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CASE STUDY

Ayurvedic Management of *Sarvanga Roga* w.s.r to Charcot Marie Tooth disease – A Case Study

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ABSTRACT

Charcot-Marie-Tooth disease (CMT) is one of the common inherited neuropathies, caused due to mutations of the genes that produce proteins that are involved in the structure and function of either the peripheral nerve axon or the myelin sheath. Its clinical presentation comprises of toe walking, pes cavus, claw hand, waddling gait, foot drop, absent or decreased deep tendon reflexes, nerve hypertrophy and loss of touch sensation predominantly in feet and legs. Prevalence rate is 1 in 2,500 individuals and it is associated with duplication (in 90%) of 17p11.2 (Hemizygous duplication). This can be correlated with *Sarvanga Roga* in Ayurveda. Here is a study of 24 years old male patient who is a known case of Charcot Marie Tooth disease complaining gradual onset of weakness in both lower limbs below the knee, foot and in both arms and hands since childhood, associated with thinning of distal muscles, difficulty in walking and muscular pain over the limbs and stiffness over joints. Considering these symptoms, case was diagnosed as *Sarvanga Roga* and treated using Ayurvedic principles. The *Panchakarma* therapies including *Bahirparimarjana Chikitsa, Basti Karma* along with *Shamana Aushadhis* has showed a sustainable improvement.

Key Words Charcot-Marie-Tooth disease, *Sarvanga Roga, Bahirparimarjana Chikitsa, Basti Karma, Shamana Aushadhis*

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INTRODUCTION

Charcot-Marie-Tooth disease (CMT) described by Charcot and Marie for the first time in 1886 at Paris, and Tooth in London, and was referred to as 'peroneal muscular atrophy'. It is clinically and genetically a heterogeneous group of hereditary motor and sensory neuropathies with a prevalence of 1 in 2,500 people, it is the most frequently inherited neuropathy and also the most common neurogenetic disorders[1]. CMT is caused due to mutation of genes that produce proteins involved in the structure and function of either the peripheral nerve axon or the myelin sheath. The various subtypes of CMT disease are classified accordingly to the nerve conduction velocities, the predominant pathology (ex - Demyelination or

CASE STUDY

axonal degeneration), inheritance pattern and the specific mutated genes[2]. The two main types are: CMT type 1 (CMT1; demyelinating form) and CMT type 2 (CMT2; axonal form).

CMT1 is the most common hereditary neuropathy, and its ratio CMT1:CMT2 being approximately 2:1. Affected individuals presents with distal leg weakness (e.g., footdrop), in the first to third decade of life, although patients may remain asymptomatic even late in life, CMT1A is the most common subtype of CMT1 representing 70% of cases [2, 3].

CMT2 tends to present later in life compared to CMT1 and affected individuals usually become symptomatic in the second decade of life, some cases present earlier in childhood, while others remain asymptomatic into late adult life. Clinically, CMT2 is for the most part indistinguishable from CMT1[3, 4].

Patients with CMT usually presents with progressive weakness/paralysis of muscles of distal legs, which can cause difficulty lifting the foot (foot drop), high stepped gait with frequent tripping or falling, balance problems. Foot deformities such as high arches (pes cavus) and curled toes (hammer toes) are common in CMT. As the disease progress, weakness may occur in distal arm, hands, causing claw hand, difficulty with fine motor skills. As it is a lower motor neuron involvement, individuals present with atrophy of the muscles below the knee, leading to so called inverted Champagne bottle legs.

Degeneration of sensory nerve axons may result in reduced ability to feel heat, cold and touch. The vibration sense and position sense (proprioception) are often decreased in individuals with CMT. The muscle stretch reflexes are unobtainable or reduced throughout. The disease also can cause displacement of curvature of the spine (Scoliosis) and even hip displacement.

