IAMJ

Case Report *International Ayurvedic Medical Journal* *ISSN:2320 5091*

AYURVEDA: A HOPE FOR HSP- HEREDITARY SPASTIC PARAPARESIS

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ABSTRACT

Hereditary Spastic Paraparesis is a group of hereditary ataxic disorders; Symptoms usually begin in the third or fourth decade, presenting as progressive spastic weakness beginning in the distal lower extremities. The two genes, spastin and alastin has been identified as the defective gene in causing HSP. In hereditary ataxias, an autosomal recessive inheritance is characterised by several affected members in one generation with healthy parents. Presence of consanguinity in parents strongly favours such inheritance. Typically these disorders manifest in childhood, adolescence or early adulthood. However a negative family history does not always rule out a hereditary disorder This article explains how this disorder begins, how it is managed in Ayurveda by taking the help of the PANCHAKARMA procedures such as Basti, Abhyanga etc.

Keywords: Hereditary Spastic Paraparesis, Abhyanga, Basti, Nasya

INTRODUCTION

Hereditary spastic paraparesis (HSP) is not a single disease entity; it is a group of clinically and genetically diverse disorders. Symptoms usually begin in the thirties or fourties, presenting as progressive spastic weakness beginning in the distal lower extremities[i]. However, there are variants with onset so early that the differential diagnosis includes cerebral palsy. Strumpell (German Neurologist) first described hereditary forms of spastic paraplegia in 1883, with Lorrain later providing more extensive detail. HSP is also called familial spastic paraparesis and Strümpell-Lorrain syndrome. Syndromes are classified as uncomplicated, or pure, when only spinal involvement occurs, and they are classified as complicated when they are associated with neurologic abnormalities, such as ataxia, mental retardation, dementia, extrapyramidal dysfunctions, visual or hearing dysfunctions, adrenal insufficiency, and ichthyosis. In its pure form , HSP is usually transmitted as an autosomal trait; most adult-onset cases are dominantly inherited. HSP has typically long term

How to cite this URL: Dr Chitte Om Virbhadra, Ayurveda: A Hope for HSP- Hereditary Spastic Paraparesis. International Ayurvedic medical Journal {online} 2016 {cited 2016 April} Available from: http://www.iamj.in/posts/images/upload/663_669.pdf

survival presumably because respiratory function is spared. Late in the illness, there may be urinary urgency and incontinence and sometimes fecal incontinence, sexual function tends to be preserved. In pure forms of HSP, the spastic leg weakness is often accompanied by posterior Colum (vibration & position) abnormalities and disturbance of bowel and bladder function. Some family members will have spasticity without clinical symptoms. In hereditary ataxias, an autosomal recessive inheritance is characterised by several affected members in one generation with healthy parents. Presence of consanguinity in parents strongly favours such inheritance. Typically these disorders manifest in childhood, adolescence or early adulthood. However a negative family history does not always rule out a hereditary disorder[ii]. Defects at numerous loci underlie both dominantly and recessively inherited forms of HSP. More than 30 HSP genes have now been identified. The gene most commonly implicated in dominantly inherited HSP is spastin. The most common childhood onset dominant form arises from mutations in the atlastin gene[iii].

Aims and objectives

1. To study about Hereditary spastic paraparesis, its pathological manifestations, symptoms in detail.
2. To assess the effect of Panchakarma therapies in Hereditary spastic paraparesis.

CASE REPORT

Preliminary Data of Patient: A 17-year-old female patient Hindu by religion belonging to middle socio-eonomic status without any premorbid status such as Diabetes mellitus & Hypertension approached to OPD (No- 14588) IP No (2510/15) of SKAMCH & RC, department of Panchakarma to DR. SWATI S. DESPANDE, HOD & Panchakarma consultant at KAMCH & RC- with complaints of Gait disturbance from the age of 5 years and Nystagmus since childhood. She is the second child(FTND) of her parents was born through 2ºconsanguineous marriage. Her mother's pregnancy was uneventful and her mother had taken folic acid & calcium supplements prescribed by the consultant gynecologist then. She was apparently normal till the age of 1½ years. She was having recurrent episodes of fever (once in 15 days) for which they took allopathic medications and got relief. Her milestones were slightly delayed. She started talking and walking at the age of 1½ years. She was given vaccinations as per schedule. When she was 5years old, her parents gradually noticed that she was falling frequently while walking and her legs were going sideways. She reduced playing with friend's outdoors as she lost her confidence in walking and running. Her mingling with friends remained normal. With the support of her friend she was able to walk confidently. At the age of 6 years she was taken to Orthopedic surgeon and he advised MRI scan with referral to Neurologist at NIMHANS. At NIMHANS, she was advised physiotherapy. They couldn't continue physiotherapy as there was no physiotherapist near to their home. At the age of 13 years she was taken to St. John's and got admitted there for 4 days. At St. John's some investigations were done and they advised some medications along with physiotherapy. As she didn't show any improvement even after few months, she was taken to Martha hospital(No improvement). Since then her parents took her to many other physicians and got no significant results. So they stopped going to doctors for the past 4-5 years. She presents with difficulty in walking with balance. Her bowel and bladder habits are regular and under control. There was tongue tie and she started talking after the surgery at 1 year of age. One elder sister aged 20 years is healthy. No family members is said to have similar complaints.

