WORLD JOURNAL OF PHARMACEUTICAL RESEARCH
SJIF Impact Factor 8.084
Volume 10, Issue 12, 2146-2156. Case Study ISSN 2277–7105

AYURVEDIC MANAGEMENT OF GIANT AXONAL NEUROPATHY – A SINGLE CASE REPORT

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Article Received on
19 Aug. 2021,

Revised on 08 Sept. 2021,
Accepted on 29 Sept. 2021

DOI: 10.20959/wjpr202112-21903

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ABSTRACT

Male patient aged 16 years diagnosed with a rare genetic disorder – giant axonal neuropathy approached Sri Dharmasthala Manjunatheshwara College of Ayurveda and Hospital, kuthpady, Udupi, Karnataka, India for treatment. Patient presented with the complaints of progressive weakness in all four limbs, difficulty to speak, mild breathing diffulty and difficulty to carryout basic functions. Detailed history was taken and complete examination, with special emphasis to respiratory, musculoskeletal and nervous system was done. Patient was assessed using amyotrophic lateral scelorosis scale and CKNAC levels were monitored. All the 12 visits and inpatient treatment of the patient, through 5 years (2016-2020) were recorded and progress of disease as well as effect of treatment was assessed. Considering that the giant axonal neuropathy has poor prognosis and rapid progression, the treatment given proved to be effective in improving some of the parameters in addition to slowing the progression of the disease. hence it can be concluded that the treatment protocol was effective in management of giant axonal neuropathy.

KEYWORDS: case report, giant axonal neuropathy, sahaja karshya, sarvanga vata.

INTRODUCTION

Giant axonal neuropathy (GAN) is an early onset fatal neurodegenerative disorder that starts as severe peripheral motor and sensory neuropathy and evolves into central nervous system impairment. GAN is inherited in an autosomal recessive manner affecting the gene encoding the protein gigaxonin and is a very rare disorder.[1] The condition can be corelated to sahaja karshya[2] or sarvanga vata.[3-4]

PATIENT INFORMATION

Male patient aged 16 years was apparently normal 9 years back. He had attained developmental milestones at right age and had no ailments/systemic illness. Parents noticed frequent falls on walking and playing, and difficulty in gripping objects. Then one day he fell unconscious, following which he developed weakness initially in right lower limb and then in left lower limbs. Later the weakness was also felt in both upper limbs especially distally. In next 5 years, along with weakness of bilateral upper and lower limbs, there was difficulty to speak and difficulty to walk. He had more difficulty to walk on inclined surface and climb stairs. There was difficulty to pass urine and frequent distension of bladder. During this span they consulted local hospitals and extensive investigations were carried out and was diagnosed with Giant Axonal Neuropathy. In August 2016, they consulted our hospital and is on ayurvedic treatment since then. In the past 4 years, his weakness has been persistent and progressive, he is unable to walk, has difficulty with performing activities like writing, dressing, cutting food etc. he also developed mild breathing difficulty and the speech is slurred. Negative history: no antecedent fever infections, ne delay in milestone achievement. Family history: consaginous marriage of parents. No other family members have similar condition.

Clinical findings

Cardiovascular system: S1, S2 heard normally, no added sounds. Nothing significant.

Gastrointestinal system: soft, non tender abdomen. Nothing significant.

Integumentory system: Nothing significant.

Examination of upper respiratory tract

- Nostril –nasal discharge, hypertrophied turbinates, nasal polyps - absent
- Nasal septum – no deviated nasal septum
- Sinus tenderness - absent
- Pharynx, tonsil –normal

Examination of lower respiratory tract

Inspection

- Shape and symmetry of chest –elliptical , bilaterally symmetrical
- Superficial skin lesions, scar marks -absent
- Respiratory movement - abdominothoracic
- Visible pulsations –absent
- Litten's sign- movement of diaphragm normal and symmetrical on both sides
- Hoover's sign (indrawing of intercostals during inspiration) – absent
- Spine –normal

Palpation

- Position of trachea –centrally placed
- Apical impulse – normal, 5th intercostal space
- Respiratory movements- normal
- Chest expansion- 2 cms
- Vocal fremitus- equal on both sides
- Intercostal and rib tenderness- absent

Percussion

- Lung resonance (percussion notes) – normal, equal on both sides

Auscultation

- Breath sounds, type of breathing and alteration of inspiration and expiration
- Normal vesicular breath sounds (inspiration longer than expiration, no gap between inspiration and expiration) equal intensity on both sides
- Added sounds – Ronchi present
- Vocal resonance- equal on both sides

  Higher mental functions
- Consciousness – conscious
- Orientation – oriented to time place and person
- Intelligence- normal
- Memory- intact
- Hallucinations- absent
- Delusions- absent