Many people with CMT disease will develop contractions and Contractures such as, Chronic shortening of muscles and the tendons around joints that prevents the joints from moving freely. Muscle cramping is common. In some individuals with CMT, may need to rely on leg or foot braces or some other orthopaedic devices to maintain their mobility. In some, nerve pain can range from mild to severe. Some people with CMT experience tremors, vision and hearing may also be affected. In some rare cases, there may be chances of developing breathing difficulties as the nerve that control the muscles of the diaphragm are affected. The severity of the above said symptoms may vary among individuals and among family members with the disease.

Diagnosis is mainly based on symptoms. Nerve conduction study, EMG (Electro Myography), Genetic testing may help. Nerve enlargement may be felt or even seen through the skin, especially at the elbow. These enlarged nerves which are called as hypertrophic nerves are caused due to abnormal thickening of myelin sheaths. Nerve biopsies (calf of leg) if done reveals reduction of myelinated nerve fibres with a predilection for the loss of large diameter fibres and Schwann cells proliferation around thinly or demyelinated fibres, forming so called “onion bulbs”.

CMT is not a life-threatening condition, but this may rarely affect the muscles that are involved in vital functions like breathing. People with most forms of CMT will be having normal life expectancy[5]. There are no medical therapies for any of the CMT's, but physical and occupational therapy can be beneficial as can bracing (ex - ankle foot orthotics for foot drop) and other orthotic devices[2].

There is currently no cure for CMT, but it can be managed with supportive therapy, hence there is need to form a standard protocol for such cases. According to Ayurveda the present case study can be considered as *Sarvanga Roga* which is explained in the context of *Pakshagata*, that *Pakshagata* is called *Ekanga Roga* and if the same affecting both half of the body i.e., all 4 limbs is called as *Sarvanga Roga*[6]. It is a type of *Vata Vyadhi* and in this case along with *Vruddha Vata Dosha* there is an association of *Kapha Dosha* as well. Hence the treatment was aimed at reducing the *Vruddha Kapha* followed by *Vatahara* line of treatment, and the results were found effective.

CASE REPORT

A 24-year-old male patient complains of gradual onset of weakness of lower limbs below the knee and foot since 10 years of age. He was diagnosed as CMT. After 2 years he noticed weakness of both arms and hands with thinning of muscles of hands, difficulty in lifting the foot, getting up from chair, walking with fear of fall. So, he started using splint to his both feet. Later he developed muscular pain over the limbs and stiffness over the joints along with flexion of metacarpals of both hands which gets aggravated during morning hours. Regularly he used to go for Physiotherapy and stopped it since 4 years. Now since 2 months all the symptoms got aggravated.

Past history: Not a known case of Diabetes mellitus and Hypertension, no history of Tuberculosis and has not undergone any major surgical procedure. No history of specific medication and no history of smoking and alcohol consumption.

Family history: Nothing significant

Treatment history: Physiotherapy and Neurodevelopment training like, Independent Standing, Gait and Balance training, Hand functions.

Ashtasthana Pareeksha

- *Nadi* – 78 beats/min
- *Mala*- once per day
- *Mutra*- 3-4 times per day/once in night
- *Jiwha - Lipta*
- *Shabdha - Prakrutha.*
- *Sparsha - Prakrutha.*
- *Drik– Prakrutha.*
- *Arkruthi - Madhyama*

Dashavidha Pareeksha

- *Prakriti – Vatakapha*
- *Satwa, Samhanana - Avara*
- *Pramana, Sara – Madhyama*
- *Saatmya, Aharashakti – Madhyama*
- *Vyayaama shakti – Avara*
- *Vayataha – Yuva*

Examinations:

CNS Examinations-

- Higher Mental Functions – Conscious and Co-operative

Oriented to time and Place

- Memory – Intact
- Speech – Normal
- Cranial Nerve Examinations – Normal
- Motor System –

