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Evaluation of dysphagia at the initial diagnosis of amyotrophic lateral sclerosis

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ABSTRACT

Objective: Dysphagia eventually occurs in amyotrophic lateral sclerosis (ALS). Swallowing in patients with ALS at their initial diagnosis was evaluated using videofluoroscopy (VF).
Methods: Nineteen consecutive patients with ALS, 14 with bulbar symptoms, and 5 without them, underwent VF. Fourteen physiologic components, 6 oral and 8 pharyngeal components, were assessed during the examination.
Results: Significantly poorer scores were observed in three of the 6 oral components and 3 of the 8 pharyngeal components in patients with bulbar symptoms. Furthermore, bolus transport from the oral cavity to pharynx, pharyngeal constriction, oral residue and pharyngeal residue were impaired in patients even without bulbar symptoms. On the other hand, pharyngoesophageal segment opening was preserved in patients even with bulbar symptoms. Bolus transport and initiation of pharyngeal swallow were correlated with the swallowing category of the ALS severity scale.
Conclusion: Defining types of impairment in patients with or without bulbar symptoms is useful for evaluating dysphagia in this disease. Although VF showed impairment of oral and pharyngeal phases of swallowing, the oral phase affected the eating habit in ALS at the initial diagnosis.

1. Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disorder that primarily involves the motor neurons in the cerebral cortex, brainstem, and spinal cord [1]. The degeneration of upper motor neurons results in symptoms that include increased deep tendon reflexes and a manifestation of pathologic reflexes. Degeneration of the lower motor neurons causes muscular atrophy, decreased muscle force, fasciculation, and bulbar palsy. As the disease progresses, both upper and lower motor neurons may be affected; therefore, various symptoms overlap. Among them, dysphagia eventually occurs independent of the time of onset [2].

Dysphagia is one of the most critical problems for ALS patients, which leads to serious nutritional deficit and results in aspiration pneumonia [2]. Therefore, careful follow-up of the clinical condition of dysphagia is required to decide on the appropriate timing of intervention. Furthermore, because the dysphagia of ALS is generally progressive and symptoms vary, evaluation of the swallowing function at the initial diagnosis is inevitable.

Videofluoroscopy has been the most reliable examination to evaluate the swallowing function of ALS patients [3]. It facilitates accurate analysis of the mechanisms of the oral and pharyngeal phases of swallowing, identification of possible changes responsible for symptoms, and planning of appropriate therapy. The aim of this study was to assess the role of videofluoroscopy in identifying swallowing abnormalities, especially from the perspective of oral and pharyngeal dysphagia, at the initial diagnosis of ALS.

2. Materials and methods

2.1. Patients

Nineteen patients were diagnosed with ALS by certified neurologists based on El Escorial criteria at the Department of

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Table 1
Summary of the swallowing category of the ALS severity scale proposed by Hillel et al. [4].

Swallowing	Rating
Normal eating habits	
Normal swallowing	10
Nominal abnormality	9
Early eating problems	
Minor swallowing problems	8
Prolonged time or small bite size	7
Dietary consistency changes	
Soft diet	6
Liquefied diet	5
Needs tube feeding	
Supplemental tube feedings	4
Tube feeding with occasional oral nutrition	3
Nothing by mouth	
Secretions managed with aspirator/medication	2
Aspiration of secretions	1

ALS, amyotrophic lateral sclerosis.

Neurology in our hospital between 2004 and 2011. They were referred to our department immediately after the diagnosis to evaluate the swallowing function. The onset of bulbar symptoms was reported by patients, and bulbar symptoms were evaluated by neurologists, otolaryngologists, and speech therapists. The status of the dietary intake was evaluated using the swallowing category of the ALS severity scale (ALSSS) (Table 1) [4]. This study was approved by the Ethics Committee of Kanazawa University School of Medicine.

2.2. Videofluoroscopic assessment

Videofluoroscopy was carried out in all patients at the time of the initial diagnosis of ALS. Three milliliters of a 140% (v/v) barium mixture was basically used; however, other contrast agents were also used based on the circumstances. All 15 physiologic components except for a component of esophageal clearance, proposed by Martin-Harris and Jones, were thoroughly evaluated from the perspective of all six oral components and all eight pharyngeal components [5] (Table 2). In addition, we also evaluated the oral and pharyngeal residues. Each component was graded from 0, which was considered normal, to 3 in the present study. Each score was obtained with the average of blind reviews by one otolaryngologist and one speech therapist.

