A Case of Amyotrophic Lateral Sclerosis Presented as Oropharyngeal Dysphagia

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Amyotrophic lateral sclerosis is a rare disease. It is a fatal neurodegenerative disease characterized by progressive muscular paralysis reflecting degeneration of motor neurons which leads to muscle weakness and muscle wasting. Respiratory failure limits survival to 2-5 years after disease onset. Several clinical manifestations including dysphagia can result in reductions in both the quality of life and life expectancy. Dysphagia occurs at onset in about one third of case, although generally it occurs in later stage of the disease. Evaluation of dysphagia includes video-fluoroscopic swallow study, radiological esophagogram, flexible endoscopic examination, ultrasound examination, conventional manometry and electromyography. We report a case of amyotrophic lateral sclerosis in a 54-year-old man presenting oropharyngeal dysphagia which was diagnosed by high resolution esophageal manometry presenting abnormality of the upper esophageal sphincter.

Key Words
Oropharyngeal dysphagia, Amyotrophic lateral sclerosis, High resolution manometry

Introduction

Amyotrophic lateral sclerosis (ALS) or Lou gehrig’s disease is a relatively rare disease. The incidence is about 1-2 per 100,000. It is a fatal neurodegenerative disease characterized by progressive muscular paralysis reflecting degeneration of motor neurons in the motor cortex, corticospinal tract, brainstem and spinal cord. It leads to muscle weakness and muscle wasting due to unsend messages to muscles. Respiratory failure limits survival to 2-5 years after disease onset. Dysphagia is one of the frequent features of ALS and can result in reductions in life expectancy and quality of life. It can lead to nutritional deficiency and aspiration pneumonia. This symptom occurs at onset of disease in about one third of case, although generally it occurs in later stage of the disease.

Neurogenic dysphagia can be evaluated by video-fluoroscopic swallow study, radiological esophagogram, flexible endoscopic examination, ultrasound examination, conventional manometry and electromyography. High resolution manometry (HRM) has been recently developed and defined the intraluminal pressure of the esophagus more completely than conventional manometry and is very useful and safe in evaluating oropharyngeal, neurogenic dysphagia.

We report a case of an amyotrophic lateral sclerosis in a 54-year-old man presenting oropharyngeal dysphagia which was evaluated by high resolution esophageal manometry.

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Conflicts of interest: None.
Case Report

A 54-year-old man was admitted to the hospital complaining of dysphagia and hoarseness of 3 months. He developed weakness in both arms 1 year before the admission, which has disabled him for lifting a heavy load over his head. He experienced shortness of breath when going up a hill or stairs. But he had no difficulty in swallowing during the initial hospital visit. Gradually, he developed difficulty in lifting his hands over his head and repeatedly stopped level walking due to shortness of breath. Thereafter, from 3 months before admission, he could not lift up his arms to the chest level, and dysphagia occurred. He had difficulty in swallowing porridge and water, and had to spit because he could not swallow his saliva. He couldn’t sleep well because of shortness of breath, which led him to stay awake every night for 1 to 2 hours. He was admitted for evaluating dysphagia and hoarseness. There was no specific findings on medical history or familial history. Vital signs were stable. He was alert, but appeared chronically ill. Nutritional status was poor and he complained of swallowing difficulty. Both upper proximal extremities showed Medical Research Council grade 3-4 weakness and especially, both arm abduction were graded 1-2.

Laboratory studies showed the following parameters: white blood cell count 10,900/mm³, hemoglobin 16.1 g/L, platelets 434,000/mm³, serum sodium 133 mEq/L, serum potassium 4.2 mEq/L, serum creatinine 1.0 mg/dL, serum blood urea nitrogen 12 mg/dL, serum albumin 4.3 g/dL, serum lactate dehydrogenase 307 IU/L, serum erythrocyte sedimentation rate 20 mm/hr and serum C-reactive protein 3.9 mg/dL. In addition, rheumatoid, anti-nuclear antibody, anti-DNA antibody, anti-neutrophil cytoplasmic antibody, anti-extractable nuclear antibody, anti-Ro/SSA antibody and anti-La/SSB antibody showed negative results with no abnormality on the chest and abdominal CT scan.

Neurologic examination revealed jaw jerk signs and positive Hoffman sign. The deep tendon reflexes of both the upper and lower limbs were positive for upper motor signs with atrophy of the tongue. Patient presented symptoms such as swallowing difficulties for liquids and solids, bulbar weakness signs of hoarseness, muscle atrophy and weakness in the trunk, neck, back and both proximal upper limbs for lower motor signs. Brain MRI was recommended but refused by the patient.

Esophagogastroduodenoscopy revealed neither a mucosal abnormality nor an unstained area by lugol spray on the esophagus and the scope passed easily through the lumen. However, we had difficulty in performing endoscopy because of the hypoxemia developed during the procedure and poor cooperation of the patient. Esophagogram showed the retention of contrast agent in the upper esophageal sphincter, which did not pass down after swallowing, and thus, remained in the pharynx and was aspirated into the airway. Nerve conduction velocity revealed sensorimotor polyneuropathy, and electromyography showed increased insertional activity, fibrillation on rest and a positive sharp wave in the first dorsal interosseous muscle, extensor digitorum communis muscle, biceps brachii muscle, deltoid muscle and tibialis anterior muscle. Additionally, the interference pattern was decreased in all of these muscles. Acute degenerations and chronic changes in widespread distribution confirmed the lower motor neuron injury. On video fluoroscopic swallow study (VFSS), movements of the tongue in the oral phase was intact and there was no sign of tongue apraxia. Swallowing reflex in the pharyngeal phase was slightly impaired and aspiration without coughing was observed for thin liquid, thick liquid, semisolid and solid. Grade 2 and 3 residue was observed in the valleculae and piriform sinus, respectively (Fig. 1).

High resolution esophageal manometry were done in a supine position after at least 8 hours of fast. The manometric protocol included a 5-minute period to assess basal sphincter pressure and 10 series of 5-mL water swallows. The patient performed only four successful 5-mL water swallows because of the dysphagia. On HRM, Upper esophageal sphincter (UES) was relaxed after...