



Case Report

Ayurvedic management in limb girdle muscular dystrophy – A case report



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ABSTRACT

Limb girdle muscular dystrophy (LGMD) is a type of Muscular dystrophy (MD), heterogeneous devastating complex genetic disorders causing progressive weakness and degeneration of muscles. LGMD is hereditary autosomal diseases characterized by weak and wasteful limb girdle muscles. The available management of LGMD in biomedicine is unsatisfactory. Here we present a case of LGMD managed with combinations of Ayurvedic oral medicines and Panchakarma procedures. The Ayurvedic diagnosis of the condition was considered as *Mansagata Vata* (~neuromuscular diseases), a type of *Vatavyadhi* (~neuromusculo skeleton disorders). The patient was treated with *Shalishastika Pinda Swedana* and *Mustadi Yapana Basti* for the duration of 16 days along with following Ayurvedic oral medicines: *Yograj Guggulu* 500 mg with 40 ml *Dashamoola Kwatha*, *Ekangaveera Rasa* 125 mg with honey, a combination of *Ashwagandha Churna* -2g, *Satavari Churna* - 2g, and *Sankha Bhasma* 500 mg with milk, *Narsinha Churna*- 3g and *Ashwagandhavleha*- 5g with milk. All medicines were given twice a day. Patient's condition was assessed for symptoms of pain, walking distance, power and reflexes of both upper and lower limb and pseudohypertrophy of both calf muscles. Serum Creatine Phosphokinase (S.CPK) level and electromyography (EMG) were also measured. There was symptomatic improvement in the patient's condition and reduction in S.CPK level. The study suggests that LGMD can be satisfactorily managed with Ayurvedic oral medicines and Panchakarma therapy.

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1. Introduction

Muscular dystrophies (MDs) are genetically heterogeneous cluster of muscle disorders with progressive weakness and muscle wasting. Duchene muscular dystrophy (DMD), Becker muscular dystrophy (BMD), congenital muscular dystrophy (CMD), Myotonic muscular dystrophy (MMD), Fascioscapulohumeral muscular dystrophy (FSMD) and Limb girdle muscular dystrophy (LGMD) are common MDs [1]. Each of these disorders differs in terms of age of onset, severity, affecting muscle groups and pattern of inheritance [2]. LGMDs are autosomal, heterogeneous neuromuscular disorders of progressive weakness and wasting of the limb girdle muscles [3]. The combined prevalence rate of the MDs range between 19.8–25.1 per 100,000 people per year [4]. Worldwide prevalence rate of LGMDs are 1 in 14,500–45,000 [3]. MD patients are known to have

mortality at younger age and there is lack of effective management [5]. There is need to explore alternative options for MDs managements. The present case is first attempted study in Pub Med indexed journals till date to explore Ayurvedic management in MDs with satisfactory outcome. The primary focus of the management was to ameliorate the symptoms of the disease. The Ayurvedic diagnosis was made as *Mansagata Vata* (~neuromuscular diseases), a type of *Nirupstambhita* (~disease due to unobstructed *Vata Dasha*) *Vatavyadhi* (~neuromuscular skeleton disorders) [6].

2. Case report

A 35 years old married female patient visited the Panchakarma outpatient department of National Institute of Ayurveda (N.I.A.) on December 4, 2018 with complaints of lower back ache, pain in bilateral knee joints and difficulty in walking and performing routine household activities since past 5 years. Patient was not able to walk more than 10–20 m on flat surface. There was gradual weakening and heaviness (like stones tied to body) in both upper and lower limbs. There was difficulty in getting up from squatting

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or sitting position, climbing up stairs, lifting heavy objects and doing overhead activities like combing or shampooing hair since last 9 years. The patient had consulted on 12/01/2016 in neurology department of Swai Man Singh medical college and hospital, Jaipur and was diagnosed as LGMD – sarcoglycopathy. She was advised conservative treatment and genetic analysis. Patient was not satisfied with the treatment and hence approached N.I.A. hospital.