- Speech distrurbances- present – slurred speech
- Handedness- right

Cranial nerve examination

- Olfactory nerve – normal functions
- Optic nerve - visual acquity – 6/36 without glasses
  - visual field – normal
- Occulomotor-troclear -abducens nerve
  - movement of eyes- normal
  - nystagmus- absent
  - light reflex- normal
  - accommodation reflex- normal
- Trigeminal nerve
  - sensory- touch pain pressure – normal
  - corneal reflex- normal
  - anterior 2/3rd of tongue- normal
  - motor- muscles of mastication - normal
  - jaw jerk – normal
- Facial nerve

Motor

- Forehead wrinkling- normal
- nasolabial fold- normal , equal on both sides
- angle of mouth- normal, no drooping
- asymmetry of face- absent
- differences in blinking and eye closure- absent
- slurring of speech- present

Power

- eye shutting- normal, possible
- Blowing cheeks- normal, possible
- Showing teeth- normal, possible
- sensory- taste- anterior 2/3rd of tongue- normal
- Vestibulocochlear nerve

- nystagmus- absent
- Rinnes test- air conduction > bone cunduction
- Webers test- equal on both sides
- Glossopharyngeal and vagus nerve
- dysarthria - absent
- dysphonia - absent
- position of uvula- centre
- palate movement- normal, equal
- gag reflex – not ellicited
- sensory- post 1/3rd of tongue- not elicited
- Accessory nerve
- sternocleidomastoid – reduced resistence
- trapezius- reduced resistence
- Hypoglossal nerve
- tongue – wasting - absent
- fasciculation- absent
- involuntary movement - absent
- deviation - absent
- power- tongue against cheek- reduced
- speech – yellow lorry- slurred
- water swallow test- possible

Motor system

- Gait – unable to walk

Inspection and palpation of muscles.

- wasting- present
- hypertrophy- absent
- no fasciculation, no myoclonic jerks, no tremors, no dystonial chorea,no ballism
- no involuntary movements

Table. No. 1: Measurement of limbs.

	Right (cms)	Left (cms)
Upper arm	27	25
Mid arm	22	22
Forearm	20	20
Upper thigh	40	40
Mid thigh	35	35
Lower thigh	31	29
Above calf	22	23
Mid calf	19	20

Muscle tone

- Upper limb – hypotonic
- lower limb- hypotonic
- Knee clonus- absent
- Ankle clonus- absent

Table. No. 2 Muscle power.

	Right	Left
Upper limb	4/5	4/5
Lower limb	2/5	2/5

Coordination

- rebound phenomenon - absent
- finger nose test - altered
- heel shin test – could not perform
- dysdiadochokinesis- present
- tandem walking- cant walk

Superficial reflexes

- corneal- normal
- abdominal- normal
- cremasteric – not ellicited
- plantar- mild flexion

Deep reflexes

- jaw jerk- normal
- biceps + (diminished)

- triceps + + (diminished)
- pectoral + (normal)
- knee jerk + (diminished)
- ankle (absent)

Primitive reflexes

- glabellar tap/ Myerson's sign – normal
- snout reflex - absent
- sucking reflex - absent
- palmomental reflex - absent
- grasping response - absent
- avoiding response – absent
- Beevor's sign – normal position of umbilicus
- Gower's sign – not performed

Sensory system

- Lateral spinothalamic tract
- pain- superficial - diminished
  - deep - normal
- temperature - diminished
- Posterior spinothalamic tract
- touch - diminished
- vibration - diminished
- joint sense - present
- position sense - present
- pressure sense – diminished
- Cortical sensations
- tactile localization - normal
- two point discrimination - altered
- sensory inattention - absent
- stereognosis – altered
- graphesthesia – altered
- Romberg's sign – could not perform

- Pseudoathetosis – absent

Timeline of treatment: from 2016 to 2020

	Medicines	**External treatment**
August 2016	Cap balamoola rasayana 2 tid Mahishadravaka 3 tsp tid Laghumalinivasantha rasa 1 bd Jwarankusha 15ml tid	Rajayapana basti Veshtana with mahamasha taila all 4 limbs Sarvanga abhyanga with mahanarayana taila Shalipinda sweda
November 2016	Cap shatavari 4 tid Cap guru rasayana 1tid Sarivadyasava 15ml tid Discharge medicines: Cap shatavari 4 tid Bhargavaprokta rasayana 1tsp bd with milk Tab dashamoola katutraya 1 tid Laghu malini vasantha rasa 1 bd Mahishadravaka 15ml tid	Veshtana with maharanayana taila
January 2017	Tab dashamoola katutraya 1 tid Laghu malini vasantha rasa 1 bd Mahishadravaka 15ml tid Kapikacchu rasayana 1 tid	Rajayapana basti Veshtana
April 2017	Bhargavaprokta rasayana 2tsp bd with milk Laghu malini vasantha rasa 1 bd Mahishadravaka 15ml tid Discharge medicines: Bhargavaprokta rasayana 2tsp bd with milk Laghu malini vasantha rasa 1 bd Mahishadravaka 15ml tid Balamoola rasayana 2 tid	Rajayapana basti Veshtana
July 2017	Shatavari rasayana 4 tid Bhargavaprokta rasayana 2tsp bd with milk Laghu malini vasantha rasa 1 bd Mahishadravaka 15ml tid	Rajayapana basti Veshtana with mahamasha taila
October 2017	Cap laksha 2 tid Shatavari rasayana 4 tid Bhargavaprokta rasayana 2tsp bd with milk Laghu malini vasantha rasa 1 bd Mahishadravaka 15ml tid Discharge –lashuna course	Sarvanga abhyanga with mahamasha taila
January 2018	Bhargavaprokta rasayana 2tsp bd with milk Laghu malini vasantha rasa 1 bd Mahishadravaka 15ml tid Laksha 2 tid Lashuna course Discharge medicines- Bhargavaprokta rasayana 2tsp bd with milk Laghu malini vasantha rasa 1 bd Mahishadravaka 15ml tid Balamoola rasayana 2 tid	Rajayapana basti Veshtana with mahamasha taila Abhyanga with mahamasha taila

