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A Case Study on Ayurvedic Management of Spinal Muscular Atrophy (SMA)

Case Report

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Abstract

Spinal Muscular Atrophy (SMA) is the second leading genetic disorder inherited in autosomal recessive pattern due to absence of SMN1 gene characterized by loss of motor neurons and progressive muscle wasting, often leading to dependent life and decreased life span. In Ayurveda, SMA can be considered as a type of *janma jaata Vata vikara* as it has been mentioned that: "*Tatra va gati gandhanayoriti vata"* that means all the movements of the body are controlled by *vata.* In *Vata vyadhi Lakshanas*, few symptoms like *Anganam sosha* (Atrophy or emaciation of limbs), *Sankocha* (Contraction), *Kanja, Pangulya, Kubjatva* (Lameness of hands and feet, hunch-back and shortness), are considered, few of which are also observed in the Spinal Muscular Atrophy. A 2yrs female patient was admitted in our I.P.D who was a known case of SMA II presented with complaints of inability in sitting for longer time without support, unable to stand and walk even with support. Through Ayurvedic principles we have treated adopting various vata hara treatment modalities & also with few palliative treatments as per the need in view of enhancing the quality living.

Keywords: Neuromuscular disease, SMA, Genetic mutation, Vata vyadhi.

Introduction

It is human outlook to dream for begetting a healthy & flawless offspring but it is unfortunate to have increasing incidences of unhealthy pediatric population with irremediable major neurological disabilities. The management of such ailments has become rather confusing to the present day clinicians and apparent remedies if present are highly not sure & expensive.

Spinal muscular atrophy is an autosomal recessive neurodegenerative disease characterized by degeneration of alpha motor neurons in the spinal cord, atrophy of skeletal muscles, resulting in progressive proximal muscle weakness and paralysis.

The Neuromuscular disease is a very broad term that encompasses many diseases and ailments that impair the functioning of the muscles, either directly, being pathologies of the voluntary muscle, or indirectly, being pathologies of nerves or neuromuscular junctions. The Spinal muscular atrophy is a rare neuromuscular disorder characterized by loss of motor neurons and progressive muscle wasting, often leading to early death. It is an autosomal recessive pattern of inheritance which is linked to a genetic mutation in the *SMN1* gene.

It is caused by homozygous disruption of the survival motor neuron 1 (*SMN1*) gene by deletion, conversion, or mutation. The most common form of SMA is caused by a deficiency of a motor neuron protein called *SMN*, for survival of motor neuron (chromosome 5 SMA, or SMN-related SMA). For normal motor neuron functioning this protein is necessary. Its deficiency is caused by genetic defects (mutations) on chromosome 5 in a gene called *SMN1*. Neighboring *SMN2* genes can in part compensate for non-functional SMN1 genes. In age of onset, symptoms and rate of progression there is wide variability. In order to account for these differences, the chromosome 5 SMA often is classified into types 1 to 4. Approximately the degree to which motor function is affected depends on the age at which the onset of SMA symptoms occur. The earlier the age of onset, the greater the effect on motor function. Children who present symptoms at birth or in infancy typically have the lowest level of functioning (type 1). SMA onset in children (types 2 and 3), teens or adults (type 4) generally relates with increasingly higher levels of motor function. Other rare forms of SMA (non-chromosome 5) are caused by mutations in genes other than *SMN* (1).

SMA is the second most common serious autosomal recessive disorder after cystic fibrosis, with an estimated incidence of 1 in 6,000 to 1 in 10,000 live births, with a carrier frequency of 1/40-1/60 (2, 3). Although no medical treatment is available, investigations have elucidated possible mechanisms underlying the molecular pathogenesis of the disease[1].

If SMA affects in an infant under 2 years of age, it is usually fatal. If the onset of disease is at an older age, the children will have a different type of SMA, and life expectancy may be normal.

