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The Role of a Speech Language Pathologist in Assessment and management of Communication Impairment in Progressive Supranuclear Palsy

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ABSTRACT

Progressive supranuclear palsy, also known as Steele-Richardson-Olszewski syndrome is a slowly progressive degenerative neurologic disease of later adult life. Clinical symptoms include gait difficulty, postural instability and falling. The vertical gaze palsy, parkinsonian symptoms, postural instability and pseudo bulbar palsy usually suggest the diagnosis of Progressive supra nuclear palsy.

Case Report: The case study involved three geriatric subjects (one female and two males), who had history of frequent falls, swallowing difficulties and poor clarity of speech. Following neuroimaging studies and clinical findings each subject was diagnosed as Progressive supranuclear palsy. Following a detailed case history assessment for speech language and swallowing skills using standardised tools; and evaluation of oral structures and their functions, were done. Each candidate was intervened for communication impairment and swallowing difficulties for 1.5-month period and outcome were discussed. Furthermore, we discussed about clinical presentation, assessment and treatment strategies for individuals with Progressive supranuclear palsy.

Since Progressive supra nuclear palsy later develops to dysarthria, dysphagia, drooling and pseudobulbar affect need for early diagnosis and rehabilitation of communication impairment is necessary.

Keywords: Progressive supranuclear palsy, compensatory strategy, communication impairment

1. INTRODUCTION

Progressive supranuclear palsy (PSP), also known as Steele-Richardson-Olszewski syndrome is a degenerative neurologic disease[9]. It is a slowly progressive disorder of later adult life exhibiting ocular and neurological signs and the cause of which is unknown. This rare multisystem degenerative disorder has a prevalence of 5-7 per 100,000 population[10]. It causes degeneration of neurons in Basal ganglia, Brainstem and Cerebellum. The most characteristic symptom is the gradual restriction of voluntary eye movements, which makes it difficult for the patients to walk downstairs, read or notice objects in their peripheral vision. Initial complaints often include gait difficulty[8], postural instability and falling[7]. The vertical gaze palsy, parkinsonian symptoms, postural

instability and pseudo bulbar palsy usually suggest the diagnosis of Progressive supra nuclear palsy. Progressive supra nuclear palsy later develops to dysarthria, dysphagia, drooling and pseudobulbar affect. Behavioural changes including irritability and depression, as well as cognitive changes are common[4]. Cognitive changes tend to be related to prefrontal lobe impairments[5]. The prevalence of PSP is higher in men[2] with mean age of onset is 63years. Men affected with PSP develops more frequent motor symptoms while women are associated with dysarthria and dysphagia symptoms[6].

In most cases, the genetic cause of progressive supranuclear palsy is unknown. Rarely the disease results from mutations in the MAPT gene[1]. The principal neuropathological findings in progressive supranuclear palsy are nerve cell loss, gliosis, neurofibrillary tangles and demyelination in basal ganglia, brain stem, and cerebellum.

Individuals with PSP develops mixed dysarthria after 2years of onset with a variable combination of spastic-hypokinetic-ataxic type. Additionally, they may also develop dysphagia leading to aspiration pneumonia[3]. These neurological impairments of person with progressive supra nuclear palsy will lead to communication deficiencies and swallowing dysfunction. Hence a systematic assessment and intervention needs to be carried out by a speech language pathologist who works with individuals with communication impairment and dysphagia to restore communication skills.

In this present study, we delineate clinical manifestation of three individuals with PSP and the role of speech language pathologist in evaluation and rehabilitation of neurodegenerative disorder like PSP.

2. METHOD

2.1 Participants

The study includes three geriatric subjects diagnosed as Progressive Supranuclear Palsy after neurological evaluation. The first subject (S1) was a female with 66 years of age with onset of condition since 1year, second (S2) and third (S3) subjects were males with 68 and 74 years with onset of condition since 2years and three years respectively. All the participants were presented with motor disturbances, history of frequent falls, poor rate and clarity of speech, and dysphagia.

