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Dysphagia

Dysphagia in Progressive Supranuclear Palsy: Radiologic Features

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Abstract. Progressive supranuclear palsy (PSP) is a progressive degenerative extrapyramidal disease that often masquerades as Parkinson's disease (PD). Similar to PD, dysphagia frequently complicates the course of PSP. Because there is only one published report characterizing dysphagia in PSP, we reviewed the neurologic features and dynamic videofluoroscopic swallowing function study results in 10 dysphagic PSP patients. Abnormalities during multiple stages of ingestion were recorded in each patient. Uncoordinated lingual movements, absent velar retraction or elevation, impaired posterior lingual displacement, and copious pharyngeal secretions were noted in all patients. Tongue-assisted mastication, noncohesive lingual transfer, excessive oral bolus lingual leakage to the pharynx prior to active transfer, vallecular bolus retention, abnormal epiglottic positioning, and hiatal hernias were noted in at least half of the cohort. Although ingestion abnormalities in PSP are similar to those previously reported in PD, the number of studied patients and observed differences were too few to clearly differentiate the two diseases.

Key words: Progressive supranuclear palsy — Parkinson's disease — Extrapyramidal — Dysphagia — Dysarthria — Bradykinesia — Gait disturbance — Rigidity — Deglutition — Deglutition disorders.

Progressive supranuclear palsy (PSP), a progressive degenerative disease of the central nervous system of unknown etiology, is one of the most common extrapyramidal syndromes masquerading as Parkinson's disease (PD). Disease onset is usually after age 50, with early presenting signs of bradykinesia, gait disturbance, and rigidity, and later complication of dysphagia and dysarthria [1–5]. However, impaired control of vertical eye movements and the relative insensitivity of basal ganglia signs and symptoms to dopamine replacement therapy are typical of PSP but not PD [6]. When tremor is present, it is almost always of a postural type rather than the typical rest or repose tremor of PD. Dementia, usually apparent at the time of diagnosis [2,3] may be present in up to 100% of PSP patients [7]. Other neuropsychologic abnormalities include personality changes, depression, and emotional lability of the pseudobulbar type [2]. Widespread neuronal loss in the basal ganglia accounts for the extrapyramidal signs while pontomedullary pathology [7] may account for some aspects of related dysphagia and dysarthria.

Multiple aberrations of ingestion have been recognized in PD [8–11]. We and others believe that bradykinesia and possibly rigidity alter eating behavior, mastication, lingual motility and transfer functions, laryngeal elevation and vocal cord movement, and all components of pharyngo-esophageal bolus transport [9,10]. Although dysphagia occurs in a majority of patients with typical PSP [7] and is the proximate cause of death in most cases [5], there is but one published report of swallowing function in this patient population [12]. We herein present our experience with dysphagia in patients with PSP.

Subjects and Methods

All dysphagic PSP patients meeting the clinical criteria of Lees [3] and followed at the Parkinson Disease and Movement Disorder Center of Crozer Chester Medical Center (CCMC) were included for review. Patients were excluded from the study if they had other central nervous system illnesses, such as stroke or metabolic encephalopathy, that could affect the clinical dysphagia or radiologic evaluation. The results for 10 patients, 7 men and 3 women, were tabulated. Ages ranged from

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63 to 83 (mean age = 70.9 ± 2.23) (Table 1). The Hoehn and Yahr (H&Y) scale [13] and the motor examination section of the Unified Parkinson Disease Rating Scale (UPDRS) [14] were used to estimate disease severity. The former scale ranges from 0 (no signs of disease) to 5 (wheelchair bound or bedridden). The H&Y group mean disease severity score was 2.7 (range 2–5); the disease duration was 4.1 ± 2.38 years (range 2–8). The UPDRS motor component, excluding resting tremor (range 0–88), was 20.6 ± 8.06; higher values represent more severe disease. At the time of each dysphagia evaluation, all but 3 patients were recieving an anti-parkinsonian medication (pergolide) with little symptomatic benefit. Every patient had previously received and discontinued carbidopa-levodopa due to lack of efficacy. A Mini-Mental status examination [15] was completed on 8/10 patients with a mean score of 24.7 ± 2.66. One patient not so tested was found to be moderately demented with formal neuropsychologic testing.

Dynamic Videofluoroscopic Swallowing Function Study (DVSFS)

A DVSFS was completed on all patients to confirm the location, severity, type, and cause of over dysphagic symptoms, and to identify any covert abnormalities. DVSFS performed by the CCMC Dysphagia Center were reviewed by one author (MCK). Mastication and transfer functions were assessed with barium-impregnated foods of varying textures ranging from pureed to solids, introduced in teaspoon amounts. Thin liquids followed, from teaspoon amounts to regulated cup and straw sips, advancing to unregulated continuous drinking. Mastication was classified into anterior munching/graded, lateral, or rotary patterns while viewed in the lateral and anterior-posterior (A-P) projections; pre-oral and oral activities were analyzed in the lateral projection. Pharyngo-esophageal transport and emptying functions were analyzed in both the lateral and A-P projections.