Kaumarabhritya is the specialty department that deals with child care in Ayurveda. One to one correlation of SMA to a single disorder in Ayurveda is

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not possible. But basing on the etiopathogenesis, and symptomatology it can be considered as conditions like *Udanavrita prana vayu vikara, janma jata chestavaha sroto vikara, Janma jata dhatugata vata vikara, Rakta, Asthi, Majja gata vata, beeja gata margavaroda janyavata vyadhi* under the conglomeration of *Vata vyadhi* (*Neurological disorder)* that have an overlap of symptoms that of SMA. Contributory factors like inappropriate *ritu* (ovulation cycle), *kshetra* (uterus), *ambu* (amniotic fluid), *beeja* (sperm and ovum)(10), *dauhridavamana*(11) (neglect of urges during *dauhruda* state of pregnant woman), Presence of *garbhopaghatakara bhavas* (12) (activities or substances which are not favorable for growth and survival of fetus) , incompatible *garbha vriddikara* bhavas (13)(requisites for growth and development of fetus) (14) and improper *garbhini paricharya* (antenatal regimen) may have unwanted effects on the fetus impeding its normal growth and development subsequently leading to many diseases, deformities and even death. Hence a formulated Ayurvedic therapy protocol can improve the condition of SMA patients providing a quality life minimizing their dependency.

Case Report:

Basic information of the patient

A 2 yrs female child patient was admitted in our I.P.D who was a known case of SMA II presented with complaints of inability in sitting for longer time without support, unable to stand and walk even with support. She was a Muslim by religion and was born to a middle class, literate, non-consanguineous parents.

Chief complaints (*Pradhana vedana visesha*)

Regression of motor milestones, Generalized weakness, floppiness, unable to hold the neck completely, can sit only with curved back with the support of upper limbs and cannot stand even with support since the age of 7 months.

History of present illness (*Vartamana Vyadhi Vrittanta*)

Child is 2nd offspring, delivered through full term normal vaginal passage, born to non-consanguineous couple with less spacing, no other significant pre-natal, natal & post-natal birth history noted. Child had attained social smile, neck holding, sitting, proning appropriately as per chronological age and was apparently normal till the age of 7 months. Suddenly she developed severe vomiting & motions and was managed conservatively in the local hospital. Gradually child started developing progressive loss of muscle strength & decreased motor activity in neck, trunk & all the limbs leading to significant regression of motor mile stones.

History of past illness (*Purva vyadhi vrittnata*)

No significant history.

Treatment history (*Chikitsa vrittanta*)

Physiotherapy for 6 months from 11 months of age after which mild improvement of neck holding & upper limb power was seen. Took Unani treatment for 3 months and then came to S.V.Ay.Hospital-Kaumarabhritya O.P.D. with a ray of hope for better relief.

Family history (*Kulaja vrittanta*)

No family history or consanguinity found

Birth history

Antenatal: Except for less spacing i...e. First child was 3 months when the present child pregnancy was confirmed.

Natal: Full term normal vaginal delivery, Birth weight – 2.5 kg

Post-natal history: Insignificant

History of Immunization: Immunized as per schedule

Personal history (*Vaiyaktika vrittanta*)

Ahara- Child Was breast fed exclusively till 6 months of age, there after milk, cerelac, rice and egg white till 11 months later on good amount of solids.

Table- I Developmental History

Neck holding	3 months
Prone	3 months
Sitting with support	4 months
Sitting without support	6 months
Standing with support	Not Attained
Standing without support	Not Attained
Walking with support	Not Attained
Walking without support	Not Attained
Dentition	7 months
Speech	1 year

Examination

Vitals were normal. No abnormality was found in cardiovascular system, respiratory system, gastrointestinal system and per abdomen. *Pitta Kapha prakriti* (Constitution)

Rogi Pariksha

Table II-*Dashavida Pariksha*

Prakriti:	*Pitta Kapha*
Vikriti	*Vata pradhana tridoshaja*
Sara	*Twak*
Samhanana	*Madhyama*
Desha	*Jangala*
Satmya	*Sarva Rasa*
Satva	*Madhyama*
Ahara sakti	*Madhyama*
Vyayama Shakti	*Avara*
Vayah	*Kumara*

Central Nervous system examination

Child was alert, conscious coherent and well oriented with intact sensory functions.