Inclusionary criteria include:

1. Three subjects had postural instability with a history of frequent falls
2. Vertical gaze palsy
3. All subjects developed Mixed Dysarthria within 2years of post PSP
4. Dysphagia and drooling
5. Cognitive impairment like decreased verbal fluency, reduced abstract thinking and apathy (noticed in first and second subjects)

2.2 Test Material and procedure

Every participant who had known relatives was asked to provide an in-depth case history. Presenting complaints reported were history of recurrent falls, poor clarity and rate of speech, dysphagia and drooling. The speech-language pathologist conducted oral peripheral mechanism examination of structures and vegetative functions after reviewing the case history. Each subject underwent comprehensive speech language assessment using Frenchay Dysarthria Assessment (FDA), Western Aphasia Battery (WAB). Additionally, maximum phonation duration, Diadachokinetic rate (alternate and sequential motion rate), s/z ratio was calculated; perceptual assessment on intelligibility of speech, fluency and voice parameters were also documented. Manipal Manual for Swallowing Assessment (MMSA) was administered to rule out dysphagia. The diagnosis was confirmed with magnetic resonance imaging studies. In FDA using 9-point rating sacle eight categories including reflex, respiration, structural evaluation of lips, jaw, velum, laryngeal and tongue with its function (at rest and speech), intelligibility of speech in word, sentence and conversation level were documented. In WAB, assessed areas were spontaneous speech, auditory

comprehension, naming and repetition skills. Each individual's aphasia quotient was determined in order to rule out aphasia. Assessment of structures and functions (including sensory and motor evaluation), assessment of swallowing phases, and tolerance to food consistencies were the four subscales included in the MMSA test.

3. RESULTS

Information's on oral structural examination, their functions, vegetative skills, results of formal and perceptual assessment, assessment of dysphagia of three subjects are given in tables 1 to 4.

Table 1: Results of Oral Peripheral Mechanism Examination (OPME) and vegetative skills

Task	S1	S2	S3
OPME	All structures were found to be normal in size and shape; except lips (deviated to left) and asymmetric velum	All structures found to be normal; except lips (poor seal) and teeth (missing teeth)	All structures found to be normal; except tongue (deviated to left side), prosthetic teeth.
Assessment of functions	All functions (tongue, lips and velum) were found to be reduced in range and strength of movement.	All functions were found to be reduced in range and strength of movement. Drooling was present with inadequate	All functions were found to be severely affected. Severe drooling and he is not able to maintain intra oral breath pressure.
Vegetative skills	Mild difficulty with intake of solid foods	Mild difficulty with intake of liquid foods	Severely affected

Table 2: Outcome of formal assessment tools.

Test tools	S1	S2	S3
FDA	Mild Dysarthric component	Moderate level of dysfunction	Severe level of dysfunction
Remark	Dysarthria	Dysarthria	Dysarthria
WAB	Good language performance	Good performance in language task	All subtest were severely affected.
Aphasia Quotient (AQ)	97.6	97.4	4
Remark	Non- Aphasic	Non- Aphasic	Severe language impairment

Table 3: Results of perceptual judgement by SLP

Task	S1	S2	S3
MPD (vowel phonation)	10-12 seconds	6-10 seconds	1-2 seconds
Remark	Respiratory insufficiency	Poor respiratory function	Poor respiratory function
DDK (Iteration of syllables)	3 syllables/sec.	1 syllable/ sec	Not able to iterate consonants
Remark	Normal repetitive movement of oral structures	Reduced	Reduced
s/z ratio	0.7sec Reduced	Difficulty producing voiced consonant	Not able to perform task
Intelligibility of speech	Good in syllable to word production	Distorted articulation	Severely affected articulation. Able to produce low back open vowels
Voice	Breathy voice quality with reduced pitch and pitch breaks	Hoarse voice with reduced loudness	Severely affected
Fluency	Reduced rate of speech with repetition errors	Effortful and reduced rate of speech with articulatory freezing	Severely affected fluency skills.