2.1. Clinical finding

Patient was conscious, well oriented, and spoke normally. There was absence of pallor, icterus, clubbing, cyanosis and edema. She had difficulty in walking (waddling gait) and positive Gower's sign. Neurological examination revealed normal Cranial nerve functions, the tone of upper and lower extremities was reduced and grade 4 power was observed in bilateral shoulder joint, knee joint and in both upper and lower limbs. Bilateral Achilles tendon reflexes were diminished. The sensory functions like pain, touch and temperature were normal. Pseudohypertrophy was observed in bilateral calf muscles. Erythrocyte sedimentation rate, Complete blood count and thyroid profile were within normal limits. Serum Creatine Phosphokinase (S. CPK) was elevated. Electromyography dated 12/01/2016 (EMG) shows the myopathic pattern. Tenfold examination of patient suggested *Vatakapha Prakriti* (~physical constitution) *Madhyama Samhanana* (~medium body constitution), *Avara Sara* (~lowermost purest body tissue), *Avara Vyayam Shakti* (~lowermost capability to carry on physical activities), *Madhyama Abhyavara* and *Jaranashakti* (~middlemost food intake and digestive power). *Mansavaha Srotodusti* was observed (Vitiation in micro-channels related to muscular tissues) in this patient. *Srotasa Pariksha* (examinations of other microchannels) revealed normal status of other *Srotasas*.

3. Timeline (Table 1)

Disease course, management and outcomes.

4. Diagnostic focus and assessment

The patient was a known case of LGMD (sarcoglycopathy type). The types of muscular dystrophy i.e. DMD, BMD, CMD and FSMMD were the potential diagnosis for the case. The age of onset excluded DMD, BMD, and CMD, while normal functioning of cranial nerves excluded FSMMD. Heaviness of the body, severe body ache similar to *Dandamushthihata* (body has been beaten with a cudgel or fist cuffs) and extreme exhaustion are the symptoms of *Mansagata Vata* [6] which were exhibited in patient. *Mansakshaya* or *Shosh* (depletion of muscular tissues) was the differential diagnosis of the disease. Specific part of body involved in pathology ruled out the condition of *Mansakshaya*. Other *Vatavyadhis* such as *Snayugata Vata* and *Mansagata Vata* which can lead to depletion of muscular tissues were also the potential diagnosis [6 verse 35–36]. But other symptoms of these *Vatavyadhis* were absent in the case. Systemic examination of other systems and *Srotasas Pariksha* were normal except *Mansavaha Srotodusti* hence these were ruled out. *Swayathu* (edema/inflammation), *Mansa Shosh* (depletion of muscular tissues, fibrosis of muscles fibers) are the common symptoms due to damages of *Mansavaha Srotasas* (micro-channels related to muscular tissues) [7]. Difficulties in climbing stairs, walking and rising from the floor also indicate *Mansavaha Srotodusti* as there is disruption of the function of *Mansa Dhatu* (~nutrients related to muscular tissues) which is transferring through these channels. Thus Ayurvedic diagnosis was established as *Mansagata Vata*, a *Nirupsthambhita Vatavyadhi*. As the chronicity of disease is more than one year it was considered as *Yapya Vyadhi* (palliative disease).

5. Treatment plan

The management was focused on providing symptomatic relief. The general management of *Vatavyadhi* was adopted. *Shalishashtika Pinda Swedana* [which provided both *Snehana* (oleation) and *Swedana* (perspiration)] and *Basti Karma* (medicated enema) were planned with oral medications. Details of Ayurvedic medications and *Panchakarma* are summarized in Tables 2 and 3.

