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Case Study

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MANAGEMENT OF PROGRESSIVE SUPRANUCLEAR PALSY- A NEURODEGENERATIVE DISEASE THROUGH AYURVEDA- A SINGLE CASE STUDY

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ABSTRACT

Progressive supranuclear palsy, also called as steele-richardson-olszewsky syndrome, is an rapidly progressive degenerative condition of unknown etiology affecting individuals in middle and late age. It resembles parkinsonism in the association of hypokinesia with rigidity, differing from it in that it is associated with a diagnostic disorder of ocular motility. This first affects vertical movements and conjugate lateral gaze involvement at a later stage. Early recurrent falls are typical. The annual incidence increases with age from 1.7 cases per 1,00,000 at ages 50 to 59 years to 14.7 per 1,00,000 at 80 to 89 years[1]. In this present case a 62 year old male presented at department of panchakarma on 3/5/2019 with diagnosis of PSP. For assessing disability, progressive supranuclear palsy rating scale(PSPRS) was used. This patient was treated with ayurvedic treatment modalities like snehana, swedana, shirodhara, mustadi raja yapana basti and oral medications. This has yielded into significant improvement in post treatment status of patient. This case study may present new possibilities of treatment and hope to the millions who is suffering from neurodegenerative diseases.

KEYWORDS: Progressive supra nuclear palsy, parkinsonism, panchakarma, Ayurveda, shirodhara, mustadi raja yapana basti.

INTRODUCTION

Progressive supranuclear palsy(psp), also known as steele-richardson-olszewski syndrome, is a neurogenerative disease that affects cognision, eye movements and posture. It was first described as a clinicopathologic entity in 1964. The disease was named progressive supranuclear palsy, referring to the progressive degeneration of the brain structures localized superior to the oculomotor nuclei, causing palsy and eventual paralysis of ocular movements. Onset in late 50's early 60's and Men affected slightly more often than women.

Risk factors and etiology

There are no proven risk factors for the development of psp except age. The cause of the psp remains unknown. The signs and symptoms of the disorder result from deterioration of cells in areas of brain, especially those that help to control body movements (mid brain) and thinking (frontal lobe). Both environmental and genetic influences have been postulated, but the findings have been inconclusive.

Genetic Susceptibility

Increased risk of psp for two independent variants of microtubule associated protein tau gene.

Pathophysiology

The disease is characterized by neurodegeneration, gliosis, and abnormal accumulation of tau protein in the basal ganglia, brain stem, prefrontal cortex, and cerebellum, the tau protein becomes resistant to proteolysis and is partially crystallized, forming abnormal deposits of tangled fibres, which are called neurofibrillary tangles.

Tau protein is important in maintaining neuronal morphology through microtubule binding. Under abnormal circumstances, the normally soluble tau protein may collect in insoluble protease resistant helical filaments. The exact triggers for the conversion from normal tau to the aggregate form are not completely understood.

Different rates, localizations and patterns of the accumulation of phosphorylated tau protein may account for the variation in clinical phenomena seen in patients with psp.

Clinical features[2]

- Most common classic phenotype of psp, is known as richardson phenotype or richardson syndrome, the most frequent initial feature is a disturbance of gait i.e., postural instability resulting in falls.

- Supranuclear ophthalmoplegia, Dysarthria, dysphagia, pseudobulbar palsy, rigidity, frontal cognitive abnormalities and sleep disturbances are additional common clinical features.
- Impaired postural reflexes (staggers backward uncontrollably).

Oculomotor Findings

- Supranuclear ophthalmoplegia or paresis is the hallmark of psp, but it may take as long as ten years to develop, average is three to four years. This distinctive ocular finding is first noted as slowing of vertical saccades, followed by a limitation of saccadic range.
- Impairment of convergence eye movements(may cause diplopia)
- Eyelid retraction, opening or closing apraxia, blepharospasm, or lid lag.

Motor Involvement

- Axial rigidity greater than appendicular rigidity, especially the neck and upper trunk.
- Prominent neck dystonia, dysarthria (monotone with slight hypo phonic quality).
- Absence of cog wheeling or tremor, widely based and unsteady gait.
- Bradykinesia with masked face and a startled expression.
- An absent, poor or rapidly waning response to levodopa is a characteristic feature of psp.

Cognitive and behavioural abnormality

- Slowed cognitive processing, sequencing, planning difficulty and apathy.
- Among behavioral abnormalities, the most common behavioral symptoms were apathy (91 %), disinhibition (36%), dysphoria (18 %), and anxiety (18%).