		Right	Left
Attitude of limb	Upper limb	Flexed wrist and digits	Flexed wrist and digits
	Lower limb	Foot drop, Flat foot	Foot drop, Flat foot
Muscle tone	Upper limb	Hypotonia	Hypotonia
	Lower limb	Hypotonia	Hypotonia
Muscle Power	Upper limb	3/5	3/5
	Lower limb	4/5	4/5
Muscle bulk - Atrophy of distal muscles of both upper and lower limbs	Upper limbs	➢ Flexor group and extensor group muscles in anterior and posterior compartment of forearm respectively. ➢ Thenar and Hypothenar muscles of hand	
	Lower limbs	➢ Anterior Compartment of leg - Extensor hallucis Longus, Extensor Digitorum Longus, Tibialis anterior. ➢ Posterior Compartment of low Calf muscles. ➢ Lateral Compartment - Peroneus longus, Peroneus brevis	
ROM	Upper Extremity	Restricted active wrist extension	Restricted wrist extension
	Lower Extremity	Restricted active dorsiflexion of foot	Restricted active dorsiflexion of foot

- Co – ordination
  - ➢ Finger nose test – not possible
  - ➢ Knee heel test – not possible
- Sensory system – Intact
- Involuntary movements – Absent
- Gait – Waddling gait
- Superficial reflexes

Corneal (B/L) – Normal

Abdominal -Normal

Babinski's sign – negative

- Deep Reflexes

Biceps(B/L) – Reduced

Triceps (B/L) – Reduced

Supinator (B/L) – Reduced

Ankle jerk (B/L) – Reduced

Knee Jerk (B/L) – Reduced

Clonus - Absent

- Fasciculations and Irritability – Absent

Investigation

- Hb% - 16.6gm/dl
- WBC – 9700cells/cumm
- Neutrophils – 65
- Lymphocytes – 28
- Eosinophils – 04
- Macrocytes – 03
- ESR – 36 mm 1st hour
- RBS – 114mg/dl

DIAGNOSIS AND TREATMENT: Based on manifested symptoms and clinical findings after proper history taking and physical examinations, the patient was diagnosed with *Sarvang Roga* w.s.r to Charcot Marie Tooth Disease and patient was administered with *Panchakarma* procedures along with *Shamana Aushadhis* as shown in Table 1 and Table 2 and *Shamana Aushadhis* during follow up in Table 3.

Table 1 Treatment

Sl no	Procedure	Duration
1.	*Sarvanga Dhanyamla Seka*	5 days
2.	*Sarvanga Salvana Upanaha*	Next 5 days
3.	*Sarvanga Shastika Shali Pinda Sweda*	
4.	*Yoga basti: Anuvasana with Mahamasha Taila* – 70ml *Niruha Basti – Madhu* :100ml *Saindhava:* 6 grams *Dhanwantara Taila* -70ml *Ashwagandha Gritha:* 70ml *Shatapushpa Kalka: 20 grams* *Balamoola Ksheera Kashaya*: 350ml	8 days

Table 2 *Shamanaushadhi*

Sl.No	*Shamanaushadi*	Dose	*Anupana*	Duration	Time
1.	*Gandharvashastadi Eranda Taila*	0-0-10ml	With equal quantity of *Ushna Ksheera*	2 days	Before food
2.	*Astavarga Kashaya*	10ml-0-10ml	*Sukoshna Jala*	10 days	After food
3.	*Dhanadanayanadi Kashaya*	10ml-0-10ml	*Sukoshna Jala*	10 days	After food
4.	*Tablet Ekanga Veera Rasa*	1-0-1	*Sukoshna Jala*	10 days	After food
5.	*Balamoola Ksheera Kashaya*	50ml -0- 50ml	*Ksheera*	10days	After food

Table 3 *Shamanaushadhi* at discharge

Sl No	*Shamanaushadhi*	Dose	*Anupana*	Time
1.	*Vidaryadi Kashaya*	10ml-0-10ml	*Sukoshna Jala*	After food
2.	*Dhanwantara Kashaya*	10ml-0-10ml	*Sukoshsna Jala*	After food
3.	*Ashwagandha Churna (10grams)* *Abhraka Bhasma (125mili grams)*	5gms-0-5gms	*Ksheera*	Before food
4.	*Tablet Shatavari*	2-0-2	*Sukoshna Jala*	After food
5.	*Ashwagandha Gritha*	20ml-0-20ml	------	Before food
6.	*Mahamasha Taila*	External Application all over body	--------	Before bath

