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Case report

Laryngeal dysfunction in Amyotrophic Lateral Sclerosis: a review and case report

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Abstract

Background: Laryngeal dysfunction can be a salient feature in the clinical symptomatology of speakers diagnosed with Amyotrophic Lateral Sclerosis (ALS). In addition to dysphonia, swallowing function is also disrupted. This paper reviews what is known about laryngeal dysfunction resulting from ALS.

Results: Presented is a case report of a female, diagnosed with ALS, whose initial symptoms were caused by laryngeal bulbar involvement that was characterized by dysphonia and dysphagia.

Conclusions: In bulbar forms of ALS, voice and/or swallowing difficulties are often the initial signs of disease. Careful examination of the muscles innervated by bulbar nerves, and tracking the rate of progressive deficit in the affected muscles, will help to solidify an accurate diagnosis. With therapy, the ability to swallow safely may still be maintained even when voice and articulation abilities are such that oral communication is inefficient.

Background

ALS, also known specifically as Motor Neuron Disease (MND) or a subtype of MND, is a progressive degenerative disease involving symptoms and signs of upper and lower motor neuron dysfunction. Carrow, Rivera, Mauldin, and Shamblin [1] described the features of four clinical varieties of MND. Progressive muscular atrophy was characterized by gradual and progressive degeneration in the lower motor neurons resulting in weakness and atrophy in the muscles of the extremities, decreased muscle tone, and fasciculations in the trunk and extremities; Primary lateral sclerosis was characterized by degeneration in upper motor neurons resulting in progressive spasticity, muscle weakness, fasciculations, and some atrophy of the skeletal muscles; Progressive bulbar palsy was characterized by degeneration of the corticobulbar fibers with bilateral atrophy of the ninth through twelfth cranial nerves, resulting in bulbar symptoms such as speech difficulties, swallowing difficulties, respiratory problems, tongue atrophy & fasciculations, and abnormal corticobulbar reflexes; Finally, ALS involved characteristics of the other three varieties of MND with atrophy of upper motor neurons and consistent speech deficits.

Weakness is the primary symptom of ALS, and this feature is usually first noted in the extremities or manifested by dysarthria (slurred speech) and dysphagia (swallowing difficulties) [2]. The average age of diagno-

sis is around 56 years and life expectancy after diagnosis is approximately three years. The usual course of ALS is unremittingly progressive, although in some cases the course of progression may be prolonged [3]. The dysarthria resulting from this disease, which usually develops into a mixed spastic-flaccid type, can be rapidly progressive and have an impact on all speech systems. Features of spasticity can include muscle weakness, hyperreflexia, extensor plantar reflexes, and pseudobulbar palsy. Flaccidity can be characterized by muscle weakness and atrophy [4]. The pattern of speech impairment resulting from the mixed dysarthria of ALS includes effortful, slow productions with short phrases, inappropriate pauses, imprecise consonants, hypernasality, strain-strangled voice, as well as decreased pitch and loudness range [5].

Kent, Kent, Duffy, and Weismer [6] described three subtypes of ALS based on the symptomatology at disease onset. Bulbar subtypes are characterized by lower motor neuron damage in the brainstem, corticobulbar subtypes are characterized by damage in the brain stem and corticobulbar tract, and corticospinal subtypes are characterized by initial symptoms in the limbs secondary to anterior horn cell and possibly corticospinal tract involvement. Bulbar and corticobulbar subtypes often manifest early difficulties in speech and/or swallowing, and bulbar subtypes seem to have a more rapidly progressive disease pattern compared to corticospinal subgroups. Bulbar subtypes also manifest a faster rate of decline of orofacial and phonatory functions [6].

In regards to the phonatory system, perceptual features can include harshness, strain-strangled voice, breathiness, tremor, and pitch abnormalities [1]. In addition, features of spasmodic dysphonia (or focal laryngeal dystonia) have also been reported in speakers with ALS [7]. In cases of bulbar involvement, dysphonia can be the initial clinical symptom, and often results in a referral to the otolaryngologist before the diagnosis of ALS has been made [8]. Typically, laryngeal structure is normal in appearance. When corticobulbar involvement dominates, there is often a pattern of hyper-adduction of the vocal mechanism, and when bulbar involvement dominates, there is often a pattern of hypoadduction

Acoustic analyses of the voice have revealed deviant fundmental frequency (Fo), amplitude and frequency perturbation (e.g., shimmer, jitter), voice range, vocal quality, and phonatory instability [9–11]. Reports of indirect or direct laryngeal observation in speakers with ALS are limited. They have included paradoxical adduction patterns, aperiodic vocal fold vibration, hyperadduction of the ventricular folds, and hypoadduction of the vocal folds [8,12,13]. Studies of phonatory performance have found large variance in both the types of deviant phonatory parameters exhibited and the severity of phonatory disruption. Predominance of spasticity or flaccidity, differential muscle involvement, and compensatory strategies has been proposed to account for this variability between speakers [9].

