

CASE REPORT

Role of Pre-Biopurification Procedures in Conventional Management of Neurogenic Myopathy : A Case Study

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Abstract

Diseases in which the muscles do not function normally because there is something wrong with their nerve supply and diseases in which the nerve supply is normal but the muscles are unable to respond normally to the nerve impulses are described as neurogenic myopathy or neuromuscular diseases which generally produce weakening and atrophy of skeletal muscles. Worldwide incidence of inheritable myopathies is about 14%. Management of myopathy aims at the conservation of structure, activity and assistance in disability. There is no specific therapy for complete cure of neurogenic myopathy in modern system of medicine. The absence of specific treatment for complete cure for neurogenic myopathies makes it all more important to consider complementary and alternative medicines approaches for the treatment. Sign and symptoms of neurogenic myopathy are nearer to *Mamsavaha Srotodusti Vikara* (Disorders of Musculoskeletal origin). According to Ayurveda, *Mamsashosha* (reduction in muscle bulk) condition occur due to Doshic imbalance and changes in *Saptha Dhathus* (Seven tissues) which are the basic elements maintaining the body structure. In this case study, the Ayurvedic treatment involving the group of Herbomineral drugs, Physical support and specific *Purvakarma* (pre-biopurification) procedures. Patient was observed for symptomatic improvement based on assessment done by Vignous & Brooke Scale. Result has shown definite acceptable improvement even in severe form and longer survival upon neuromuscular diseases.

Keywords: Neurogenic myopathy; *Purvakarma*; Agni; Ayurveda

Introduction

Neurogenic myopathies are a group of muscular diseases that weaken the muscles which move the human body. Myopathies are characterized by progressive muscle weakness, death of muscle cells and tissue. These myopathies are caused by mutations, or changes, in genes the blueprints for making proteins that are necessary for our bodies to function correctly. In the inheritable myopathies, genetic mutations cause defects in various proteins necessary for muscle tone and contraction [1]. Symptoms of myopathies include mild weakness of voluntary muscles, especially in the hips and legs, hip displacement, delays in reaching developmental milestones, problems with running, jumping and climbing stairs & frequently develop contractures. This situation is one of the most complicated dilemmas faced by the orthopedics. The purpose of the management includes perpetuation of constitution, function and help from pain and disability. Ayurveda comes under AYUSH system of medicines which is about 5000 years old. Ayurveda also names three elemental bodily humors, the *doshas* (called Vata, Pitta and Kapha), and states that a balance of the *doshas* results in health, while imbalance results in disease. The Plan of management of this case is based on the principles of Ayurveda which includes pre-procedures of Panchkarma (Bio-purification) and internal herbal medicines.

Case History

An Eight years old boy of Gorakhpur region, reported to OPD of *Kaumarbhritya/Bal Roga* department of Sir Sunder Lal Hospital, Banaras Hindu University, Varanasi, Uttar Pradesh, India with complaints of unable to stand without support and unable to move with/without support according to age. There was no significant past history except typhoid and jaundice four years back which cured within 7-8 days without any complication. In family history, all members were healthy except grandfather who suffered from non-insulin dependent diabetes mellitus (NIDDM) and grandmother who suffered from thyroid disorders. In birth history, prolonged labor (Stage-II -8 hrs) with spontaneous vaginal delivery (SVD) with vertex presentation, delayed cry after birth

(according to mother), full term/Appropriate for gestational age (AGA), and birth weight- 2.865kg. In development history, all milestones were delayed. He consulted a physician in his local place Gorakhpur for relief from these complaints and advised muscle relaxant drugs and calcium supplement without any diagnosis and not get any relief from these medicines. Then he approached our Sir Sunder Lal Hospital for traditional and better management. The symptoms of the patient during 1st visit in our hospital were unable to stand properly without support, unable to move with/without support, and development of contractures in joints.

On General examination

Appetite was decreased and incomplete evacuation of bowel, 1-2 times/day, hard, yellowish without blood, mucous & foul smell while other parameters were within normal limits.