Neuroimaging

- Neuroimaging studies using CT and MRI of the brain demonstrate generalized and brainstem atrophy, particularly involving the midbrain.
- The radiologic "hummingbird "sign, results from the prominent midbrain atrophy with a relatively preserved pons, resembling a hummingbird on midsagittal MRI of the brain. And shows selective atrophy of the midbrain tegmentum with relative preservation of tectum and cerebral peduncles resembling the head of a mickey mouse. This is known as the 'mickey mouse sign', also called as 'morning glory sign'.

Diagnostic criteria[3]
(National institute of neurological disorders and stroke (NINDS)/ society for PSP)

Possible psp	Probable psp	Definite psp
• Gradually progressive disorder. onset at age 40 or later. • Supranuclear vertical gaze abnormalities with pursuit movements downwards or upwards with a restriction of less than 50% compared to normal or the two fallowing criteria:slowed verticle saccades and postural instability with falls in the first year of disease onset. • No evidence of other diseases that could explain the foregoing features, as indicated by mandatory exclusion criteria.	Obligatory presence of supranuclear vertical gaze palsy (upwards or downwards) and postural instability with falls in the first year of the disease.	Possible or probable psp with pathologic confirmation.

Management

There are no treatments that alter the natural history of disease in psp and no drugs that provide significant symptomatic benefits as seen with levodopa in idiopathic Parkinson disease. In some individuals the slowness, stiffness, and balance problems of PSP may respond to some degree to antiparkinsonian agents such as levodopa, but the effect is usually minimal and short-lasting.[4]

Ayurvedic treatment has been found effective in many neurodegenerative disorders and helped in minimising the symptoms of the same. Most of the neurodegenerative disorders present a picture of avarana which in turn leads to dhatukshaya resulting in vata prakopa. After considering this, the present case of psp was treated with panchakarma along shamanoushadhi & rasayana therapy

CASE REPORT

A male patient aged 62 years with no H/O systemic illness approached opd of dept. of panchakarma, SJIIM Bengaluru opd no. 6137 on 3/5/2019 who was already diagnosed with progressive supra nuclear palsy on 30/5/2018. His chief complaints were sluggishness in carrying out day-to-day activities, slowness in walking speed, imbalance of the body and disturbed gait since 2013. As per the information got from patient's wife, patient showed less interest in daily routine activities. Recurrent fall, tend to fall backwards, increased episodes of sudden unprovoked cry, gradual loss of memory specially related to his job i.e., tailoring since 2017 was absorbed. Patient also c/o rigidity in upper and lower

limbs, slurred speech and while walking feel any raised obstacles in his path. Initially patient neglected the condition but later after persistence of the symptoms he consulted neurologist in Victoria hospital, Bengaluru and diagnosed as progressive supra nuclear palsy and advised oral medications such as tab calcium 500mg, pantoprazole 40mg, optineuron, syndopa plus 125 mg and amantidine 100mg in 2018.

On observation, face was expressionless and mask like, has poor body balance with tendency to fall backwards. He was able to walk only with support and gait was slow and wide based. Has difficulty to sit and stand by self from a chair and sleep was disturbed. On systemic examination, vitals were normal. Mini mental status examination showed mild deterioration of cognitive functions. Cranial and ocular examination revealed diminished horizontal saccades but conjugate vertical gaze was affected more. Blink rate being 3 times per minute. MRI done on 30th may 2018 revealed age related atrophic changes in many regions of the brain.

Assessing subject on ayurvedic parameters revealed him to be of vata pittaja prakruti with madhyama sara and samhanana and normal jatharagni. The nidana noted were excessive consumption of katu, amla & lavana rasa, atichinta, krodha, udvega, late night working (ratri jagarana) and the symptoms observed like memory loss (smriti kshaya), spontaneous episodes of cry. Involvement of rasavaha, raktavaha, mamsavaha, majjavaha and manovaha srotas were noted.

Pre-intervention outcome measures

For assessing disability, progressive supranuclear palsy rating scale (PSPRS) was used. It comprises of 28 items in six categories: daily activities (by history), behaviour, bulbar, ocular motor, limb motor and gait/midline. Scores range from 0 to 100.

Intervention

After taking informed consent of the patient, was advised with Ajamodadi churna 5 gm BD with warm water & rasnaerandadi kashaya 15ml bd before food orally. Sarvanga Dashamoola kashaya parisheka was performed for 5 days. Patient was administered eranda taila 50 ml in empty stomach for vatanulomana & was advised shirodhara with Ksheera bala taila and brahmi taila for 7 days, followed by sarvanga abhyanga with Ksheerabala taila and swedana and Mustadi rajayapana basti in kala basti pattern for a span of 16 days. On discharge shamanoushadhi were advised.

