.