Results

Abnormalities were observed during several stages of ingestion in all patients. (Fig. 1) The oral preparatory stage abnormalities included graded mastication in all patients. In 3/10, this pattern was ineffectual, resulting in largely unmasticated food prior to active transfer initiation. In 9/10 patients, mastication was accompanied by forceful lingual pressing of food against the hard palate. Additional lingual function abnormalities included delayed or uncoordinated lingual movements in 10/10, with noncohesive transfer in 6/10, decreased posterior lingual displacement during the transfer in 9/10, and delayed or uncoordinated transfer in 10/10 patients. Absent velar retraction or elevation in 10/10 patients further obstructed or inhibited the transfer maneuver. Excessive posterior lingual bolus leakage was noted in 5/10 patients before swallow onset. Delayed swallow onset with food or liquids present in the valleculae occurred in 6 patients. During the pharyngeal swallow, supraglottic penetration was noted in 3/10 and glottic penetration in 2/10 patients. Frank aspiration was not observed. After the pharyngeal swallow, dysmotilities included vallecular retention in 4/10 and pyriform sinus retention in 3/10 patients. The epiglottic resting position appeared normal (vertical) in 5/10 and horizontal in 3/10, with absent displacement in 1 patient. Residua superior to the cricopharynx with adjacent cervical osteophytic spurs was noted in 1 patient. Copious, tenacious pharyngeal secretions were evident in every patient. Vocal cord appearance and motility were abnormal in 4/9 patients; this feature could not be assessed accurately in 1 patient. Normal vocal cord movement was seen in 5/9 patients, but delayed in 3/9 and absent in 1. Vocal cords were estimated to be enlarged in 4/10 patients.

Table 1. PSP patient data at time of dysphagia evaluation

Number	10
Sex	
Male	7
Female	3
Age	Mean ± STD 70.9 ± 2.23
Disease duration (yr)	Mean ± STD 4.1 ± 2.38
UPDRS motor score[a]	Mean ± STD 20.6 ± 8.06
Mini-Mental status score	Mean ± STD 24.7 ± 2.66

[a]UPDRS = Unified Parkinson Disease Rating Scale.

Multiple abnormalitieis of esophageal motility were recorded in all patients. Mild to moderate postswallow esophageal food residual and tertiary contractions were observed in all patients. In the lower esophagus, an open lower esophageal sphincter (LES) was noted in 2/10, a hiatal hernia in 6/10, slowed LES closure in 2/10, absent LES closure in 2/10, and gastroesophageal reflux disease (GERD), established by a water siphon test in 4/9 patients; 1 patient was not so tested because of an increased aspiration risk.

Discussion

This review of our experience with PSP highlights numerous abnormalities during multiple stages of ingestion. Our results are similar to those reported previously in PD [8–11]. Indeed, using videofluoroscopy alone, the motoric differences between PSP and PD are too few to distinguish the two diseases.

In PD, we previously described impaired pre-oral motor dysfunctions during observations of independent feeding including defective regulation of food and liquid quantities and impulsive feeding, primarily in our more severely affected patients. [10] Neumann et al. [16] recently described ‘‘mouth stuffing and rapid drinking’’ in at least half of their PSP subjects. Golbe [17] mentioned a similar type of pre-oral feeding pattern by noting that PSP patients had to be reminded ‘‘not to overload the fork and mouth.’’ Because observation of independent feeding was inconsistently completed, due in part to de-