OBSERVATIONS AND RESULTS

After 10 days of treatment muscular pain over the limbs and stiffness over the joints markedly reduced, stiffness in the back region reduced. During follow up *Shamana Aushadhi* was administered for a duration of 3 months which showed a gradual improvement, after that patient was able to sit and work for long hours without any discomfort, walk without any difficulty and fear of fall while walking markedly reduced. Gait distinctly improved, stiffness in the low back region reduced completely. Muscle bulk of lower limbs noticeably improved when compared to upper limbs.

DISCUSSION

According to Ayurveda this case of CMT can be paralleled with *Sarvanga Roga* explained in the context of *Pakshagata. Karmakshaya* or *Pakshagata* of half of the body is called as *Ekangaroga*, the same if affects both halves of the body it is called as *Sarvanga Roga*[6]. In this case patient had weakness or *Karmakshaya* of all the four limbs (*Sarvanga)* along with stiffness and contractions in due course of illness because of shortening of muscles and tendons around the joints (which presents the joint from moving

freely). Thinning/Atrophy/Wasting of the muscles of limbs were seen as the lesions is LMN.

Though this *Sarvanga Roga* is *Vataja*, as the symptoms like pain, stiffness, weakness used to aggravate in the early morning hours shows the association of *Kapha* with the *Vata* need to be considered. This associated *Kapha* should be addressed first in the treatment, while making sure that *Vata* is under "*Anuloma Gati*". So *Ruksha Sweda* i.e., *Dhanyamla Seka* was started as *Bahirparimarjana* which is one of the *Ruksha Ushna Sweda* having *Amla Rasa,* does *Deepana, Amapachana. Shrama Klamahara* and its specific actions are *Kaphavata Shamana*. *Gandharvahastadi Eranda Taila* was given orally for the purpose of *Vatanulomana*. As *Acharya Charaka* explains whenever there is association of Pitta or Kapha along with *Vata*, one should opt for *Snehayuktha Mrudu Virechana* for the purpose of *Vatanulomana*[7] and also to prepare the *Kosta* for *Basti Chikitsa*.

Later patient felt free from pain and stiffness during morning hours, the treatment was shifted from *Ruksha* to *Snigdha* therapies with *Sarvanga Shastikashali Pinda Sweda* and *Salvana Upanaha*. *Salvana Upanaha* is a type of *Niragni Sweda* and mainly indicated in *Kapha Samsrista Vata* as the drugs in *Salvana Upanaha* are *Vata Shamaka* and *Ushna Veerya*. Here the drugs are observed by the *Romokoopa (*Hair follicles) and spreads all over the body through *Swedavaha Srotas* reducing the *Lakshanas* like *Stambha (*Stiffness*)* and *Gauravata (*Heaviness*). Sarvanga Shastika Shali Pinda Sweda* where *Shastika Shali* having *Snigdha, Picchila, Sheeta* and *Mrudu Guna* along with *Ksheera* having *Guna* of *Brimhana* (Nourishing), *Balya* (Strengthening) when administered as *Sweda (*Sudation*)* mitigates the *Ruksha (*Dry*), Khara (*Rough*)* and *Vishada* (Clearness) *Gunas* of *Vata* imparting *Bala* (Strength) to the *Mamasa Dhatu* (Muscle tissue). *Yoga Basti* comprising *Anuvasana Basti* with *Mahamasha Taila* and *Niruha Basti* with *Balamoola Ksheera Kashaya,* which is again an *Madhura Rasa Pradhana, Madhura Vipaka* and having *Snigdha* (Unctuousness), *Guru* (Heaviness) *Guna* does *Brimhana* (Nourishing) and *Vatahara. Shamanushadi* which are *Kapha Vatahara* were advised initially and later *Vatahara*, *Brimhana* and *Rasayana Aushadhas* was administered.