Simultaneously, the penetration-aspiration scale (PAS), an 8-point scale to quantify selected aspects of penetration and aspiration in order to clarify the depth of airway invasion and whether or not there is material entering the airway, was defined for the brief evaluation of dysphagia (Table 3) [6].

2.3. Statistical analysis

The difference in scores between patients without bulbar symptoms (BS-negative) and those with symptoms (BS-positive) was compared using the Mann–Whitney *U*-test. A correlation between two variables was analyzed using Spearman's rank correlation. All analyses were carried out using SPSS 19.0 software (SPSS Inc., Chicago, IL, USA). In all tests, $p < 0.05$ was considered significant.

3. Results

3.1. Patients

Nineteen patients were evaluated, consisting of 12 men and 7 women. The mean and median ages of the 19 patients were 64 and 66 years old, respectively, ranging from 44 to 79 years old. Physical examinations by a neurologist and otolaryngologist at the initial presentation revealed 5 patients with disorder of only the extremities, 6 patients with only bulbar disorder, and 8 patients with a combination of both disorders. The mean duration from the onset of symptoms to the diagnosis of ALS at our hospital was 8.0 months for bulbar symptoms and 11.1 months for other symptoms involving the limbs, showing a significant difference ($p < 0.05$).

3.2. Oral swallowing impairment on videofluoroscopy

The mean score of each oral component is shown in Table 2 and Fig. 1. The components of Lip Closure (LipC), Lingual Elevation (LingE), and Tongue to Palatal Seal (TPS) were preserved in both BS-negative and BS-positive patients. However, both components of Bolus Preparation/Mastication (BP) and Initiation of Pharyngeal Swallow (IPS) were preserved in BS-negative patients while they

Table 2
Physiologic swallowing components.

Component	Abbreviation	Mean score ± standard deviation		*p*-value
		Bulbar symptoms (−) ($n = 5$)	Bulbar symptoms (+) ($n = 14$)	
Oral				
1. Lip Closure	LipC	0.00 ± 0.00	0.21 ± 0.42	0.27
2. Lingual Elevation	LingE	0.20 ± 0.44	0.28 ± 0.72	0.88
3. Tongue to Palatal Seal	TPS	0.20 ± 0.44	0.28 ± 0.72	0.88
4. Bolus Preparation/Mastication	BP	0.00 ± 0.00	0.57 ± 0.51	0.03
5. Bolus Transport/Lingual Motion	BT	0.40 ± 0.54	1.14 ± 0.66	0.04
6. Initiation of Pharyngeal Swallow	IPS	0.00 ± 0.00	0.93 ± 0.73	0.01
Pharyngeal				
7. Soft Palate Elevation and Retraction	SPE	0.00 ± 0.00	0.43 ± 0.51	0.08
8. Laryngeal Elevation	LaryE	0.00 ± 0.00	0.71 ± 0.61	0.01
9. Anterior Hyoid Excursion	AHE	0.00 ± 0.00	0.93 ± 0.61	<0.01
10. Laryngeal Closure	LaryC	0.00 ± 0.00	0.43 ± 0.51	0.08
11. Pharyngeal Contraction	PC	0.60 ± 0.54	1.07 ± 0.61	0.14
12. Pharyngoesophageal Segment Opening	PESO	0.00 ± 0.00	0.21 ± 0.42	0.27
13. Tongue Base Retraction	TBR	0.20 ± 0.44	1.00 ± 0.55	0.01
14. Epiglottic Inversion	EI	0.20 ± 0.44	0.36 ± 0.63	0.67
Esophageal				
15. Esophageal Clearance	EsoC	N/D	N/D	N/A
Residue				
Oral	OR	0.80 ± 0.44	1.36 ± 0.63	0.08
Pharyngeal	PR	1.40 ± 0.54	1.86 ± 1.02	0.35