



POEMS SYNDROME- COMPLEMENTARY APPROACH IN AYURVEDA: A CASE REPORT

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ABSTRACT

POEMS syndrome is a rare multisystem disorder that develops due to an underlying plasma cell dyscrasia. Peripheral neuropathy is a prominent symptom and often the first identified common clinical condition. In the present study, a diagnosed case of POEMS syndrome, a 50-year-old male patient who presented with pins and needle sensation in B/L soles, electric shock like sensation and heaviness of both lower limbs below knee joints since one year; imbalance while walking and tendency to fall on tripping over small objects from 06 months of onset of the disease. Examination revealed absent triceps, knee and ankle jerk; and quadriplegia of all limbs. Sensory system examination showed decreased pain, touch and temperature sensation on bilateral lower limbs below knee level and lateral aspect of left thigh. Loss of vibration sensation on both lower limbs from ASIS (Anterior superior iliac spine) to toes and mild impairment in JPS (Joint Position Sense) in bilateral toes also noted. Allodynia, swelling and blackish discoloration present on B/L lower limbs. Considering the signs and symptoms, the disease was diagnosed as *Vatakaphadhika Vatarakta*. Symptomatic treatment focusing on external therapy (*Dhanyamladhara* – Pouring of *Dhanyamla* over body) was done for 14 days. The intervention resulted in significant clinical improvement as evidenced by the reduction in NPS (Neuropathic Pain Scale Score) from 80 to 38 and improvement in vibration and hot sensation assessed using Neuropathy Analyser. There was remarkable improvement in swelling and pain in the patient. This case report is an attempt to introduce POEMS syndrome in the light of Ayurveda and to emphasise the promising role of integrative therapeutic approaches in improving patient's quality of life.

Key words: POEMS Syndrome, Ayurveda, Demyelinating Neuropathy, *Dhanyamladhara*, Neuropathic Pain Scale

INTRODUCTION: POEMS (polyneuropathy, organomegaly, endocrinopathy, M-protein, and skin changes) syndrome is a rare multisystemic paraneoplastic disorder.¹ It was first narrated by Crow in 1956 and then by Fukase in 1968.² The acronym POEMS was coined by Bardwick et al in 1980 based on the 5 main features of the disease.³ This syndrome has an incidence

of approximately 3 in 100,000, with male predominance.⁴ The diagnosis of POEMS syndrome, based on the Dispenzieri diagnostic criteria, requires the presence of both mandatory criteria (a polyneuropathy and a monoclonal plasma cell-proliferative disorder, usually lambda restricted), and at least one major and one minor criterion.⁵

Table No:1 Diagnostic criteria for POEMS syndrome

MAJOR CRITERIA	MINOR CRITERIA
<p>Mandatory:</p> <ul style="list-style-type: none"> • Polyneuropathy • Monoclonal plasma proliferative disorder <p>Other criteria:</p> <ul style="list-style-type: none"> • Sclerotic bone lesions • Castleman’s disease* • Elevated VEGF(Vascular endothelial growth factor) 	<ul style="list-style-type: none"> • Organomegaly (splenomegaly, hepatomegaly or lymphadenopathy) • Endocrinopathy (adrenal, thyroid, pituitary, gonadal, parathyroid, pancreatic) • Oedema • Skin changes hyperpigmentation, hypertrichosis, glomeruloidhaemangioma, plethora, acrocyanosis, flushing, white nails • Papilledema • Thrombocytosis/Polycythaemia <p>Other symptoms and signs</p> <ul style="list-style-type: none"> • Clubbing • Weight loss • Hyperhidrosis • Pulmonary hypertension • Diarrhoea

*Castleman disease (CD) is a heterogeneous group of lymphoproliferative disorders that share common morphological features on lymph node biopsy.