Astavarga Kashaya has *Bala, Sahachara, Eranda, Shunti, Rasna*, etc. It has *Katu Tikta Rasa, Ushna Veerya, Vata Kaphahara* property[8]. *Danadanayana Kashaya* contains *Danadanayana (Kuberaksha Mula), Shunti, Shigru, Rasana, Pippali* etc. It has *Tikta, Katu Rasa, Ushna Veerya* and *Kaphanubhandha Vatahara* property[9]. *Ekangveer Rasa* is a Herbo mineral medicine. It has ability to pacifying vitiated *Vata Dosha* as it is having *Madhura Rasa, Snigdha Guna* (Unctuousness), *Ushna Veerya* (Hot potency) and *Madhura Vipaka.* It pacifies vitiated *Kapha Dosha* by *Tikta* (Bitter), *Katu* (Pungent)*, Kashaya Rasa* (Astringent)*, Laghu Guna* (Light)*, Ruksha Guna* (Dry)*, Ushna Veerya* (Hot potency) and *Katu Vipaka*. It is mainly indicated in *Pakshaghata, Ardita*, and other *Vata Vyadhi*[10]. *Balamoola* was

given in the form of *Ksheerapaka* as explained by *Acharya Susrutha* in *Chikitsa Sthana,* that *Balamoola* when administered in dose of one *Pala* or half *Pala* along with *Ksheera.* It is advised in those who desires to have *Bala*[11]. *Vidaryadhi Kashaya* has *Vidari, Eranda, Punarnava, Devadaru*, etc, has *Vata Pittahara. Hridya* and *Brimhana* and beneficial in *Shosha, Angamardha*, etc[12]. *Dhanwanthara Kashaya* contains *Yava, Kola, Kulattha, Dashamoola, Triphala, Manjistha*, etc, mainly indicated in *Sarva Vatavikara*[13]. *Ashwagandha Gritha* contains mainly *Ashwagandha* which is *KaphaVata Shamaka* and it acts as *Balya* and *Brimhana*[14]. *Shatavari* given in tablet form also has *Madhura* and *Tikta rasa, Guru* (Heavy), *Snigdha Guna* (Unctuousness), *Seetha Veerya* (Cold potency), does *VataPitta Shamana* and has properties like *Balya* (Nourishing), *Rasayana* (Rejuvenation), *Shulahara*, hence can be advised in *Dourbalya* (Weakness), *Dathukshaya, Kshayaroga* and in *Vata Vyadhi*[15]. *Ashwagandha Churna has Madhura, Kashaya, Tikta Rasa, Laghu*, *Singdha Guna, Ushna Veerya*, and does *Kaphavata Shamana,* has properties like, *Balya, Brimhana, Rasayan* [16]. *Abhraka Bhasma* has *Madhura Rasa*, *Singdha, Sheeta Seerya*, and has properties like, *Vatahara, Deepana, Pachana, Balavardhana* and brings *Sthirata* to the body[17]. *Maha Masha Taila* advised for external application is *Ushna, Guru, Balya, Brihmana* and *Vatashamana*. It contains *Masha* and *Dashamoola* as major ingredients. It is highly useful in muscular weakness and muscular atrophy[18].

The patient was advised to take *Pathya* such as, 1 cup of *Shastika Shali* rice boiled and cooked in milk and other *Vata Kapha Aahara's* and also advised to avoid *Apathya's* like curd, lentils etc, which increases *Vata and Kapha Dosha.* Hence the outcome was a combined effect of Both *Panchakarma* therapies including *Bahirparimarjana Chikitsa, Basti Karma, Shamana Aushadhis* and *Pathya.*

CONCLUSION

The above said *Panchakarma* therapies along with *Shamana Aushadhis* has given a sustainable improvement in managing *Sarvanaga Roga* w.s.r to Charcot Marie Tooth disease, a type of *Kapha Samsrista Vata Vyadhi*. This intervention helped the individual in improving quality of health to lead an independent life.

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