6. Outcome measures and follow up

Patient was assessed for Pain, walking ability, power and reflexes of both upper and lower limbs, pseudohypertrophy of calf muscles, S.CPK level and EMG at baseline and endline (completion of *Panchakarma* procedures). Lower back region and bilateral knee joints pain were at grade 5 on visual analogue scale on 4/12/18 it reduced to 3 on 17/4/19 and further reduced to 0 on 13/09/2019. Walking ability on flat surface assessed by 6 min walk test was improved from 10 to 20 m on 04/12/2018, 500 m on 17/04/2019 and 800 m on 13/09/2019. Additionally mild improvement was experienced by patient in waddling gait. Measurements of calf muscles pseudohypertrophy were 54.56 cm of right leg and 52.8 cm of left leg on 04/12/2018 which reduced to 51 cm of right leg and 49 cm of left leg on 17/04/2019 and further reduced to 42 cm in right leg and 41 cm in left leg on 13/09/2019. The power was at grade 4 on medical research council scale on 04/12/2018 which improved to grade 5 on 17/04/2019 and on 13/09/2019 in bilateral shoulder joint, knee joint and in both upper and lower limb. Bilateral Achilles tendon reflex were 2+ on 17/04/2019 and on 13/09/2019. Patient reported that there was no heaviness in the body and she could perform the routine house work with ease. Mild difficulty in climbing up stairs and lifting heavy objects was experienced by patient on 13/09/2019. The S. CPK and EMG were reassessed after the therapy. The value of S. CPK level was reduced from 1450 IU/L on 11/01/2016 to 909 IU/L on 30/05/2019. EMG didn't show any changes after the therapy. Other blood investigations were within normal limits [Supplementary Table 1].

7. Discussion

The main pathology in LGMD is gene mutations causing autosomal, heterogeneous hereditary disorder. In *Ayurveda* there is concept of *Aadibala Pravrita Vyadhi* (~hereditary disorders) caused by *Vata Dosh* [7, Sutra Sthan chapter 24 verse 6]. *Vata* plays prime role in the organogenesis, tissue formation and systemic maturation during fetal development [8] and on vitiation through *Swaprakopaka Karana* (i.e. *Beeja Dosh*), *Dhatukshaya* (~depletion of body tissue) and *Margavarana* (obstruction in the pathway of *Vata*) causes *Kubja* (hunch backed), *Pangu* (crippled), *Muka* (dumb) etc. *Vatavyadhis*. [7, chapter 2 verse 54]. In present case vitiating *Vata Dosh* must have focused on *Mansa* in fetal life and developed *Mansagata Vata*. *Ekanga Swayathu* (Pseudohypertrophy) in calf muscles was developed due to *Prakupita* (vitiating) *Doshas* and mainly *Vata Dosh* which affected the *Twak* (skin and subcutaneous tissues), *Mansa* (muscles), *Shonita* (blood), etc. and lead to *Swayathu*. [8, chapter 18 verse 6/1]. At the time of first encounter on 04/12/2018 *Sthanika Shleshka Kapha* depletion (localized depletion of *Kapha Dosh*) symptoms and vitiating of *Vata Dosh* at lumbar and knee region, which probably caused knee joint pain and lower back ache. In present case line of treatment for *Dhatukshaya* *Vatavyadhi* of chronic nature was planned. As the patient was suffering with *Mandagni* (suboptimum digestive power), oral Ayurveda medicine was given before *Panchakarma* to digest the *Ama* (~undigested food that vitiating the *Doshas*). The line of treatment of *Vatavyadhi* comprises of *Snehana*, *Swedana*, *Mriduvirechana* (~mild purgation)

Table 1
Timeline.