General Examination: On the day of examination found to be moderately built and nourished, afebrile, normotensive, coated tongue, Ht- 146.5 cms, Wt- 50kg, BMI-23.29, other parameters such as pallor, cyanosis, icterus, clubbing, lymphadenopathy, edema was normal.

Systemic Examination

- **CVS:** On auscultation; S_1 S_2 heard, No murmurs
- **RS :**
- Inspection
  Shape of chest - bilaterally symmetrical
  Respiratory rate – 16/min
- Palpation
  Trachea - centrally placed
- Auscultation
  Normal Vesicular Breath Sounds heard
- **PA**
- Inspection :
  No distension
  Umbilicus centrally placed
  No scars
  No visible peristalsis
- Palpation:
  Soft, Tenderness in right illiac & hypogastric region, No organomegaly
- **CENTRAL NERVOUS SYSTEM**
- Higher Mental Functions
  Consciousness – Fully conscious
  Orientation to - time, place & person-intact
  Memory - immediate, recent & remote-intact
  Intelligence - Intact
  Hallucination & Delusion - Absent
  Speech disturbance - absent
  Handedness - Right
- Cranial Nerve Examination
  Olfactory- Smell sensation-intact
  Optic - a) Visual acuity- intact
  b) Colour vision-intact
  c) Visual field- intact
  d) Light reflex- intact
  e) Accomodation-impaired

Occulomotor,Troclear & Abducent Nerve
-Eyeball movement-Possible in all directions in right eye. Left eye- nystagmus
-Pupil-position, shape, size & symmetry- NAD
-Ptosis-Absent

Trigeminal
Sensory-Touch, pain and pressure sensation intact
-corneal reflex-present
Motor-clenching of teeth -possible
-lateral movement of jaw- possible
Reflex-corneal-present
- jaw jerk- couldn't elicit

Facial

a)Forehead frowning	normal
b)Eyebrow raising	normal
c)Eye closure	possible
d)Teeth showing	no deviation of angle of mouth
e)Blowing of cheek	normal
f)Naso labial fold	equal on both sides

Vestibulo-cochlear
-Rinne's test-AC>BC
-Weber's test-no lateralisation of sound

Glossopharyngesl and Vagus
Position of uvula- centrally placed
Taste sensation- intact

Spinal accessory
Shrugging shoulder- possible against resistance
Neck movement -possible against resistance

Hypoglossal
Protrusion of tongue -possible
Tongue movements -possible

Motor System

1)Involuntary movements – Absent

2)Muscle bulk

Muscle bulk	Left	Right
Upper arm	30cms	31.5cms
Fore arm	20.5cms	21cms
Thigh	50cms	50cms
Calf	27cms	28cms

3)Muscle tone

Right hand	– Normal
Left hand	–Normal
Right leg	-Spasticity present
Left leg	–Spasticity present

4)Muscle strength	Rt.	Lt.
a) Elbow -Flexion	5/5	5/5
-Extension	4/5	4/5
b) Wrist -Flexion	5/5	5/5
-Extension	5/5	5/5
c) Finger abduction	5/5	5/5
d) Opposition of thumb	5/5	5/5
e) Test of grip	5/5	5/5
Lower limb	Rt.	Lt.
Hip -adduction	4/5	4/5
-abduction	4/5	4/5
-flexion	5/5	5/5
-extension	3/5	3/5
Knee -flexion	5/5	5/5
-extension	5/5	4/5
Ankle -dorsiflexion	5/5	5/5
-plantarflexion	5/5	5/5

5)Coordination

UL	Finger nose test Co ordination present
LL	Knee heel test

6)Involuntary movement- Absent

7)Gait- Waddling gait

Tandem walking - positive

Romberg sign - negative

Gover's sign - negative

8)Reflexes

Superficial

a) Corneal	Intact
b) Abdominal	Intact

	Rt	Lt
Deep		
a) Biceps jerk	++	++
b) Triceps jerk	++	++
c)Knee jerk	+++	+++
d)Ankle jerk	++	++
e) Clonus - patella	absent	absent
- ankle	absent	absent

f) Jaw jerk -	couldn't elicit
Plantar response- Babinski sign - +ve	

Measurements

LOWER LIMB	Right- 84 cms	Left-81 cms
UPPER LIMB	Right-68.5cms	Left-66.5 cms

Sensory system

1)Superficial:

Touch	present
Temperature	present
Pain	present

3. Deep:

Crude touch	present
Vibration	present
Joint sense	present
Position sense	present
Pressure sense	present