On Systemic examination

- **CVS, R/S and GIT**- No significant abnormality.
- **Central nervous system**- Patient was conscious and well oriented to time, place & person.
- **Musculoskeletal examination**

Muscle bulk (B/L)	Upper limb	Lower limb
Biceps	Atrophied	-
Deltoid	Atrophied	-
Trapezius	Atrophied	-
Calf	-	Atrophied
Soleus/Gastronemius	-	Atrophied
Muscle tone (B/L)	Hypertonic	Hypertonic

Table 1: Examination of muscle bulk/tone: All muscle bulk were normal except

Muscle power (B/L)	Upper limb	Lower limb
Biceps	Grade 2	-
Deltoid	Grade 2	-
Trapezius	Grade 2	-
Calf	-	Grade 2
Soleus/Gastronemius	-	Grade 2

Table 2: Examination of muscle power: All muscle power was in grade 4 except

Muscle DTR	Right side	Left side
Biceps	Increased	Increased
Knee	Increased	Increased
Ankle	Increased	Increased
Planter	Increased	Increased

Table 3: Examination of deep tendon reflexes (DTR): All DTR was normal except

Motor system

- Crepitus- absent
- Nutrition-moderate
- Involuntary movement-absent
- Contractures-present

CBC	CPK	CT SCAN/MRI(Head)
Hb-12.2 gm/dl	301.4 U/L	No Pathology seen , Within normal limits
TLC-7600/cumm		
DLC- N40 L47 E7 M6 B0		
Platelets-245000/micro lit		

Table 4: Investigation

Muscle Biopsy: Skeletal muscle biopsy show normal muscle fibers, small group of atrophic fibers of variable size are seen, admixed in between normal fibers. Sarcolemmal muscles are peripheral & inflammatory infiltrate not seen. The features suggestive of Neurogenic Myopathy.

Plan of Management

In view of the history and assessment of the patient, management was designed according to involvement of *Dosha* (Fundamental bodily elements) and *Dushya* (Bodily toxins). In Ayurveda there is the theory of *Agni* (digestive fire) which maintains the cellular metabolism which in turn nourishes the *Dhathus* (Tissues) which are *Rasa*, *Raktha*, *Mamsa*, *Meda*, *Asthi*, *Majja* and *Shukra*, which builds the body and each *Dhatu* (tissue), is having its digestive fire [2]. Any inequity or vitiation in this fire will be leading to reprehensible absorption of nutrients leading to the condition of weakness. In this case there is an increase in *Mamsa Dhatu Agni* (muscle Fire), which will be leading to its destruction and in turn reduction in muscle bulk and increase in *Medas* (fat) which is the next *Dhatu* (tissue) after *Mamsa* leading to a reduction in muscle power [3]. Vitals of this case were monitored daily pre and post Purvakarma (Pre-biopurification) therapy by the multichannel pulse oximeter. *Shirobasti* (A type of Snehana of Head having cap full with medicated oil and sit for 30 minutes) with *Maha Narayana*⁴ oil and *Jyotishmat* (*Celastrus paniculatus*) oil, followed by *Katibasti* (A type of Snehana of Waist and Back having cap full with medicated oil) with *Maha Narayana* oil and *Bala* (*Sida cordifolia*) oil for 25-30 minutes. After that *Sarvanga Snehana* (body massage) with *Maha Narayana* oil and *Bala* (*Sida cordifolia*) oil⁶ followed by Modified *Shastik Shali Pinda Sweda* (A type of Brhmana Swedan which gives nutrition to the tissues which are undergoing depletion and degeneration [5-7]. It is a time tested treatment administered to those ailing from musculoskeletal and neuromuscular diseases) using *Shastik* rice cooked in decoction of *Dashmool* for 20-25 minutes, then *Avgahana* (*Tub Bath with medicated decoction*) with *Dashmool* for 35-40 minutes according to the strength and lastly *Shiropichu* (A type of Snehana of Head having cotton or gauze piece soaked with medicated oil) with *Jyotismati* (*Celastrus paniculatus*) oil and *Bala* (*Sida cordifolia*) oil for 2.5-3 hrs. This Procedure was scheduled for 21 days [5,6,8].