Year	Incidence/Intervention
2009	Progressive weakness of both lower limb causing difficulty in getting up from squatting or sitting position and difficulty in climbing stairs.
2012	Progressive weakness in both upper limbs causing difficulty in lifting heavy objects, doing overhead activities such as combing and shampooing hair. Patient also noticed prominence of both calf muscles.
2014	Patient suffered from lower back ache, pain in bilateral knee joints and difficulty in walking and activities related to routine house work.
12/01/2016	Patient consulted in neurology department of Swai Man Singh medical college and hospital, Jaipur for the complaints. Serum CPK was value was 1450 IU/L and EMG suggested of myopathic pattern. Patient was diagnosed as LGMD (sarcoglycopathy). She was advised conservative treatment and genetic analysis. Only multi-vitamins were prescribed to the patient.
04/12/2018	Patient was unsatisfied with the management hence approached Panchakarma outpatient department of National Institute of Ayurveda, Jaipur for the complaints of lower back ache, pain in bilateral knee joints and difficulty in walking and household activities since past 5 years. Lower back region and bilateral knee joints pain was grade 5 on visual analogue scale (VAS). She had difficulty in walking (waddling gait) and Positive Gower's sign. Measurements of calf muscles (pseudohypertrophy) were 54.6 cm in right leg and 52.8 cm in left leg. Walking distance on flat surface assessed by 6 min walk test (6MWT) was 10–20 m. The power was at grade 4 on medical research council scale (MRC) in bilateral shoulder joint, knee joint and in both upper and lower limb. Bilateral Achilles tendon reflex were diminished. The Ayurvedic diagnosis of the condition was considered as <i>Mansagata Vata</i> , a type of <i>Vatavyadhi</i> . The management was focused on providing symptomatic relief. The general management of <i>Vatavyadhi</i> was adopted. <i>Shalishastika Pinda Swedana</i> and <i>Basti Karma</i> were planned with oral medications.
04/12/2018	Selected Ayurvedic oral medications were – <i>Yograj Guggulu</i> 500 mg with 40 ml <i>Dashamoola Kwatha</i> , a combination of <i>Ashwagandha Churna</i> (powder of <i>Withaniasomnifera</i> Dunal)-2g, <i>Satavari Churna</i> (powder of <i>Asparagus racemosus</i> WILD.)- 2g, and <i>Sankha Bhasma</i> (calx of conch shell)- 500 mg with milk, <i>Narsinha Churna</i> 3 g and <i>Ashwagandhavleha</i> - 5g with milk. All medicines were prescribed twice a day.
10/12/2018–26/12/2019	<i>Shalishastika Pinda Swedana</i> and <i>Mustadi Yapana Basti</i> was carried out for 16 days
04/01/2019	<i>Narsinha Churna</i> was discontinued and rest of the medications were continued till date.
29/03/2019	<i>Ekangaveera Rasa</i> 125 mg with honey, twice a day was incorporated in daily regime and continued till date.
(2/4/19–17/04/2019)	<i>Shalishastika Pinda Swedana</i> with <i>Mustadi Yapana Basti</i> were repeated for 16 days
17/04/2019	Patient was assessed for pain, walking distance, power and reflexes of both upper and lower limb with measurement of calf region. Lower back region and bilateral knee joints pain were at grade 3 on VAS. Walking ability was improved to 500 m on 6MWT. Calf muscles measurements were reduced to 51 cm in right leg and 49 cm in left leg. The power was at grade 4 on MRC in bilateral shoulder joint, knee joint and in both upper and lower limb. Bilateral Achilles tendon reflex were 2+.
30/05/2019	The S. CPK and EMG assessments were repeated after the treatment. S. CPK level was decreased from 1450 IU/L on 11/01/2016 to 909 IU/L on 30/05/2019. No change in EMG was observed. Other blood investigations were within normal limits.
13/09/2019	Lower back and bilateral knee joints pain was reduced to 0 on VAS. Walking ability was improved to 800 m on 6MWT. Mild improvement in waddling gate was reported by patient herself. Calf muscles pseudohypertrophy was further reduced to 42 cm in right leg and 41 cm in left leg. The power was at grade 4 on MRC in bilateral shoulder joint, knee joint and in both upper and lower limbs. Patient reported that there was no heaviness in the body and she could perform the routine house work with ease. Mild difficulty in climbing up stairs and lifting heavy objects was reported by patient.