3) Cortical

a. Tactile localisation- present
b. Tactile discrimination- present
c. Stereognosis-present
d. Graphesthesia-present
e. Brisk Knee reflex- suggestive of upper motor neuron lesion
f. Babinski sign - +ve
g. Impaired tandem walking - Ataxia
h. Waddling gait - muscle weakness
i. Muscle bulk is same on both sides - excludes MD
j. Gover's sign - -ve
k. Limb length difference-suggestive of neurological conditions

Dashavidha pariksha

Prakruti	*- Kapha pitha*
Vikruti	*- Pravara*
Hetu	*- Beeja dosha (atlastin gene)*
Dosha	*- Vata, Pitha, kapha*
Dushya	*- Majja, Snayu, Sira ,*
Prakruti	*- Kapha pitha*
Desha	*- Saadharana*
Kaala	*- 12 yrs*
Bala	*- Rogi- madhyama*
Roga	*- Pravara*
Sara	*- Madhyama*
Samhanana	*- Madhyama*
Pramana	*- height-146.5cms,weight-50kgs*
Satmya	*- Madhyama (Madhura amla rasa)*
Satva	*- Madhyama*
Ahara sakthi	*- Abvyaharana sakthi-madhyama Jarana sakthi – madhyama*
Vyayama sakthi	*- Madhyama*
Vaya	*- Bala*

Samprapthi ghataka

Dosha	- Tridosha with vata pradhanatha
Dooshya	- Rasa, Mamsa, Snayu
Agni	- Jataragni
Srothas	- Rasavah, mamsavaha,majjavaha, asthivaha
Srotho dushti prakara	– Sanga
Udbava sthana	- Beeja doshaja

Vyaktha sthana	- Shaka
Adhishtana	- Masthishka
Marga	- Madhyama
Sadhyasadhyatha	- Yapya

Samprapthi: Beeja dosha, Due to beeja dosha Vata prakopa , Affecting majjavaha srotas, sira and snayu Resulting in weakness of both legs and gait disturbance

Differential **Diagnosis**

Disease	Inclusion Criteria	Exclusion criteria
Multiple sclerosis	Spasticity f lower limbs, ataxia, Nystagmus	Complaints didn't occur as episodes, Band sensation around limbs, episodes of limb paraesthesiae , Lhermitte's sign were absent.
Ataxia telangiectasia	Ataxia	Telangiectasias,Recurrent sinus & respiratory infections, delayed development of motor skills were absent.
Friedreich's Ataxia	Ataxia, Nystagmus, frequent falling	Deep tendon reflexes present. Titubation,Dysmteria , cariomyopathy absent
Hereditary Spastic Paraparesis	Chronic symmetric gait ataxia, Nystagmus, progressive spasticity and weakness in legs	

VYAYACHEDAKA NIDANA

Disease	Inclusion criteria	Exclusion criteria
Pangu	Both legs are affected	
Ardita	Netradinam vaikrutham	Ruja and other symptoms absent
Mamsagata Vata	sthabdhata	Sashoola granthi,dandamushti hatam, guru angatha,absent
Asthimajja gata Vata	Mamsa bala kshaya	Sandhi asthi shoolam absent
Anukta Vata Vyadhi	Sthabdhata of both lowerlimbs.	

Available Reports of investigations

MRI Spine - Normal

MRI Brain - Normal

Nerve conduction study - Normal

Diagnosis: The Dx of HSP is total history and clinical based Dx can be confirmed with the study of DNA & Gene.

Intervention

- Abhyanga with maha masha taila+ Mahanarayana taila followed by Nadi sweda for 14 days
- Mustadi Raja Yapana Basti for 8 days
- Shastika shali pinda sweda for 14 days
- Nasya with ksheerbala taila 101 (8 drops in each nostrils) for 7 days
- Balarishta 3tp tid A/F
- Dhanvantaram gutika 1-1-1
- Cardorium plus 2tp tid A/F
- T. BVC with gold 1-0-0 with honey

Post Panchakarma Treatment Status of the patient

The patient was hospitalised for one month and given Panchakarma treatment. There was significant change in the strength of lower limbs. She gained more confidence in walking with balance and the speed of walking improved.

The patient was discharged with some oral medication and physiotherapy was advised by SKAMCH & RC physiotherapy unit in charge. The patient comes for follow up walking herself and under supervision of consultant with oral medications.

DISCUSSION

In most cases of HSP, the primary problem may be disturbance of the ends of the long axons, with little or no loss of myelin and no abnormal myelin. The Abhyanga, Nadisweda and Shashtika Shali Pinda Sweda helps to reduce the spasticity and improves the balance of the patients while walking. Mustadi Raja Yapana Basti further helps in reducing the symptoms and gives a sustained relief to the patient.

CONCLUSION

Hereditary Spastic Paraparesis can be well treated in the lines of Vatavyadhi Chikitsa. The known complications of HSP are: Gastrocnemius-soleus contracture, Cold feet, Fatigue, Back and knee pain and Stress and depression. Hence the treatment should be planned according to the condition of patient aiming at improving the quality of life of patient. Panchakarma Chikitsa is definitely a ray of hope for patients suffering from HSP.

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