Oral medication

The drugs were used along with the Purvakarma:

- *Bitter Vachamool* (*Acorus calamus* rhizome) rub 60 times on stone & Almond¹⁶ 4 in number in liquid form with milk and given with honey two times per day before meal [9].
- Germinated *Methika* seeds (*Trigonella foenum*) 3 gm in morning before food [10].
- Cap Evion (Vit. E) 1 cap on alternate day [11].
- *Praval Bhasma* (100 mg) divided into two parts two times per day after food [12].
- *Agnitundi Vati* (*Kupilu*): ½ tab two times per day after food [13].
- *Guduchi* (*Tinospora cardifolia*) *Ksheer Pak* 50 ml two times per day after food [14].

Pathya (compatible food) was advised with *Nidan Parivarjana* (avoiding all aggravating factors) along with light diet , occupational therapy, physical therapy, social support and counseling of parents. Patient was observed for complication during whole course of treatment.

Observation

The result of this case is based on the grading of different musculoskeletal functions according to Brooke scale and Vignous scale.

Joints, Spine and Angle Test

Grade 0 (Absent), 1 (Present), 2 (Severe)

- **Joint contractures:** Before Purva karma therapy, the grade for all joint contractures was 2 but after therapy it reduced to grade 1.
- **Straight leg raising test & Femoral ante version:** Reduced from grade 2 to grade 1.
- **Spine changes (Scoliosis & Kyphosis):** Reduced from grade 2 to grade 1.
- Before Purva karma, patient requires hand for support in sitting but after therapy, trunk rounded occasional use of hands for support.

Upper/Lower Extremities Muscle Strength

Grade 0 (Absent), 1 (Definite weakness), 2 (Severe Weakness), 9 (cannot evaluate)

- **Hand grip:** Reduced from grade 2 to grade 0.
- **Biceps/Brachioradialis/Pronator teres/Shoulder extensors:** Reduced from grade 2 to grade 1.
- **Hip adductors/Angle dorsiflexion:** Reduced from grade 2 to grade 1.
- **Rectus femoris/Ham strings/Gastrocnemius/Clonus:** Reduced from grade 1 to grade 0.
- **Walk on Heel/Walk on toes:** Reduced from grade 9 to grade 2.
- **One leg standing:** No improvement i.e. grade 9.

Upper Extremities Function

Active range of motion- Grade 0 (Full), 1 (limited), 2 (Severely limited)

- **Elbow/Wrist extension/Supination/Pincer grasp:** Reduced from grade 2 to grade 1.

- **Thumb abduction/Thumb extension/Finger extension/Release of Block:** Reduced from grade 1 to grade 0.
- Before therapy, left deviation of thumb was present with no voluntary control but after therapy, significant reduction in deviation of thumb was noted.

Standing/Walking with/without Braces

Observation of gait: Grade 0 (not present), 1 (definite), 2 (Severe)

- **Pelvic obliquity/Hip adduction/Standing balance:** Reduced from grade 2 to grade 1.
- **Crouch/In & out toeing/Equinus:** Reduced from grade 1 to grade 0.
- Before therapy, Patient could not stand straight for long time even with support but after therapy, patient can stand for long time with support.

Ambulatory Status

Before therapy, patient crawls and uses wheelchair for Non-independent mobility but after therapy, walks with braces or other aids, no wheelchair required.

The Brooke scale for grading upper extremity function: Before therapy, patient cannot raise hands to the mouth and has no useful function of hands but after therapy patient can raise arms above the head only by flexing the elbow or using accessory muscle

The Vignous scale for grading lower extremity function: Before therapy, patient Stands in long leg braces but unable to walk even with assistance but after therapy, patient walks only with assistance or walks independently with long leg braces

Discussion

Ayurvedic *Acharyas* cautiously believe this condition as *Adibala-Pravrit Mamsa-Vata-Kshaya* due to *Srotorodha*. There is diminution of *Mamsagni* paving the way of Ama development [15]. It is followed by vitiation of *Kapha Dosha*. This complex pathogenesis is accountable for progressive wasting and necrosis of the affected muscle fibers.