Date	Treatment adopted	Dose & duration
From 3/5/2019	Ajamodadi choorna	5gm bd B/F
	Rasnaerandadi kashaya	15 ml bd B/F
4/5/2019 to 8/5/2019	Sarvanga dashamoola kashaya parisheka	Once a day for 5 days
9/5/2019	Kosta shodhana with eranda taila	50ml in empty stomach
11/5/2019 to 17/5/2019	Shirodhara with ksheera bala and brahmi taila fallowed by sarvanga abyanga with ksheera bala taila and swedana	For 7 days
19/5/2019 to 3/6/2019	Mustadi raja yapana basti with alternate anuvasana basti with ashwagandha gritha	16 days (kala basti)

No. of basti	01	02	03	04	05	06	07	08	09	10	11	12	13	14	15	16
date	19/5/2019	20/5	21/5	22/5	23/5	24/5	25/5	26/5	27/5	28/5	29/5	30/5	31/5	1/6	2/6	3/6/2019
Type of basti	A	A	N	A	N	A	N	A	N	A	N	A	N	A	A	A

On 4/6/2019 patient got discharged and medications on discharge were

Shamanoushadhi given	Dose	Duration
Tab. Brihat vata chintamani rasa	OD A/F	3 Months
Saraswatharista	15 ml bd with equal quantity of water,a/f	3 months
Ashwagandha avalehya	10gm, b/f	3 months

RESULTS

After the completion of the treatment, there was significant improvement in patient-

- Rigidity in limbs and neck was markedly improved
- Improvement was seen in activities of the daily routine.
- Speech was improved & ocular movements too.
- Patient was able to sleep for more than 6 hours.
- Marked improvement in facial expression and reduced episodes in recurrent crying.
- Improvement was seen in walking without aid and reduction in spontaneous fall.

Scores on PSPRS showed significant improvement. The patient scores in daily activities area reduced from 12 out of 24 to 8. In the mentation area scores reduced from 9 to 5 out of 16 points. Out of 8 for the bulbar area, scores reduced from 4 to 2. Scores in the ocular motor area improved from 6 to 3 out of 16. In the limb motor area which had a score of 16, score reduced from 7 to 4 and for the gait area scores reduced to 7 from 17 out of 20 points.

Showing comparison between pre and post intervention in progressive supranuclear palsy rating scale.

S. No.	Criteria	Pre- treatment score	Post treatment score (30 days)	Follow up (3months)
1.	Withdrawal	0	0	0
2.	Irritability	2	1	0
3.	Dysphagia for solids	0	0	0
4.	Using knife & fork, buttoning clothes, washing face & hands	3	2	2
5.	Falls	4	3	2
6.	Urinary incontinence	0	0	0
7.	Sleep difficulty	3	2	1
8.	Disorientation	3	2	1
9.	Bradyphrenia	2	2	1
10.	Emotional incontinence	2	0	0
11.	Dysarthria	2	1	1
12.	Dysphagia	2	1	1
13.	Ocular motor	2	1	1
14.	Voluntary upward command movement	2	1	0
15.	Voluntary downward command movement	3	2	1
16.	Voluntary left & right command movement	0	0	0
17.	Eye lid dysfunction	1	0	0
18.	Limb rigidity	2	1	1
19.	Limb dystonia	3	2	1
20.	Finger tapping	0	0	0
21.	Toe tapping	1	0	0
22.	Apraxia for hand movement	1	1	0
23.	Tremors	0	0	0
24.	Neck rigidity/ dystonia	2	1	1
25.	Arising from chair	4	3	2
26.	Gait	3	2	2
27.	Postural stability	4	3	2
28.	Sitting down	3	2	1
	Total score	54	33	21

DISCUSSION

Explanation of the exact mode of action of any treatment on modern parameters is difficult, however based on Āyurvedic classics, a hypothetical mode of action has been proposed below.

Vāyu is considered to be the cause of all type of movements of the body and plays a major role in the pathogenesis of various diseases.[4] It aggravates due to *Dhātuksaya* (tissue diminution) or by *Āvarana* (occlusion of its channel by other tissues).[5] Most of the neurodegenerative disorders present a picture of *Vāyu* dominant symptoms where *Vāyu* primarily gets aggravated due to *Āvarana* (occlusion), which in turn leads to *Dhātuksaya* (tissue diminution) leading to increased aggravation of *Vāyu*. This feedback mechanism make neurodegenerative diseases almost incurable, if treated late.

