



Treatment of Dysphagia in Parkinson's Disease

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Case

A 72-year-old right-handed man first developed a slight shuffling quality to his gait in approximately 2012. He also began to experience some difficulty adjusting his feet when standing on the golfing tee, preparing to hit the ball. However, he did not seek any medical evaluation until 2014, at which time he was referred for neurological evaluation because of increasingly noticeable neurological dysfunction.

The neurologist initially suspected the presence of a cervical myelopathy because of the presence of neck stiffness and pain, along with brisk lower extremity reflexes and bilateral Babinski responses. X-rays of the cervical spine demonstrated multilevel degenerative changes, but an MRI was deferred because the patient was claustrophobic. The neurologist commented that the patient was not experiencing any dysphagia or choking. Baclofen produced some transient benefit but was eventually discontinued. The patient was subsequently placed on a brief trial of carbidopa/levodopa 10/100 as a diagnostic test for possible Parkinson's disease (PD), but he only took the medication at a dose of one-half tablet once or twice daily for 3–4 days before discontinuing it because of some mental “foginess” and lack of benefit.

By mid-2015 the patient's gait dysfunction had progressed, and cervical fusion at the C3–C7 levels was undertaken. Postoperatively, the neck stiffness resolved, and there was some slight improvement in leg function. However, impairment of fine motor dexterity, particularly when performing repetitive tasks, deteriorating ability to type, and impairment of handwriting became progressively more

noticeable and reduced arm swing, and slight bilateral upper extremity tremor emerged. An MRI of the brain was unremarkable. Some difficulty swallowing had become evident even prior to the neck surgery, but following surgery, the dysphagia became even more pronounced, and the patient also developed some voice hoarseness.

The patient was referred to the movement disorders clinic, and by that time, the parkinsonian features were more readily apparent, and a diagnosis of PD was made. The patient was still experiencing dysphagia and at times would choke when swallowing. Carbidopa/levodopa 25/100 was initiated, with definite improvement in motor function within 1 month. He even resumed driving, which he had given up 8 months previously. However, his dysphagia did not improve. In addition to difficulty swallowing, the patient also mentioned that he would sometimes cough up or regurgitate undigested food or pills that he had swallowed up to an hour earlier. This description raised the question of a possible Zenker's diverticulum, and the patient was referred for gastroenterology evaluation.

A fluoroscopic upper gastrointestinal radiographic study was subsequently performed and demonstrated the presence of a large, wide-mouthed Zenker's diverticulum extending from the posterior aspect of the esophagus (Fig. 10.1). The mouth of the diverticulum measured approximately 1.8 cm, beginning at the C3–C4 level. The pouch measured approximately 2.3×1 cm, and a large amount of retained contrast was noted within the diverticulum throughout the examination. The distal esophagus appeared unremarkable and no gastric abnormalities were identified. Endoscopic surgery was subsequently performed, and the large, wide-mouthed Zenker's diverticulum was visualized, along with a prominent cricopharyngeal bar. The diverticulum was repaired and cricopharyngeal myotomy performed without incident, with subsequent resolution of the dysphagia.

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Fig. 10.1 A large, wide-mouthed Zenker's diverticulum, with a pouch measuring approximately 2.3×1.0 cm containing a large amount of retained contrast, extends from the posterior aspect of the esophagus

Discussion

Many patients with PD have some difficulty swallowing. The exact percentage is uncertain because large variability has been reported in different studies, probably owing to differing definitions of dysphagia and differences in the assessment tools utilized. Subjective survey studies suggest that anywhere from 30% to over 80% of persons with PD may have some awareness of swallowing impairment, whereas studies that use objective measures, such as radiographic or manometric techniques, arrive at higher figures of prevalence, often exceeding 95%. It also is evident from such studies, however, that not all individuals who have objective abnormalities involving swallowing actually experience or are aware of any difficulty swallowing. Studies even suggest that silent aspiration may be present in up to one-third of PD patients.

