Case Study: Dysphagia among Older Adults

Amira Rana, MD, Medical Resident, Department Of Medicine, University of Toronto, ON.

Anselmo Mendez, BSc, BScN, Medical Student, Jagiellonian University, Medical College, Kraków, Poland.

Shabbir M.H. Alibhai, MD, MSc, FRCPC, Staff Physician, Department of Medicine, University Health Network; Assistant Professor, Departments of Medicine & Health Policy, Management, and Evaluation, University of Toronto, Toronto, ON.

Key words: myasthenia gravis, dysphagia, pyridostigmine, dysarthria, ptosis

Case Presentation

Mrs. K was an 88-year-old woman who arrived in the emergency department complaining of sudden onset of dysphagia that began three days previously when she started to choke on thin liquids and was unable to swallow solids. She denied having odynophagia. She also complained of associated difficulty speaking for 3 days. There was no complaint of limb weakness. At baseline she was ambulatory at home with the assistance of a cane. She denied any problems with swallowing or speech before this episode and also denied any bowel or bladder problems. She was admitted to the hospital with a presumed diagnosis of an acute stroke.

Mrs. K's past medical history was significant for hypertension and bilateral localized breast cancer that was diagnosed 10 years ago and treated with bilateral subtotal mastectomies.

On arrival at the hospital, she was in no acute distress and her vital signs were stable with a blood pressure of 138/70 mmHg and heart rate was regular at 85 beats per minute. Central nervous system examination revealed partial bilateral ptosis worsening toward the end of the exam. She had slurred speech with a nasal quality and difficulty swallowing clear liquids. Her dysphagia and dysarthria worsened with repetition. Motor exam revealed 5/5 power in the large muscles of upper and lower extremities except for 4/5 for shoulder abduction and hip adduction bilaterally. Her deep tendon reflexes were normal. Cranial nerve, cerebellar, and sensory examinations were normal. The rest of her review of systems was noncontributory.

Direct laryngoscopy was performed and did not reveal any glottic or cervical esophageal pathology. A swallowing study demonstrated aspiration of clear liquids and difficulty swallowing liquids primarily in the oropharyngeal phase.

Other investigations revealed a normal complete blood count and normal electrolytes, renal function, and thyroid studies. Magnetic resonance imaging of the brain did not reveal any evidence of acute stroke or space-occupying lesion. Since the initial basic workup did not reveal any identifiable cause and the patient's symptoms of dysphasia and dysphagia progressed, a consultant neurologist suggested neuroelectrophysiological studies that included repetitive nerve stimulation (RNS) and single-fibre EMG studies (SFEMG). Repetitive nerve stimulation studies were normal but the SFEMG studies were abnormal, indicating impaired neuromuscular transmission. This was followed by testing for acetylcholinesterase antibodies which were >8 nmol/L (normally <0.39).

Thus, the patient was diagnosed with myasthenia gravis (MG) on the basis of the clinical and neurodiagnostic evaluation. She was given plasmapharesis on alternate days along with oral pyridostigmine 60 mg daily and a short course of oral prednisone. A nasogastric tube was placed to initiate feeding as the patient was unable to swallow safely on her own. Her symptoms improved considerably within 2 weeks of starting the above therapy. The nasogastric tube was removed and the patient started to eat solids and liquids without difficulty. Her ptosis and phonation markedly improved and she was discharged home with follow up in the neurology clinic.

Discussion

Dysphagia is a common condition that family physicians frequently encounter among older adults. Although stroke is the leading cause of this disorder among older persons, other possibilities should be considered. The physician should first determine whether the dysphagia is consistent with esophageal, oropharyngeal, obstructive, or neuromuscular symptoms (Table 1, Figure 1). Proper classification leads to a successful diagnosis in up to 85% of patients.[1]