The neurological hallmark of the syndrome is a sub acute onset demyelinating neuropathy which often presents as the common first clinical feature that can be rapidly disabling and painful.⁶ The aim of treatment is to eradicate the underlying plasma cell dyscrasia but there are no established treatment guidelines for POEMS Syndrome. Supportive care considerations include management of endocrinopathy, neuropathy, thrombosis, and infection.⁷

PATIENT INFORMATION:

De-identified demographic and other patient information

A 50-year-old married male, employed as a driver, belonging to middle economic status, with k/h/o systemic hypertension for 5 years and dyslipidaemia for 2 years on regular medication, diagnose as POEMS Syndrome from Sri Chitra Tirunal

Institute for Medical Sciences and Technology on March 2021 came to our OPD on 07/10/2021. He presented with complaints of pins and needle sensation in B/L soles, electric shock like sensation and heaviness of both lower limbs below knee joints since 1 year; imbalance while walking and tendency to fall on tripping over small objects from 6 months of onset of the disease. He also presented with swelling of both lower limbs. The patient adhered to a mixed diet with irregular timing of intake. He had loose bowels since 1 month and regular bladder movements. He is a non-smoker and denies habitual usage of alcohol. He did not engage in regular exercise.

Main concerns and symptoms of the patient

The patient experienced an insidious onset of pins and needle sensation in B/L soles(L>R) which was less during early morning hours and more on activities. He developed electric shock like sensation when someone accidentally hit on his feet. He felt as if walking over a cotton-wool

which became difficult in the course of time. There was slippage of chappals with his knowledge such that he used to hold them with toes to prevent from slipping. No difficulty in navigating toes or removing of footwear was noted. He had shortness of breath. There was distension of abdomen with protrusion of umbilicus. He noticed a weight loss of 10 kg within a span of 3 months. The patient denied any h/o of involuntary movements/fever/jaundice/diplopia.

Medical, family and psycho-social history including relevant genetic information

There was no family history of a similar illness or any neurological illness. The patient was tormented by his health condition as well as financial crisis.

Relevant past interventions and their outcomes

Nothing relevant to present condition

CLINICAL FINDINGS:

The patient was overweight and well-nourished with a height of 176 cm and weight of 82 kg, resulting in a BMI of 26.5 kg/m². Vital signs were within normal limits. General examination revealed B/L pitting pedal oedema. There was no lymphadenopathy. Blackish discoloration of feet below the ankle was noted. Neurological examination showed a normal higher mental function and no cranial nerve involvement was found. Motor system examination revealed absent triceps, knee and ankle jerk and quadriparesis of all limbs. Sensory system examination showed pain, touch and temperature sensation decreased in bilateral lower limbs below knee level and lateral aspect of left thigh. Loss of vibration sensation on

TIMELINE

both lower limbs from ASIS to toes and mild impairment in JPS in bilateral toes also noted. Allodynia also present on B/L lower limbs. There were no signs of cerebellar dysfunction or extrapyramidal signs. No sign of meningeal irritation. Cardiovascular examination showed dyspnoea on exertion of grade 1 severity. Respiratory system examination showed AEBE (Air Entry Bilaterally Equal). Gastrointestinal system examination revealed reducible umbilical hernia. The patient reported a NPS score of 80 before treatment. Vibration & thermal perception assessment using Neuropathy Analyser showed moderate loss in vibration perception B/L and hot perception (B/L) and normal cold perception study. Ayurvedic examination (*Dasavidhapareeksha*) revealed:

Prakriti- Sharira Prakriti: KaphaPitta
Manasa Prakriti: TamaRaja
Vikriti-
Dosha: VataKaphapradhanaTridosha
Dhatu: Rasa, Raktha, Mamsa, Medas, Asthi
Upadhatu: Sira, Snayu
Mala: Pureesha
Sara- Medosara
Samhana- Madhyama
Pramana- Madhyama
Satmya- Madhyama with Katu and Lavana Rasapreeti(predominantly pungent and salty taste food intake)
Satva- Madhyama
Aharasakthi- Abhyavaharanashakti(quantity of food intake): *Madhyama*
Jaranashakti(power of digestion): *Madhyama*
Vyayamasakthi- Avara
Vaya - Madhyama (middle aged)

Table 2: Timeline showing the disease course, diagnosis, treatment and outcomes.