and *Basti* [6 Verse 75–84]. Likewise *Virechana*, *Niruha Basti* (Medicated enema mainly comprises of decoction of medicinal plant for *Vatavyadhi* etc) and *Shamana* (palliative) therapies are indicated in the management of *Mansagata Vata* [6 Verse 93]. For *Dhatukshaya*, the main treatment includes *Snehana*, *Swedana*, *Basti* along with Ayurvedic oral medications. In chronic *Vatavyadhis*, *Nanatamaj Vata* and for *Avrita Vata Yapana Basti*, *Guggulu*, *Rasayana* and *Shilajeeta* are indicated [6 Verse 240–241]. Thus following the textual line of management, *Shalishastika Pinda Swedana* and *Mustadi Yapana Basti* were planned along with oral medication. *Shalishastika Pinda Swedana* has the property of *Brunhana* because it is a kind of *Snigdha Swedana* (~provides nourishment to muscles and bones). *Mustadi Yapana Basti* is indicated for increasing strength, *Jeevana Shakti* (~vigor or quality life) and *Vrishya* (~fertility). It is useful in *Sopha* (~inflammation or pseudohypertrophy), in *Katishoola* (low back ache), pain in calf and thigh region and *Vatarakta* (~various disease of rheumatic spectrum). It also has

Rasayana properties. It can break the pathogenesis of the disease by removing obstruction and purification of channels and depletion of *Dhatu*s by its *Brunhana* nature. *Mustadi Yapana Basti* is able to control most muscle, bone and bone marrow diseases [6, *Siddhi Sthan* chapter 12 verse 15]. The oral medication -*Yograj Guggulu* is useful in *Aamvata* (rheumatic disorders), *Vatarakta*, various *Sandhi* – *Majjagata Vatika* disorders (~neurological, rheumatic and musculoskeletal and bone marrow diseases), it increases *Agni* (metabolic fire) and *Bala* (physical strength) [9]. *Dashamoola Kwatha* have *Tridoshaghana* properties (alleviate the deranged humors) and treats the depletion of tissues [10]. *Ashwagandha* [11] and *Satavari* [11, verse 291] both have *Rasayana* and *Balya* (anabolic) properties. *Sankha Bhasma* correct the *Agnimandhya* (improves the digestive fire), *Balya* (anabolic) in nature *Amlapitta Nashaka* (anti-hyperacidity) and alleviates the deranged *Vata Dosha* [12]. It is also useful in hyperacidity which may be caused in some patients due to use of *Narsinha Churna*. *Narsinha Churna* is useful in

Table 2
Oral Ayurveda medication used for the case of Limb girdle muscular dystrophy.

Name of the drug	Anupana	Days of treatment	Dose
<i>Yograj Guggulu</i>	Twice a day with <i>Dashamoola Kwatha</i>	4/12/18–7/10/2019	500 mg
<i>Dashamoola Kwatha</i>	Twice a day	4/12/18–7/10/2019	40 ml
combination of <i>Ashwagandha</i> and <i>Satavari Churna</i> with <i>Sankha Bhasma</i>	Twice a day with luke warm milk	4/12/18–7/10/2019	2g 2g 500 mg
<i>Narsinha Churna</i>	Twice a day with luke warm milk	4/12/18 to 4/1/19	3g
<i>Ashwagandhavleha</i>	Twice a day with luke warm milk	4/12/18–7/10/2019	5g
<i>Ekangaveera Rasa</i>	Twice a day with honey	29/03/19–7/10/2019	125 mg

Table 3
Panchakarma procedures used for the case of Limb girdle muscular dystrophy.