In the pathological process of PSP, aggregation of *tau* protein in the brain tissues can be correlated with one type of *Āvarana* leading to *Dhātuksaya* in the form of gliosis and neurodegeneration. In the present case, the initial pathology involved can be interpreted as *Kaphāvrta Udāna Vāyu* and *Kaphāvrta Vyāna Vāyu*,[6] which finally leads to *Dhātuksaya*.

At the time of presentation, patient's age, disease progression and MRI findings suggestive of atrophy in different parts of brain were indicative of pathology of *Vāyu*. Hence, treatment was started in accordance to the line of treatment for *Āvarana* (occlusion).charaka has stressed on srotoshuddhi, vatanulomana and rasayana in general management of avarana.

In the first step, treatment was started with kayaseka with dashamoola Kashaya to relieve the shola (pain) and stambha(stiffness) caused by kapha vata dosha, it is also considered as deepana pachana of aama and acts as balya along with this ajamodadi choorna and rasnaerandadi Kashaya are advised internally which helps in amapachana at the jataragni and dhatu level and helps in relieving shoolayukta vata dosha. Kosta shodhana with eranda taila is advised as it is the choice of treatment for aggravated *Vāyu*. It causes downward movement of *Vāyu* and hence helps to overcome the *Āvarana*.[7,8]

In a degenerative disorder to replenish the tissue elements, oleation therapy and rejuvenation therapy is

required and hence in the second step of treatment *shirodhara*, sarvanga abhyanga, bhaspa sweda, mustadi rajayapana basti and palliative treatment were administered with an aim to minimize the effects of gliosis and neurodegeneration.

Shirodhara is a process where medicated liquids like Kashaya, taila etc are poured on forehead continuously for a specific period. As the properties of thaila dhara include maintance of equilibrium in body and mind, enhances strength, taste in the food, memory and retaining power, prevents the deseases of eye, produces good sleep by stimulating the hypothalamus there by regulating the function of pituitary gland. So here shirodhara is done with ksheerabala and brahmi taila, as ksheera bala taila acts as jeevana, brimhana, swarya and anti-oxidant hence helps in the management of neurodegenarative deseases. brahmi is mentioned under medhya rasayana enhances memory, intelligence and longevity and also acts as acetylcholinesterase inhibitor & prove to be a useful memory restorative agent.[9] The tailapaka method enriches the alkaloid content of brahmi, so the penetrability of oil through the high vascular scalp is more than normal estimated 2%.

Sarvanga abyanga is adopted in this case as it helps to relieve the disease caused due to vatadosha and causes dridata to the body. As psp is kind of jara vyadhi abyanga is appreciated to be jarahara.

Basically the condition needs rasayana therapy, mustadi raja yapana basti plays very crucial role in the management of psp as it promotes bala, mamsa & shukra. It is sadyobalajanana & rasayana. Rajayapana basti which is having rasayana,balya and dhatu vriddhikara properties might have improved the excretory function of colon. Good regulation of blood flow under different condition is specific characteristic of colonic mucosa (kvietys 1980). It may also be possible that basti may be acting as stimulator for many intraluminal, luminal and whole body function. Regulatory peptides like serotonin, enteroglucogon, vasoactive intestinal polypeptides are produced in colon.administration of medicament in the liquid form or lipid soluble form into the rectum stimulates the rectum by distension or it may act systematically after getting absorbed by passive or active diffusion in mucous membrane of rectum. Thus the drugs absorbed into circulation can acts directly on neurons and modify neural function or they may reflexly acts by sending afferent impulses to central nervous system via chemo receptors, baro-receptors or the peripheral nerves.[10] The drug of yapana basti can also affect the nutrition due to its balya, brimhana and rasayana properties.

The Brihat vata chintamani rasa, the swarnakalpa acting like ‘chintamani’, the one who takes complete care in vatavikaras. It is excellent vatashamak, rasayan, balya and hruday samrakshak kalpa.[11] Saraswatarista enhances dhruti, medha,bala and kanti. It is paramaojaskara & best in reliving karkasha swara and aspasta bhashana.[12] ashwagandha avalehya acts as best rasayana and in sahasra yoga it is mentioned as ‘…ardhamasa prayogena kunjarena samam balam’.

CONCLUSION

Progressive supra nuclear palsy can be compared with kaphavruta vyana vayu and kaphanavryta udan vayu described in ayurvedic classics. The above mentioned combination of amapachana, sarvanga abhyanga and pariseka,shirodhara and mustadi rajayapana basti along with oral medications has given positive result in the patient of PSP. The main aim of the treatment in PSP is to improve the quality of life further preventing deformity. Psychological manifestations like irritability, emotional incontinence reduced drastically in the patient. Patient was symptomatically improved with rigidity, sleep difficulty, disorientation and got rid from frequent falls.

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