Year / Month	Observation and Management
2020-2021	The patient developed gradual onset of pins and needle sensation of B/L foot and electric shock like sensation over both lower limbs below knee joint. Difficulty in walking and tendency to fall developed
February 2021	He took consultation in secondary care OPD for above complaints. He was diagnosed with Peripheral Neuropathy by a general physician. Symptoms progressed.
March 2021	He took consultation at SCTIMST. After a detailed examination and appropriate investigations, he was diagnosed as a case of POEMS Syndrome.
May 2021	Took 20 cycles of radiation therapy for Plasmacytoma at RCC Trivandrum. He was advised Tab Gabantin NT (1-0-1), Tab Wysolone 20 (1-0-0) however symptoms persisted
Oct 2021- Nov 2021	He came for Ayurvedic management with complaints of pins and needle sensation in B/L soles, electric shock like sensation over B/L lower limbs below knee joint, heaviness of both lower limbs below knee joints; imbalance while walking and tendency to fall on tripping over small objects. Assessment using Neuropathy Analyser showed moderate loss in vibration perception (B/L) and in hot perception (B/L) and normal cold perception study. NPS score was 80. <i>Dhanyamladhara</i> was done for a period of 14 days. NPS score became 38 and there was improvement in vibration and hot sensation assessed using Neuropathy Analyser.

DIAGNOSTIC ASSESSMENT

Diagnostic methods

The patient's clinical presentation and disease progression led to the diagnosis of POEMS disease supported by investigations performed at SCTIMST, Trivandrum.

- 1.USG Abdomen and Pelvis (10/02/2021)
Grade 1 fatty liver, splenomegaly, left renal cortical cyst, Grade 1 prostatomegaly and fat containing umbilical hernia.
- 2.MRI Spine screening (07/03/2021)
Residual iliac bone lesion with diffuse marrow infiltration.
Expansive lytic lesion in left iliac bone with pseudotrabeclulation, sclerotic rim and few areas of cortical breakthrough in

posterior margin and iliac aspect of left SI joint.

- 3.Bone marrow biopsy (26/03/2021)
Mild plasmacytosis.
- 4.Serum protein electrophoresis (26/03/2021)
He was found to have elevated free IGG lambda light chains in serum protein electrophoresis.
Based on the symptomatology, the case was diagnosed as *Vatakaphadhika Vata-rakta* as per Ayurveda.

Diagnosis

Being a disease with multisystem involvement, the following are the probable diagnosis:

Table No: 4 Probable Diagnosis of the patient

Symptoms in patient	Dosha/dhatu involved	Probable Diagnosis
<i>Soochibhiriva nistoda</i> (pins and needle sensation)	<i>Vata, Rakta</i>	<i>Vatarakta, shonitavruta vata</i>
<i>Teevra Ruk</i> (severe pain)	<i>Vata, Pitta, Rakta</i>	<i>Vatadhika and Raktajavatarakta,</i>
<i>Chimchimayana</i> (tingling sensation)	<i>Vata,Rakta</i>	<i>RaktadhikaVatarakta, Shonitavruta Vata</i>
<i>SparsaAkshamatva</i> (Allodynia)	<i>Vata, Rakta, Pitta</i>	<i>PittadhikaVatarakta</i>
<i>Gurutva of Adhakaya</i> (Heaviness of lower limbs)	<i>Kapha</i>	<i>KaphadhikaVatarakta</i>
<i>Gati vaishamya</i> (Difficulty in walking)	<i>Kapha, Vata</i>	<i>Kaphavrutavata</i>
<i>Krishnata of pada</i> (Blackish discoloration of feet)	<i>Vata, Rakta</i>	<i>Vatadhikavatarakta,</i>
Splenomegaly	<i>Vata, Pitta, Rakta</i>	<i>Raktasthanadusthi, Avaranaupadrava, Pleehodara</i>
Grade 1 fatty liver	<i>Rakta, Medas</i>	<i>Rakthasthanadusthi, Yakritodara</i>

Diagnostic reasoning including differential diagnosis

In this case, we can see significant *dusthi* at the level of *Rakta dhatu, upadhatu* as well as its *sthana*. This *Rakthadusthi* in turn causes *avarana* of *Vata* leading to *Vatarakta*.

Depending upon the *Rupa*(symptoms), the disease was diagnosed as *VataKaphadhikaVatarakta*.

Prognostic characteristics when applicable

POEMS Syndrome is a rare multisystem disorder that has high morbidity and mortality if left untreated.

Improper diagnosis and delayed management of *Avaranavyadhior* of one year chronicity leads to complications like *Hridroga, Gulma, Pleeha, Agnisada, Vidradi* etc.