Table 4: Clinical findings of Dysphagia through MMSA test

Sno.	Subscales	S1	S2	S3
Subscales 1	Assessment of Structure	All structures were found to be symmetrical except lips and velum (deviated to left)	All structures found to be in symmetry; except poor lip seal and missing teeth.	Structures found to be in asymmetry at rest
Subscales 2	Assessment of function- sensory evaluation	Scored 0	Scored 0	Scored 1
	Remark	She indicated sensations to respective articulator suggestive of normal sensory functions	He indicated sensations to respective articulator suggestive of normal sensory functions	Impaired as he was not able to indicate the sensation
	Assessment of function- motor evaluation	Scored 1	Scored 1	Scored 2
	Remark	Mild to Moderate impairment She had difficulty in velar function and pharyngeal wall movement	Moderate Impairment He had difficulty in performing all motor functions by oral structures.	Severe impairment

Subscales 3	Assessment of phases of swallowing	Scored 1	Scored 2	He was not able to perform task
	Remark	She had difficulty in swallowing of thick liquids and solid foods	He had difficulty maintaining lip seal and trouble swallowing thin, thick and solid foods	
Subscales 4	Tolerance of consistencies	Poor tolerance to solids	Poor tolerance to thin and thick liquids	Poor tolerance to all food consistencies
	Remark	Dysphagia in oral and pharyngeal phase	Dysphagia in oral and pharyngeal phase	Reported to have aspiration

Oromotor exercises, improve vegetative functions, respiratory skills to phonation task, improve loudness, intelligibility of speech, and overall communication enhancement were among the treatment options described in table 5 that were used and advised to the individuals for 1.5-month period.

Table 5: Outlines the treatment approaches and their corresponding outcome measures.

Subjects	**S1**	**S2**	**S3**
Baseline	Reduced tongue movements Dysphagia for solids	Poor lip seal and tongue movements Dysphagia for liquids	Severely affected functions with drooling
Treatment strategies 1	Oromotor Exercises		
Goal 1	Improve Tongue movements (elevation, protrusion and lateral movements)	Improve lip strengthening exercise and tongue movement	Improve lip, tongue function Strengthening exercises
Outcome	She is able to elevate and protrude tongue with normal range of movement	He is able to maintain lip seal with verbal instruction and improved in range of tongue movements	Reduced dribbling of saliva
Goal 2	Improve vegetative functions		
Outcome	Improvement in blowing, sucking and chewing skills	Improved in biting, and chewing skills	Poor improvement noticed
Goal 3	Advised for postural management (Guided for upright posture and body alignment)		
Outcome	Intake of pureed and semisolids improved	Reduced dribbling of food	Poor improvement noticed
Goal 4	Improve respiratory functions (task: MPD, relaxation and respiratory exercises)		
Outcome	Improvement noticed in sustained phonation task and rate of speech	Improved from baseline	Increase number of elicitation/ vocalisations of open vowels
Goal 4	Communication Enhancement		
Outcome	Good articulation in conversation, increased loudness	Reduced distorted errors of articulation with loudness increase	Improved non-verbal communication

4. DISCUSSION

Early signs of PSP were observed in S1, early identification and rehabilitation helped her to regain near normal vocal parameters, normal articulation and improvement in respiratory phonatory skills. S2 had a mid-stage sign of PSP as onset was reported since 2years and reassessment of FDA showed mild impairment. S3 had a post onset of more than three years and he had severe language impairment compared to other subjects. Communication training helped him improve his non-verbal communications (eye gaze pointing, verbal vocalisation). The caretaker was counselled regarding posture correction and head positing during oral feed. This helped in reduced frequency of aspiration in S3. Therefore, in order to enhance quality of life, speech-language intervention is crucial. Nevertheless, further study involving a greater number of subjects is required to define the language characteristics along with reading and writing deficits of these disorders.

5. CONCLUSION

Speech Language Pathologist works effectively in assessment and management as well as take part in counselling of individuals with neurodegenerative disorders. Speech language assessment also have utility for differential diagnosis of PSP from other neurogenic disorders.

Declaration by Authors

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