Motor power was absent completely in lower limbs when compared to trunk and upper limbs. Areflexia was seen in both upper and lower limbs. Deep tendon reflexes were absent. Muscle tone decreased in neck & both upper and lower limbs. The muscle power in lower limbs was 0/5, and upper limbs was 2/5. Child was unable to turn neck in prone position.

Investigations

MRI Lumbar spine finding was normal

Molecular Genetic screening: Homozygous deletion of exon 7 on SMN1 gene

Differential Diagnosis

Acute flaccid paralysis
DMD
Poliomyelitis
Gullian Barre syndrome
Amyelotropic lateral sclerosis

Final Diagnosis

Diagnosis was confirmed as Spinal muscular atrophy by differential diagnosis, and genetic testing.

Analysis of signs and symptoms- Presentation

- **Loss of Motor functions** - Only **motor neurons** are lost thus the sensory functions are Retained.
- **Floppiness** – weak muscle tone almost like a floppy baby
- Difficulty in **sitting/standing/walking**, and regressed or absent milestones,
- **Areflexia** -The symmetrical **atrophy of the proximal muscles** (than distal muscles of the legs, arms sometimes the trunk, in disease progress
- **Muscle Atonia** -Loss of strength of the respiratory muscles.
- **Bell-shaped torso** (when only abdominal muscles used for respiration)
- Difficulty sucking or swallowing, **poor feeding**

Table IV- Probable Pathophysiology and its Management

Roga Prakriti	***Samprapti Ghataka***	***Samprapti Vighatana***
Dosha	*Vata Pradhana Kapha Pitta*	*Vasti*
Dooshya	*Mamsa, Medo and Majja*	*Abhyanga (*Massage*) and shashtika shali panda swedana (*nutritive sudation*)*
Agni	*Manda*	*Deepana and pachana drugs along with Udvartana.*
Srotas	*Mamsa, Medo, Majja, cheshtavaha srotas*	*Vatahara oushadi, Medhya dravyas and Madhura tikta anna.*
Sroto dusti type	*Sanga (Obstruction)*	*Sroto shodhana by Asthapana Vasti*
Vyaktasthana	*Sarvanga*	*Abhyantara- Vata hara aushadi, Balya dravyas, mamsa, majja, Prayoga, Sashtika Sali panda sweda*
Roga	*Spinal Muscular Atrophy (Chestavaha sroto gata vata vyadhi)*	*Vata vyadhi chikitsa Sootra (Snehana, svedana, Basti, and snehapana)*
Sadhya-sadhyata	*Yapya*	Long term and maintenance protocol

Treatment Protocol

Treatment regimen comprised of 14 days of abhyangam, Shastika Sali pinda swedam, vasti and 14 days of Meru vasti. Three such courses were done with an interval of 14 days.

Treatment given

1. ***Snehana*** -*Abhyangam* for 14 days with *Maha narayana tailam, Dhanwantharam tailam, ksheerabala tailam.*
2. **Swedana** -*Shashtika Shali Pinda Swedam* for 14 days.
3. **Vasti** _ *Madhu, saindhavam, satapushpa, gomutra Balamoola qwatha, Dashamoola qwatha, ashwagandha, rasna, mahanarayana taila, Ksheerabala taila* for 11 days.
4. **Meru vasti** – *Maha vishagarbha Tailam + Ksheerabala tailam* for 14 days
5. **Physiotherapy**

Internal Medication (*Abhyantara Aushada Prayoga)*

1. *Bruhat vata chintamani Ras* -½ tab three times in a day
2. Tab. *Yogaraja Guggulu*- ½ three times in a day
3. *Pravala bhasma 10mg+Trivanga bhasma 5mg+Yasti churnam 200mg three times in a day with honey*
4. *Bala Moola qwatha* 15ml two times a day

Pathya

Balya, Brimhana, Mamsa Rasa, shali dhanya, Yoosha.

Apathya

Ruksha, Asuchi, Abhishyandi, Vatakara ahara and vihara.