THERAPEUTIC INTERVENTION

Type of therapeutic intervention (Allopathic drugs)

The patient was on the following conventional medications: Tab Concor 2.5 (1-0-1), Tab Gabantin NT (1-0-1), Tab Wysonone 20 (1-0-0), Tab Rosuvas 10 (0-0-1)

Type of therapeutic intervention(Ayurveda)

As the patient was advised to not take any other oral medications, no internal Ayurvedic medicines were given. *Dhanyamladhara* was done for 14 days.

Consent: Written informed consent was taken from the patient before the treatment and also for publication.

DHANYAMLADHARA

Dhanya means cereals, *Amla* means sour and *Dhara* means stream pouring. So *dhanyamladhara* is described as an Ayurvedic treatment procedure in which fermented liquid is poured over the body of patient in monitored streams for a fixed duration of time without interruption with liquid as mild hot. It is mentioned in *Dharakalpaadhyaya* of *Sahas-*

rayoga.⁸ Dhanyamladhara comes under the category of sudation or induced sweating therapy.

Methodology: Pulse rate, heart rate & B.P are monitored before and after treatment and recorded.

Materials required: Dhara vessel with stand, pouring vessel and dhanyamla along with heating facility.

Postures: 7 postures if possible or lying side and sitting postures.

Time for each sitting: 30-40 mts

Number of therapists required: Minimum of 2 therapists are needed.

Method: First apply rasnadichoornam over bregma. Then pouring of dhanyamla over the body in mild temperature in suitable positions for above mentioned time period. The procedure is stopped

when patient starts sweating. Relaxed for a while. Then advised light diet only, before and after the procedure. Uniform temperature is to be maintained. Avoid exposure to cold immediately.

FOLLOW-UP AND OUTCOMES

The outcomes were measured before and after the treatment, significant improvement in clinical outcomes with respect to NPS and Neuropathy Analyser were obtained after the completion of therapeutic intervention. NPS score which was 80 before the treatment reduced to 38 after the treatment. There was marked improvement in swelling of B/L lower limbs. The pre and post evaluation of temperature sensation and hot sensation using Neuropathy Analyser is as follows:

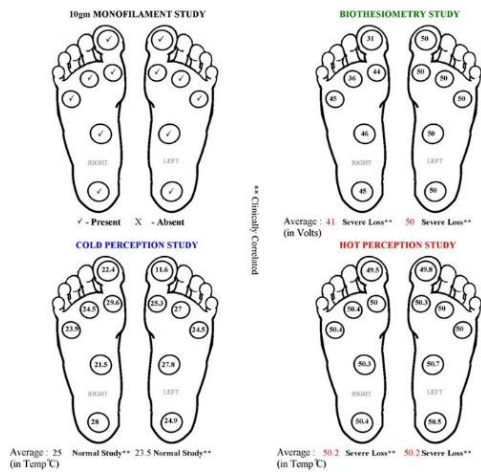


Figure :1 Before treatment

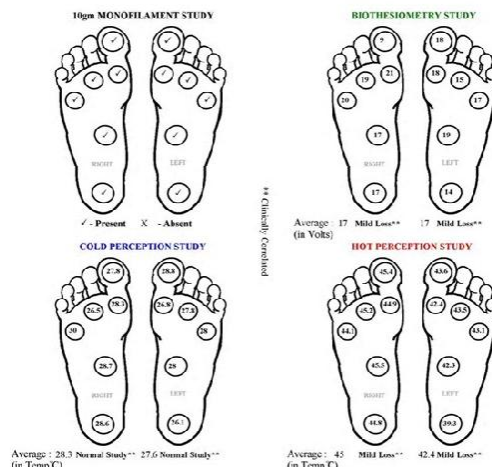


Figure 2: After treatment

DISCUSSION

POEMS syndrome is one of an infrequent and challenging plasma cell disorder, both in the diagnostic and therapeutic management of the disease. Currently, the literature on POEMS is few with most evidence being individual case reports. POEMS syndrome is distinguished from other paraproteinemic and inflammatory neuropathies by its multi-organ involvement thought to be caused by elevated pro in-

flammatory and angiogenic cytokines. Current treatment modalities target underlying plasma cell clone, a monoclonal antibody targeting VEGF which has had unpromising results.