Panchakarma procedures with method of preparation	Method of application	Days of treatment
<i>Shalishashtika Pinda Swedana</i> - 250g <i>Shashtika Shali</i> is cooked in 1.5 L of decoction of <i>Bala Moola</i> and milk. Cooked mixture is made into bolus in cotton cloth. <i>Bala Moola</i> decoction and milk mixture is used as reheating medium for bolus	Whole body Massage with <i>Dashamoola</i> oil was done for 10 min followed by whole body massage for 45 min with bolus of <i>Shashtika Shali</i> . Then <i>Abhyanga</i> with <i>Dashmoola taila</i> was repeated for next 10 min.	16 days (10/12/18–26/12/18) 16 days (2/4/19–17/4/19)
<i>Mustadi Yapana Basti</i> - <i>Madhu</i> (honey) 50g, <i>Saindhava Lavana</i> (rock salt) 5g, <i>Panchatikta Ghrita</i> 50 ml, <i>Ashwagandha</i> oil 50 ml, <i>Satapushpa</i> (<i>Anethum sowa</i> Kurz) <i>Kalka</i> 15g, milk processed with <i>Mustadi Yapana Basti Kwatha</i> drugs 300 ml and <i>Mansarasa</i> (soup of goat meat) 50 ml were mixed in standard sequence for <i>Basti</i> preparation to form a homogenous emulsion which was indirectly heated on a water bath.	Given before meal by <i>Basti Yantra</i> Route of administration – anal region	16 days (10/12/18–26/12/18) (total 16 <i>Bastis</i> were given as <i>Yapana</i> formulation no separate <i>Anuvasana Basti</i> was needed for it), then again for 16 days (2/4/19–17/4/19)

low back pain, *Kshaya* (depletion of body) and all types of *Vatika*, *Pattika* and *Kaphaja Vikara* (–diseases due to derangements of all humors) [9, Chapter 67 verse 15–24]. It is also effective in *Mansakshaya* condition [13]. *Ekangaveera Rasa* is effective in *Vataja* disorders, *Vatakaphaja* disorders and *Pakshaghata* (Hemiplegia) [13, *Kharaliya Rasayana Prakarana*, p 470]. Repeated uses of *Snehana* and *Swedana* and other *Vatanashaka* (alleviating the *Vata Dosha*) process may lead to accumulation of *Malas* (waste metabolites) in *Srotas* and hinders the normal process of *Vata Dosha* [6, verse 85–86]. Here *Anulomana* (mild purgation) *Deepana* and *Pachana* (Drugs having the property to improve the digestion) and *Vatakapha Nashaka* (*Vatakapha Dosha* alleviating) drugs such as *Ekangaveera Rasa* are effective. *Ekangaveera Rasa* is a compound of *Kuchala* (*Strychnos nux-vomica* L.) which is useful in imparting the strength to muscles fibers [13, *Kharaliya Rasayana Prakarana*, p 408]. *Ekangaveera Rasa* is also having the property of *Brunhana* (Having the nourishing property to the tissues) and thus helpful in alleviating the degeneration of muscles. Hence the *Ayurvedic* line of management fulfills all the necessarily treatments used in the progressive diseases such as anti-inflammatory drugs, physical therapy and synthetic steroids which are often associated with side effects in long term use [14]. As this is a *Aadibalapravritta Vyadhi* (hereditary disease), this is *Yapya* (palliative) in nature. It can be managed by repairing the genes. *Panchakarma* procedures and *Rasayana* drugs may affect the epigenetic process and thus brings hopes of management in this disease by genetic alteration and expression in positive way [15,16]. It is indicated that basic nutrients and heavy metals can induce epigenetic process [17]. The present case findings are important because they indicate a better and safer treatment strategy with satisfactory outcome for a disease condition that requires lifelong supportive treatment.

8. Conclusion

This case study demonstrated the satisfactory management in LGMD with *Panchakarma* procedures and *Ayurvedic* medicinal interventions and provides foundation for future research studies.

9. Patient consent

Written informed consent for publication of this case study had been obtained from the patient.

10. Patient's perspective

Patient was satisfied with the provided treatment.

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None.

Conflict of interest

No conflict of interest.

Author contributions

Kshipra Rajoria: Methodology, investigation, resources, writing- original draft, review draft.

Sarvesh Kumar Singh: conceptualization, investigation, resources, writing original draft, review draft, supervision.

Suman Dadhich: writing original draft, investigations.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.jaim.2021.07.002>.

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