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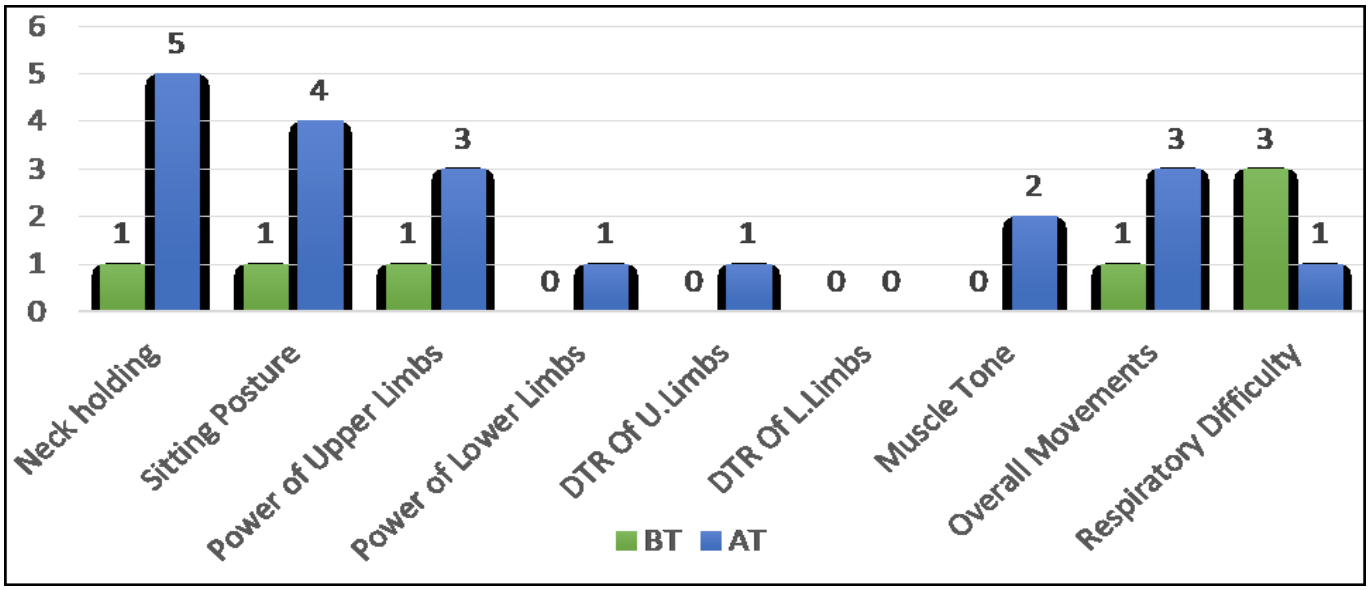
Table-V Progress after 3 sittings

S.No	Condition	Before Treatment	After treatment
1	Neck holding	Partial	Complete Able to extend neck and rotate side wards in prone position
2	Sitting posture	Rounded back	Straight back Spinal incurvation decreased
3	Power of Upper limbs	Unable to lift 1/5- visible movement, no joint movement	Able to lift 3/5- joint movement (+) Against gravity
4	Power of Lower limbs	0/5- no muscle movement	1/5- visible movement, no joint movement
5	Deep tendon reflexes of UL	Absent	Mildly present 1/5
6	Deep tendon reflexes of LL	Absent	Absent
7	Overall movements	Upper limbs, trunk are movable	Trying to move forward in sitting position
8	Respiratory difficulty	Frequent cold (on Nebulization)	No further episodes of cold noted (Nebulization not re-quired)

Image-I-Pictorial presentation of Symptoms



The symptoms were assessed by MRC scale for muscle power (23).

Discussion:

Understanding SMA –an Ayurvedic perspective / Insight

Diagnosis: SMA can be related to ***Udanavrita pranavata vyadhi* (22)**.

Prana vayu is occluded by udana vata then there will be loss of motor function, complexion, strength and ojas. It may also result in death. Margavarodha or sanga probably might be due to kapha.