In Ayurveda, the prime factor that is responsible for the pathogenesis of any disease is *Dosha-Dushya Sammurchana* and is defined using the *Nidana Panchaka*. Manifestation of a disease occurs in the body as a result of the defective *srotas* or

kha vaigunya favouring the *Dosha-Dushya* conglomeration. While looking into POEMS syndrome, we can see imbalance in *Vata&Pitta* doshas, characterised by a predominant derangement in *Rakta Dhatu* in conjunction with perturbation in *Rasa, Medo* and *Asthi dhatu*.

Avarana is one of the most complex fundamental concepts of Ayurveda. The clinical

features of this disease which includes polyneuropathy, organomegaly (splenomegaly, hepatomegaly), monoclonal plasma cell disorder, skin changes etc which can be understood as the *upadravas* of *Avarana* like *Pleeha, Gulma, Vidradi* etc.

Table No:3 Probable Ayurvedic View of POEMS Syndrome

POEMS SYMPTOMS	PROBABLE AYURVEDIC VIEW
Polyneuropathy	<i>Vata prakopa, Raktavahasrotodushti</i>
Monoclonal plasma proliferative disorder	Over production of plasma cells may be considered as <i>vatadushti</i> and <i>raktavriddhi</i> . <i>Agnisada</i> and production of <i>vikruta rasamay</i> lead to production of <i>malabhava</i> such as M-protein
Sclerotic bone lesions	<i>Asthi dhatu kshaya</i>
Elevated VEGF	<i>Raktavahasrotodushti</i>
Organomegaly(splenomegaly, hepatomegaly)	<i>Yakrit</i> and <i>Pleeha</i> are described as <i>moolasthanas</i> of <i>Raktavahasrotas</i> . May also occur as complication of <i>Avarana</i>
Oedema	Change in vascular permeability may be considered as <i>Raktavahasrotovaigunya</i>
Skin changes	<i>Raktadushti</i>
Polycythemia	<i>Rakta vriddhi</i>
Endocrinopathy Papilledema Hyperhidrosis Diarrhoea	<i>Pitta prakopa, pitta sthanadushti</i>
Weight loss	<i>Raktagatavatalakshana</i>

The *Nidana* identified in the patient includes *Lavana, Amla, Katu Rasa Pradhana Ahara* (salty, sour and pungent dietary intake) which vitiates *Pitta* and *Kapha* along with *Rakta.YanaVahana*(excessive traveling), *Vegadharana* and *Chintha* (stress) leads to *Vata dusthi*. The vitiated *Rakta* also results in *navarana* of vitiated *Vata*. This *Dosha dusthi* might have resulted in *Rasa, Raktha, Mamsa, Medo, Asthi* and *Mano dusthi*. As a result of this *Dhatu Dushti*, the *Upadhatu* i.e *Sira* and *Snayu* also got vitiated. Thus, the disease manifested with *TeevraRuk, Chimchimayana,*

Soochibhirivanistoda, Sparsa Akshamatva, Gurutva of *adhakaya* and *Gati vaishamy* which are *main prakopalakshanas* of *VataKapha Pradhana Tridoshadushti* along with *Raktadushti* leading to the diagnosis of case as *Vatakaphadhika Vatarakta*. In *Raktajavikaras*, one of the main treatment principles is *Seka*. *Seka* is also indicated in *Sama vataavastha* as well as in *Vatarakta*. *Acharya Susrut* while stating *tiryakgathadhamani* explains that they perceive the effects and potencies of *abhyanga, parisheka, avagaha* etc and they give the sensation of pleasant or unpleasant

ant contact the body perceives. This hints at the function of peripheral nerves which gets stimulated from external environment. Hence here *Dhanyamladharawas* selected owing to its *KaphaVata Samaka* property as well as it does not aggravate *Pitta* being *seetha* in *sparsa*.

Marked improvement was observed after *Dhanyamladhara*. NPS score reduced from 80 to 38 and there is positive outcome in vibration and hot sensation assessed using Neuropathy Analyser. There is overall improvement in physical and mental status of the patient.

CONCLUSION: This case report provides valuable insight into the understanding of a rare disease like POEMS syndrome in Ayurveda. Being a chronic disease, POEMS syndrome requires long term treatment and follow up to understand the long-standing effects of Ayurvedic intervention. Overall, this case report highlights the potential of Ayurveda in complementing the management of POEMS syndrome.

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