All three stages of swallowing – oral, pharyngeal, and esophageal – can be impaired in PD. Within the mouth, impaired function of the lips, tongue, and other oral muscles due to rigidity and bradykinesia can result in impaired bolus formation, delayed initiation of swallowing, the need for repetitive tongue pumping, and residual food being left in the mouth. In the pharyngeal phase of swallowing, pharyngeal dysmotility may lead to misdirection of swallows and residual food material being left in the vallecular and pyriform sinuses. A potentially serious complication due to these abnormalities is the development of aspiration. In fact, some

studies suggest that aspiration may occur in over 50% of persons with PD. Symptoms that suggest oropharyngeal dysfunction include difficulty keeping food in the mouth, hesitation in swallowing, a sensation of food sticking in the mouth or throat, nasal regurgitation, the need for multiple attempts to successfully swallow, painful swallowing, laryngeal abnormalities such as hoarseness or a “wet” voice after eating, and symptoms of aspiration such as coughing with eating or drinking. Recurrent episodes of pneumonia might also raise the question of aspiration. Oropharyngeal abnormalities can be best visualized with a modified barium swallow (MBS) study, in which the patient is asked to swallow both solid and liquid food mixed with barium under fluoroscopy. It is important to recognize, however, that although the MBS also visualizes the cervical esophagus, it does not adequately view the entire length of the esophagus.

Esophageal abnormalities have also been described in the setting of PD and may be present in up to 85% of patients. Esophageal dysfunction in PD may include slowed esophageal transit, segmental esophageal spasm, diffuse esophageal spasm, ineffective contractions, and even aperistalsis. Achalasia has also been reported in individuals with PD. A sensation of food sticking below the clavicle is suggestive of esophageal dysfunction. It is important to remember that the MBS does not adequately visualize the entire extent of the esophagus. Therefore, if the MBS does not reveal the cause of dysphagia in a person with PD, additional investigation with a barium esophagram, which employs liquid barium rather than food substances and visualizes the entire esophagus, should be performed.

Zenker's diverticulum actually is a false diverticulum that forms in the posterior aspect of the hypopharynx, just above the cricopharyngeal muscle, which serves as the upper esophageal sphincter. The cricopharyngeal muscle normally relaxes in response to the force of a food bolus and then constricts after the bolus passes in order to prevent reflux of the bolus. Formation of a Zenker's diverticulum is believed to be the result of inadequate relaxation of the cricopharyngeal muscle, with consequent increased intraluminal pressure just above the sphincter that eventually produces the outpouching that becomes a Zenker's diverticulum. An increased frequency of Zenker's diverticulum formation has been reported in patients with PD, presumably due to cricopharyngeal muscle dysfunction and perhaps even cricopharyngeal dystonia with consequent cricopharyngeal bar formation. In one study, 22% of patients with PD who were referred for evaluation of dysphagia were found to have either a cricopharyngeal bar or a Zenker's diverticulum. Symptoms suggestive of a Zenker's diverticulum include choking on food, regurgitation of food particles minutes or even hours after swallowing, bad breath, hoarseness, and a sensation of mucous in the back of the throat. A barium esophagram is the preferred study for visualizing a Zenker's diverticulum. Diagnosis of

Zenker's diverticulum is important because treatment in the form of surgical removal or closing of the diverticulum, along with cricopharyngeal myotomy, is curative.

Although curative treatment was available in this case and may also be in other instances, such as when anterior cervical osteophytes impinge on the esophagus or when dysphagia is caused by severe gastroesophageal reflux, for most PD patients with oropharyngeal dysphagia due to rigidity and bradykinesia, curative treatment approaches are not available. Some individuals will benefit from adjustment of anti-parkinson medication. For most, however, speech/swallowing therapy is the treatment of choice. Various swallowing techniques, including tilting the head, effortful swallows, and expiratory muscle strength training, may improve swallowing and reduce the risk of aspiration. Changing food viscosity by employing thickened liquids may also reduce aspiration potential. Treatment programs primarily designed to improve speech may also benefit swallowing.

Three clinical points can be made from this case. First, it is important to remember that dysphagia is a frequent gastrointestinal component of PD that may be present and pose risk to the patient even when the patient is unaware of it. Therefore, questions about its presence should be asked at every visit including to the caregiver. Second, it also is important to remember that, although dysphagia in PD most often is the consequence of impaired coordination of the oropharyngeal musculature due to rigidity and bradykinesia, other processes also may be responsible. In this case, it was a Zenker's diverticulum along with cricopharyngeal bar formation, but other processes also may be to blame, such as anterior osteophytes of the cervical spine impinging on the esophagus, severe gastrointestinal reflux, or even achalasia, in which the presence of Lewy bodies has actually been

reported. The third point to remember is that the pathology producing dysphagia in PD may not always be oropharyngeal in origin, but may reside in the esophagus. In such cases, a barium esophagram may be needed to visualize the pathology.

Suggested Reading

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