As per modern science it is a genetic disorder in which the **motor neurons** in the **anterior horn of the spinal cord** are damaged subsequently motor functions (movements) are affected because of loss of alpha motor neurons. *Vata* being responsible for both sensory and motor functions (*sarva hi cheshtah vatena sa pranah praninam smritah)* in the body. In this condition the task of vata is affected due to vitiation of vata.

Vata vitiation occurs due to *Dhatu Kshaya or Margavarodha. Margavarodha causes dhatu sosha* (8). - *(karotyavatamargatvaad rasadi kopasoshayet)*

The vitiated vata in the body will vitiate the Garbha, Shukra, Arthavam (Bija) (*Garbha sukra rajonasha spandanam gatrasuptataa)*

In the *Bijas* the effected part in the *Bija* correspondingly effects similar part of the fetus (19). (*Yasya yasya havayavasya bhije Bijabage* ...

Majjavahini dushyanti viruddanam cha sevanat).

Common vata treatment (9):

(*Sarpistaila Vasa Majja…..Prasastham Vataroginam*)

Bahya: *Abhyanga, Sashtika Sali Pindasweda, Meru vasti, Vasti.*

Abhyantara: *Vata hara Oushadhi, Balya, Medhya rasayanas, Mamsa and Majja prayoga.*

Role of physiotherapy (20):

(*Lagavam karma…vyayamadupajate)*

The treatment protocol intended to the present child and executed was basing on the following principle.

To remove *margavarodha* (*Kapha avarana*) if any and followed by kevala or samanya vata chikitsa.

Ayurvedic treatment for SMA is primarily aimed at arresting / slowing down the progress of the illness and helping alleviate the symptoms. However, in many cases an attempt was made to reactivate the neurons in the spinal column.

The vitiated Vata produces specific diseases because of the specific nature of the causative factors and the seats of manifestation, specific treatment should be given based on site, dhushyas etc. as per individual person

Common line of treatment for vitiated Vata - Ghee, oil, muscle fat, bone marrow, seka, abhyanga, vasti, snigda, sweda, to stay in windless place, cover with blankets, mamsa rasa, milk food and sweet, sour and salt food. All these will be good for the Vata vitiated persons.

Some specific treatments:

Majja sukrasamuttanamoushadam….matraya (21)

Mahasnehostimajjaste…

- *Mamsa prayoga* (15)
- *Nadiswedam (*16)
- *Majja prayoga* (17)
- *Sahacharadi tailam* (18)

Meru Vasti- with Maha Vishagarbha tailam (4) - Vishamusti or Kupilu or poison nut is one of the ingredient of Vishagarbha tailam, which is capable of activating neurons by virtue of Strychnine. In this context it activates alpha motor neurons of anterior horn cells in spinal cord (5,6,7).

Treatment Limits the progress of the motor neuron degeneration, optimizing neuron activity, Preventing further complications by Vathanulomanam & rasayanam

Dhatvagni niyamana is achieved which further Stops further muscle atrophy, Promote muscle tonicity by Dhatu poshana and Improves tissue immunity –

Balya and Medhya Rasayanas boosts inherent mechanism.

Abhyanga, Pinda sweda, kati vasti helps in fortification of neuromuscular coordination (stimulating the nerve endings by nourishing thus maintaining proper motor functions).

Conclusion:

In this patient a remarkable improvement in muscle tone, power & regaining back of few motor skills with our interventions in a short span gives a hope of further progress of symptoms and helps to improve quality of life (QOL).Treatment intervention in early stages help in getting a major benefit at later ages. Previously it was believed that neurons do not repair or rejuvenate after injury, but the new concept of neuroplasticity says that CNS have the ability to repair their neurons by axonal sprouting to take over the function of damaged neurons (14).

Going by the results of this case study, we can conclude - Ayurvedic modality of treatment is helpful for a better management of SMA.

Through **Multi-disciplinary approach - Ayurveda** as baseline therapy associated with other evolving therapies like physiotherapy, occupational, behavior therapies would certainly can do a lot for the improvement of QOL..

Hence, further research can be done on a larger sample.

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