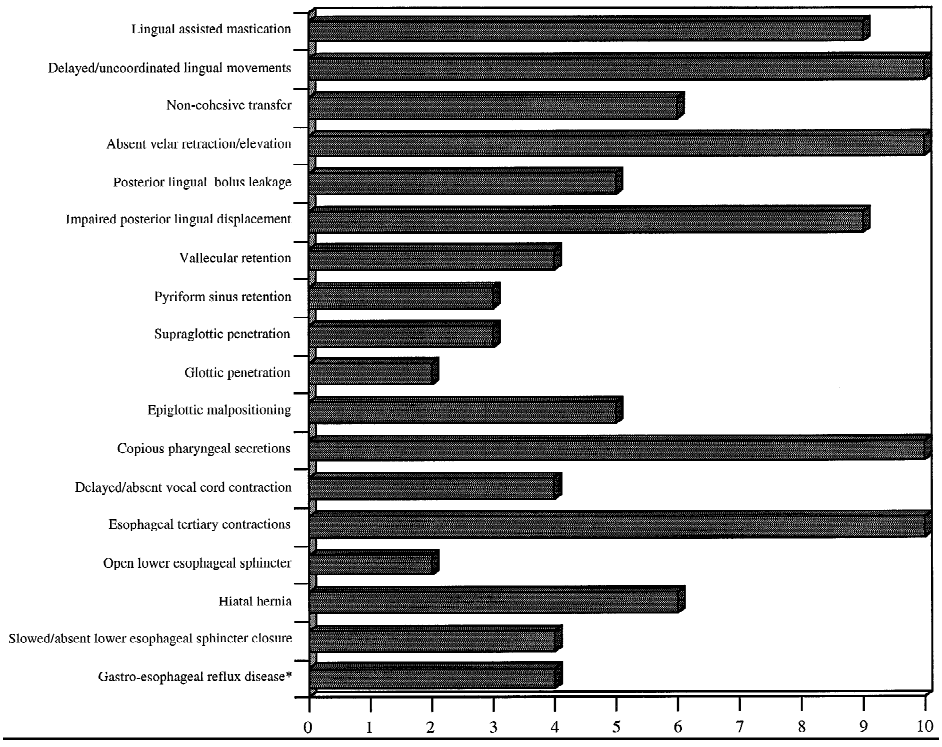

Fig. 1. Occurrence of abnormalities during ingestion in patients with PSP (n = 10). *Only 9 patients evaluated.

teriorated self-feeding skills or upper extremity instability, pre-oral ingestion stage abnormalities were not tabulated. When we retrospectively questioned our 4 surviving PSP patients and all surviving caretakers, no patient, but 4/7 caretakers acknowledged this behavior. This variant of tachyphagia in PSP and PD differs from that described by us in patients with Huntington's disease (HD) [18]. In bradykinetic-rigid syndromes, eating is relatively rapid and oral intake exceeds the expected transfer/swallowing frequency. Consequently, excessive food accumulates in the mouth waiting to be swallowed. In HD, feeding is rapid but a linkage between swallowing and feeding is maintained to the detriment of mastication. In each instance, the deficits increase choking and aspiration risk. Tachyphagia, at least in basal ganglia diseases, may reflect a sensorimotor integration disorder.

Both PSP and PD patients manifest impaired bolus preparation and tongue movements prior to swallow initiation. Posterior lingual bolus leakage, delayed swallow onset, impaired epiglottic and laryngeal movement, and ineffectual clearing of the vallecular and pyriform sinuses are likewise similar in both diseases. However, PSP transfer activity appears more adversely affected by velar elevation or retraction, an abnormality also noted by Sonies [12]. This abnormality may be partly related to nonextrapyramidal central nervous system pathology causing pseudobulbar speech and affective behavioral changes associated with PSP but not PD. Copious, tenacious pharyngeal secretions may also be more prominent in PSP.

Esophageal dysmotility including delayed, slowed transport and tertiary contraction waves with redirection, and lower esophageal sphincter abnormalities occurs commonly in PSP and PD. Indeed, all PSP patients in our series and 87% of our dysphagic PD patients [11] manifested esophageal dysmotility. Patulency was noted in 1/10 PSP patients and in 1/71 PD patients, reported previously [11]. The dorsal motor nucleus of vagus (DMNV), which dominates central nervous system motor control of the distal esophagus, is almost invaribaly depopulated and depigmented in PD. Involvement of this nucleus is either absent or minimal [7] in PSP suggesting that proposed esophageal dysmotility in basal ganglia diseases does not arise from a pathologic DMNV.

Although dysphagia complicates typical PSP, there is but one published report of aberrant ingestion. Sonies [12] conducted dysphagia evaluations including a videofluoroscopic swallowing study function of 22 PSP patients. Most patients were aware of their dysphagia, rarely displayed silent aspiration or lingual tremors, and manifested multiple oropharyngeal swallow videofluoroscopic abnormalities. Less than half of the investigated cohort had esophageal dysmotility. Using these characteristics, Sonies believed that PSP and PD could be differentiated. However, given the small number of PSP

patients with dysphagia assessments reported to date, we believe there is insufficient data to validate this separation. Also, we cannot confirm the contention that lingual tremors or a self-awareness of dysphagia segregate the PSP from the PD patient. In our experience, PD patients are also aware of their dysphagia, particularly if their disease is less severe and of shorter duration [10].

Dysphagia therapy in PSP is an arduous task made more difficult by several clinical factors that separate it from PD. Though both are progressive degenerative basal ganglia diseases, the course of PSP is much more rapid. The median survival is 6–8 years in PSP [2,4] and 13–14 years in PD [19]. Unlike PD, effective medications that may complement dysphagia therapy are unavailable for PSP. Levodopa improves deglutition in some PD patients [9] but has little if any impact on any aspect of the bradykinetic-rigid syndrome of PSP [2]. The characteristic severe limitation of downgaze and cervical dystonic neck posturing, sustaining the neck either in extension or the neutral erect position, handicap the PSP patient during meals; the former limits the ability to find food on the plate and the latter increases aspiration risk by limiting anterior neck flexion during the swallow. Both abnormalities also impede treatment efforts. In PD, cervical posture favors flexion, a feature that is likely to reduce aspiration risk. Finally, realized treatment benefits may be short-lived because of a progressive dementia that is almost universal in typical PSP [7] but that occurs in less than half of PD patients [20]. The dementia of PSP patients typically includes impaired memory, and slowed, defective motor planning and execution [21]. Consequently, clinical dysphagia management may require more frequent but shortened sessions. Despite these sensorimotor and cognitive limitations, dysphagia therapy remains warranted and welcomed by patients and caretakers.

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