Myasthenia gravis is often underdiagnosed among older adults because it may present with a vague or unusual clinical pattern.[2] However, the prevalence of MG is greater than commonly appreciated. It is the most common disorder of neuromuscular transmission. The highlight of this disorder is a fluctuating degree and variable combination of weakness in ocular, bulbar, limb, and respiratory muscles. Weakness is the result of an antibody-mediated, T-cell-dependent immunological attack directed at the proteins at the postsynaptic membrane of the neuromuscular junction (acetylcholine receptors and/or receptor-associated proteins). More than 50% of individuals present with ocular symptoms of ptosis/diplopia, and half of these individuals eventually develop generalized weakness within 2 years. About 15% present with bulbar symptoms such as dysphagia, dysarthria, and fatiguable chewing. Less than 5% present with isolated proximal limb weakness.[2]

Table 1: Differential Diagnoses of Dysphagia

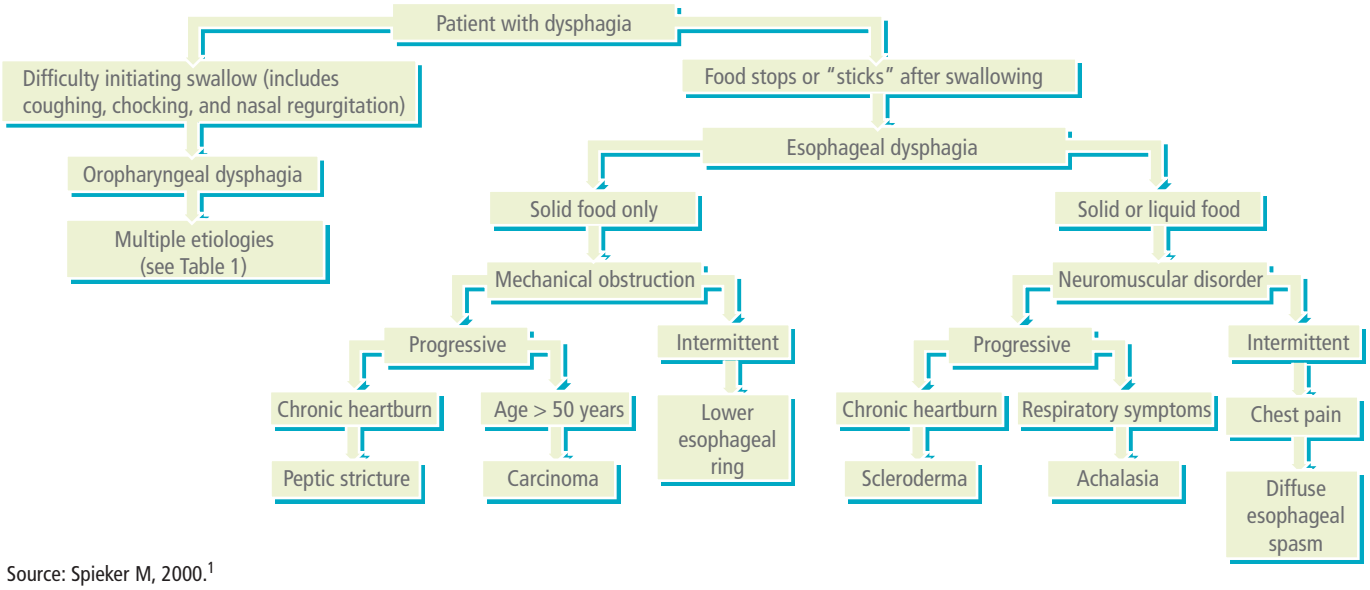
Oropharyngeal dysphagia	Esophageal dysphagia
Neuromuscular disease	*Neuromuscular disorders*
Diseases of the central nervous system	Achalasia
Cerebrovascular accident	Spastic motor disorders
Parkinson's disease	Diffuse esophageal spasm
Brain stem tumours	Hypertensive lower esophageal sphincter
Degenerative diseases	Nutcracker esophagus
Amyotrophic lateral sclerosis	Scleroderma
Multiple sclerosis	*Obstructive lesions*
Huntington's disease	Intrinsic structural lesions
Poliomyelitis	Tumours
Syphilis	Strictures
	Peptic
Peripheral nervous system	Radiation-induced
Peripheral neuropathy	Chemical-induced
Motor end-plate dysfunction	Medication-induced
Myasthenia gravis	Post-infectious
Skeletal muscle disease (myopathies)	Lower esophageal rings (Schatzki's ring)
Polymyositis	Esophageal webs
Dermatomyositis	Foreign bodies
Muscular dystrophy (myotonic dystrophy, oculopharyngeal dystrophy)	Extrinsic structural lesions
Cricopharyngeal (upper esophageal sphincter), achalasia	Vascular compression
Obstructive lesions	Enlarged aorta or left atrium
Tumours	Aberrant vessels
Inflammatory masses	Mediastinal masses
Trauma/surgical resection	Lymphadenopathy
Zenker's diverticulum	Substernal thyroid
Esophageal webs	
Extrinsic structural lesions	
Anterior mediastinal masses	
Cervical spondylosis	

