CLINICAL VIGNETTE

Early Dysphagia with Atypical Parkinsonism

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A 71-year-old right-handed male presented with dysphagia. He had intermittent difficulty swallowing solids for the previous 6 months. He denied difficulty initiating the swallowing process but felt like food got stuck several seconds after eating. He denied shortness of breath, vomiting, choking, early satiety, loss of appetite, a burning sensation in the chest, voice changes, or cough. On exam, he was well-nourished and in no distress with normal vital signs. His physical exam was normal, including detailed neurologic exam, with no tremor, normal gait and muscle tone.

In the 12 months after his initial presentation, he developed difficulty projecting his voice, his walking pace slowed, and he had one mechanical fall. He also reported poor energy and felt more depressed. His handwriting became smaller and harder to read. His swallowing worsened with liquids in addition to solids; he had several choking episodes.

His neurological exam had changed significantly. He had square wave jerks noted on picture gaze, impaired convergence with near objects and decreased blinking rate. He had moderate hypomimia and hypophonia. There was no dysarthria. He had moderate cogwheel rigidity of the left arm and his handwriting was small. His arm swing and stride length on the left were slightly diminished in comparison to the right. He had bradykinesia of the left greater than right when performing hand and toe movements and increased muscle tone in his neck.

Labs and Imaging

Labs included a normal comprehensive metabolic panel and a normal complete blood count.

Barium swallow showed frank aspiration with thin liquids as well as signs of gastroesophageal reflux to the level of the upper esophagus.

Brain MRI showed mild midbrain volume loss with possible hummingbird sign. It also showed chronic small vessel ischemic changes.

Initial Treatment Course

He was started on carbidopa/levodopa for presumed Parkinson disease (PD). The dose was titrated up without significant benefits from the medication. His bradykinesia and dysphagia did not improve.

He also started on a proton pump inhibitor to help manage the reflux noted on his barium swallow study but did not improve. He worked with a speech and swallow therapist, but despite diet modifications, his aspiration worsened with most meals.

His short-term memory worsened and was started on donepezil.

Discussion

Our patient had minimal response to carbidopa/levodopa therapy. This raised concerns about the initial diagnosis PD. As his symptoms progressed, square wave jerks and impaired convergence were noted on eye exam. Square wave jerks can significantly impair vision as the eye oscillates horizontally when attempting to fix gaze on an object.[1] These exam changes, his poor response to dopaminergic agents, and the midbrain volume loss on his MRI suggested progressive supranuclear palsy (PSP) as a more likely diagnosis. PSP patients have atrophy in the rostral midbrain that can resemble the bill of a hummingbird, thus the term "hummingbird sign" which was noted on his MRI.[2]

PSP is a four-repeat tauopathy that can lead to neuronal loss and neurofibrillary tangles primarily in the basal ganglia, cerebellum, and brainstem.[1] Classic features of PSP include: supranuclear gaze palsy, axial rigidity, dementia, and pseudobulbar palsy.[1] Supranuclear gaze palsy impairs vertical gaze and can impact the ability to look up or down.[1] The criteria for PSP include a progressive disease process that starts at or after age 40 that leads to a vertical supranuclear gaze palsy and/or slowing of vertical saccades with postural instability resulting in falls within the first year after onset of symptoms.[3] Histopathologic findings are needed for definitive diagnosis of PSP.[3]

Our patient was over 40 and had a fall within the first year of symptom onset. However, he did not have the classic eye findings associated with PSP when he initially presented. Litvan reported vertical supranuclear gaze palsy present on initial exam in a minority of patients with PSP.[4] It is important to consider PSP in patients with findings suggestive of PD who do not respond to dopamine agonists.

Our patient's initial presenting symptom was dysphagia. Dysphagia is often seen in more advanced stages of PD, but it can be seen earlier in some cases.[5] Determining when dysphagia starts can be challenging. Dysphagia in patients with PD is often silent with no obvious signs of aspiration, such as cough or voice changes.[6] This can lead to underreporting of dysphagia

for patients with PD and PSP as well as delays in implementing measures to help the dysphagia.[6]

Daniels reviewed the prevalence of dysphagia in patients with various neurological disorders[6] and estimated prevalence of dysphagia between 50-63% for patients with PD.[6] The prevalence of dysphagia for patients with PSP was 16% in the early stages and 83% in later stages of the disease.[6]

The mechanisms for dysphagia in patients with neurological disorders, such as PD and PSP, are likely multifactorial. Our patient started to have signs of dementia and cognitive decline by itself can impact the eating process. Muscle rigidity and bradykinesia of structures, such as the tongue, can slow the swallowing process and increase the risk of aspiration pneumonia.[5] Patients with PD also swallow more often during inhalation and at lower tidal volumes than healthy adults who generally swallow during exhalation.[7] Poor coordination of swallowing and breathing may be partly responsible for higher aspiration risk in patients with PD.[7] Litvan found pneumonia to be the leading cause of death for their patients with PSP.[4] Aspiration pneumonia is also a major cause of morbidity and mortality for patients with PD.[7]

Medications are generally of limited benefit for patients with PSP.[1] Even for PD, which has better responsiveness to dopaminergic agents, improvement in dysphagia with levodopa therapy is generally poor.[5] Several studies that showed mild improvement with levodopa postulated that the medication helped individuals with swallowing impairment affecting the oral phase but had less influence on the pharyngeal and esophageal phases.[5]

Without effective medications for dysphagia, along with the significant mortality and morbidity from aspiration pneumonia, swallowing therapy is important. Swallow therapy includes specific postures (such as a chin tuck posture), using thickened liquids, changing the taste of the food bolus, trying carbonation of liquids, and learning exercises that strengthen muscles involved in the eating process.[6] Though these interventions are not curative, they do offer some hope of decreasing aspiration events for individuals suffering from these progressive neurological conditions.

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