	Laksha 2 tid	
March 2018	Lashuna course Bhargavaprokta rasayana 2tsp bd with milk Laghu malini vasantha rasa 1 bd Mahishadravaka 15ml tid Balamoola rasayana 2 tid Laksha 2 tid Discharge – lashuna course Bhargavaprokta rasayana 2tsp bd with milk Laghu malini vasantha rasa 1 bd Mahishadravaka 15ml tid Balamoola rasayana 2 tid	Rajayapana basti Veshtana with mahamasha taila Abhyanga with mahanarayana taila
June 2018	Shatavari rasayana 4 tid Mahishadravaka 15ml tid Naradiya lakshmi vilasa rasa 1 tid Dhanwantara kasasudha kalpa 5ml tid Kumara kalyanaka rasa 1bd Discharge- Laksha 2 tid Bhargavaprokta rasayana 2tsp bd with milk Shatavari rasayana 4 tid Mahishadravaka 15ml tid Naradiya lakshmi vilasa rasa 1 tid Dhanwantara kasasudha kalpa 5ml tid Kumara kalyanaka rasa 1bd	Rajayapana basti Veshtana with mahamasha taila
September 2018	Laksha 2 tid Cap yashtimadu 2 tid Dhanwantara kasasudha kalpa 5ml tid Shatavari rasayana 4 tid Kumara kalyana rasa 1 od Discharge - Cap yashtimadu 2 tid Shatavari rasayana 4 tid Kumara kalyana rasa 1 od Mahisha dravaka 15ml tid	Rajayapana basti Veshtana with mahamasha taila
April 2019	Cap balamoola 4 tid Kumarakalyana rasa 1 bd Discharge: Cap balamoola 4 tid Kumarakalyana rasa 1 bd Ajamamsa rasayana 1 tsp bd	Veshtana with mahamasha taila Balamoola parisheka Nasya with ksheerabala taila Rajayapana basti
January 2020	Lashuna course Cap balamoola 4 tid Kumarakalyana rasa 1 bd Ajamamsa rasayana 1 tsp bd Discharge:	Rajayapana basti Veshtana with mahamasha taila

Lashuna course: cap lashuna in increasing dosage from 12-24-36-48 (4 days each) with milk.

Table no. 3 Assessment using ALS SCALE.

	1st visit 2016	12th visit 2020
Speech	2	3
Salivation	3	4
Swallowing	3	4
Handwriting	2	3
Cutting	1	3
Personal hygiene	1	3
Turning in bed	2	3
Walking	0	0
Climbing stairs	0	0
Dyspnoea	1	4
Orthopnoea	3	4
Respiratory insufficiency	4	4

Table no. 4 assessment of motor function using ALS scale.

	1st VISIT 2016	12TH VISIT 2020
Shoulder flexion	4	3
Extension	4	3
Abduction	4	3
Adduction	4	3
Elbow flexion	4	3
Extension	4	3
Wrist flexion	3	3
Extension	3	3
Hip joint flexion	3	2
Extension	3	2
Adduction	3	2
Abduction	3	2
Knee flexion	2	2
Extension	2	2
Ankle plantar flexion	1	2
Dorsiflexion	1	2

Table no. 5 CKNAC levels

	CKNAC
AUG 2016	94
JUNE 2018	355
SEP 2018	354
APR 2019	441
JAN 2020	245

DISCUSSION

Giant axonal neuropathy is a rare and genetic disorder and the treatment protocol is not well documented in either modern science or in Ayurveda. This casestudy records the symptoms, presentations, timeline of treatment through 5 years and also the assessment. It is observed that the basic functions of the patient improved with respect to subjective parameters. There is also marked improvement in objective parameter i.e CKNAC.

CONCLUSION

Hence it can be concluded that principle of treatment explained in Ayurveda can be used in management of rare genetic disease like giant axonal neuropathy.

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