Source: Spieker M, 2000.[1]

Single-fibre electromyography studies are a sensitive diagnostic test for neuromuscular transmission abnormalities and the frontalis muscle has great value for jitter recordings. Jitter is the variability in time of the second action potential relative to the first when two muscle fibres innervated by the same axon are stimulated. In our case the test revealed 88% abnormal pairs of muscle fibres (normal is <10%).

Treatment is based on severity of disease and ranges from the use of corticosteroids and anticholinesterases (such as pyridostigmine) to emergency measures such as plasmapheresis and intravenous immunoglobulins.[2]

Figure 1: Evaluating Dysphagia



Source: Spieker M, 2000.[1]

Conclusion

If an older individual presents with symptoms and signs of oropharyngeal dysphagia and neuroimaging does not reveal a stroke, the clinician should investigate for possible neuromuscular disorders. Diagnostic investigations include:

1. Tensilon™ test: Edrophonium hydrochloride is an acetylcholinesterase inhibitor. It can be used for patients with obvious ptosis or ophthalmoplegia[3] but use should be avoided among older people because of the risk of symptomatic bradycardia or bronchospasm, especially for individuals with heart disease or asthma, respectively. Therefore, this test was not performed in our case.
2. Repetitive nerve conduction study and single fibre EMG can narrow the differential and rule in/rule out neuromuscular junction disorders.
3. Acetylcholinesterase antibodies are positive in 80–85% of persons with generalized MG and 55% of those with ocular myasthenia.[2,4]
4. MuSK antibodies: Antibodies to muscle specific receptor tyrosine kinase (MuSK) is positive in 40–50% of patients with seronegative MG. This test, however, is not routinely available.[4]
5. Once MG is confirmed by the above clinical and diagnostic criteria, a CT scan of the chest is indicated to rule out a thymoma.

The successful treatment of dysphagia depends on the careful investigation of the cause of the disorder. Older adults are especially at risk of misdiagnosis because of subtle symptoms and the presumption that stroke is the most likely causative factor for dysphagia among older individuals. The case study demonstrated that the correct diagnosis led to directed management resulting in prompt improvement of the patient's clinical symptoms and signs.

No competing financial interests declared.

References

1. Spieker MR. Evaluating dysphagia. Am Fam Physician 2000;61:3639–48.
2. Vincent A, Clover L, Buckley C, et al. Evidence of underdiagnosis of myasthenia gravis in older people. J Neurol Neurosurg Psych 2003;74:1105–8.
3. Daroff RB. The office Tensilon test for ocular myasthenia gravis. Arch Neurol 1986;43:843–4
4. Meriggioli MN, Sanders DB. Myasthenia gravis: diagnosis. Semin Neurol 2004;24:31–9.

Key Points

- Myasthenia gravis tends to be underdiagnosed among older adults presenting with dysphagia.
- Keys to diagnosis are a good history and careful physical assessment.
- Among older adults, acetylcholinesterase antibodies, EMG, and, in some cases, MuSK antibodies are important for diagnosis.
- CT scan of the chest should be done once MG is confirmed to rule out thymoma.
- Treatment depends on severity ranging from anticholinesterases and corticosteroids to emergency plasmapheresis and intravenous immunoglobulins.