The main line of management is to correct the *Dhathu*, *Agni* and increase the muscle bulk. This is done with special internal medications and external treatment modalities which increase the vascularity and hence increasing the nourishment. It's a long term process with a strict regime arresting the muscle destruction and rejuvenating the myofibers. This patient of neurogenic myopathy was treated with Local *Abhyanga* (*Shiro/Kati Basti*), whole body massage (*Sarvanaga Snehan*) followed by *Brihman Swedana* (*Shalishasti Pinda Sweda*) and *Avagahana* line of treatment. *Abhyanga* (Local/Whole body) is an already established effective management as proper massage because this procedure decreases the release of the cortisone stress hormone and leading to the relaxation of muscle [16,17]. The lipid soluble substances are absorbed through skin, they can penetrate the skin, and after reaching the skin they create a weak electrical stimulation which enhances the absorption creating the blood circulation faster [18].

The combined effect of *Abhyanga* and *Shashtishali Pinda Sweda* as well as *Avagahana* is beneficial to patient as it increases blood circulation to muscles, provides nutrition to muscles, Increases strength of muscles, decreases spasticity of muscles and contractures of joint, improvement of power of muscles and reassurance application done in proper way can help in reduction of motor neuron hyper-excitability by reducing the alpha motor neuron activity [19]. The oils used in various procedures contain many herbs used as neuro-muscular tonic because of their *Vata* balancing properties.

Acorus Calamus and Almond is herbal tonic for the central nervous system [20]. Pandey, *et al.* evaluated the methanol and acetone extract of *Acorus calamus* (Vacha) for their CNS activity in mice. They showed the spontaneous locomotors activity for immobility by time using through forced timed swim test, diazepam induced sleeping time and motor impairment assessment using rotarod for CNS depression/analytic activity of ACME and ACAE in mice while Kulkarni, *et al.* in their study, suggests that almonds possess a memory enhancing activity in view of its facilitatory effect on the retention of special memory in scopolamine induced amnesia [21]. They concluded that almonds lowered the serum cholesterol in rats. They were also found to elevate the Ach level in the brain and ultimately improve the memory of rats [20]. Vyas S, *et al.* evaluated the analgesic and anti-inflammatory activities of *Trigonella foenum-graecum* (seed) extract due to presence of natural steroids. Analgesic and anti-inflammatory effects were examined in a partially purified fraction (MTH) of the *Trigonella foenum-graecum* seed extract [10].

Praval being a natural source of rich calcium. Calcined coral is widely used in Ayurvedic medicine as a supplement in the treatment of variety of bone metabolic disorders associated with calcium deficiency. *Praval Bhasma* is effective in the prevention of calcium deficient spinal contractures and bone deformities [12].

Agnitundi Vati contains Strychnine which stimulates all parts of the CNS and particularly the anterior horn cells of spinal cord causing greatly increased reflex excitability [22].

Vitamin E is the major lipid-soluble component in the cell antioxidant defense system. It has been found that alpha-tocopherol mainly inhibits the production of new free radicals, while gamma-tocopherol traps and neutralizes the existing free radicals. Oxidation has been linked to numerous possible conditions including: cancer, arthritis, muscle degeneration and cataracts. Thus, vitamin E might help prevent or delay the chronic diseases associated with reactive oxygen species molecules [11].

Guduchi (*Tinospora Cordifolia*) have been used in general debility, digestive disturbances, and loss of appetite in children. It is used to improve immune system and body resistance to infections [14].

Conclusion

The treatment modalities adopted for this case has shown significant improvement in term of contractures, hand grip, muscle strength, tone & ambulatory status. The therapy is cost effective and need further, longitudinal and large sample size study for confirmatory statistical result with the adopted management for treatment of neurogenic myopathies

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