In cases of bulbar ALS, dysphagia can result from the involvement of the trigeminal, facial, hypoglossal, glossopharyngeal, or vagus cranial nerves [14]. Hillel and Miller [14] have described a temporal progressive pattern of affected musculature during the course of bulbar ALS. According to their observed pattern, tongue and lips were affected first (resulting in possible oral stage swallowing difficulties). The group of muscles affected secondly included the palatal, jaw, and pharyngeal muscles. Facial, upper trunk, and laryngeal muscles formed the third group of affected muscles followed by the last group consisting of extra-ocular muscles.

As a result of the order in which muscles are being affected, swallowing difficulties are one of the earliest complaints reported by patients manifesting bulbar ALS. Early problems include managing foods with tough textures, foods that fragment easily, thin liquids, and saliva control as well as increased eating time [8]. More advanced swallowing difficulties are characterized by a significantly reduced nutritional intake, severe problems controlling liquids during the swallow, and increased saliva control problems. Hillel and Miller [14] categorized the swallowing deterioration in patients with bulbar ALS into five stages:

I. Normal eating habits

II. Early eating problems

III. Dietary consistency changes

IV. Needs tube feedings

V. Nothing Per Oral

Due to early tongue and lip involvement, oral phase difficulties are usually encountered first, followed by pharyngeal stage problems due to pharyngeal and laryngeal muscle disorders. Signs of laryngeal involvement can include failure of the larynx to move superiorly and anteriorly during the swallowing reflex and incomplete closure of the larynx during elevation [8]. Pharyngeal and laryngeal complications can put the patient at risk for aspiration, and eventually result in the need for tube feedings.

The following case history illustrates how cranial nerve involvement and laryngeal dysfunction is sometimes the initial sign of ALS. Of significance in this case is the fact

that the ALS was first misdiagnosed as Multiple System Atrophy. Because MSA is a Parkinson related syndrome, the initial dysphonia was suspected to be the result of hypokineses via rigidity in the laryngeal musculature rather than a progressive lower motor neuron disease. The subsequent deterioration in swallowing abilities and severely dysarthric speech led to a neurological re-evaluation and formulation of an accurate diagnosis.

Case Presentation

JC was a 72 year-old white female referred to the speech-language pathologist from a neurologist Clinic in July of 1999 with complaints of dysphonia secondary to a neurological disease. The patient's only major medical history included spinal surgery for a lumbar disk that occurred approximately 2 years prior to the onset of the current symptoms. The neurologist initially diagnosed her with Multiple System Atrophy due to posture and balance difficulties noted during examination. She was later diagnosed with ALS. Primary involvement included the bulbar nerves. JC had a college level education and had worked as a real-estate broker for the past 30 years.

In December of 1998, JC reported experiencing a "flu-like" cold, after which she noticed an onset of speech difficulties. These speech problems had been getting progressively worse until the time of referral, and were characterized by the patient as voice quality changes and mild "blurring" of words. JC related that she had never experienced voice or speech problems prior to the onset of the recent symptoms. JC also reported mild swallowing problems. A modified barium swallow exam in March of 1999 found oral-stage delay with trace invasion into the larynx that could be cleared with a second swallow. Speech evaluation in July of 1999 indicated moderate to severe dysarthria with the respiratory and phonatory systems being more affected than articulation. The dysarthria type was characterized as flaccid, with compensatory hyperactivity in the laryngeal musculature (e.g., severe ventricular compression). Objective articulation measures yielded a sentence intelligibility score of 96%.

Voice Evaluation

Voice quality was perceptually rated as significantly hoarse, strained and pressed. In other words, the aberrant voice was noticeable to anyone. Glottal fry and mild hypernasality were also present. There was no apparent breathiness to her voice. Fundamental frequency was low for her sex and age (123 Hz). Sequential motion rates were slow and regular, characterized by difficulty of voice onset for the vowel. The loudness of her voice was soft for conversational speech. Timing of phonation was characterized by frequent complete voicing interruptions during running speech and vowel prolongation. Acoustic measures confirmed the perceptual findings as it relates to a significant increase in amplitude and frequency perturbation (see table 1). Long-term control of amplitude and frequency were also highly variable. Amplitude measures were positive for tremor. Although objective measures of noise-to-harmonic ratio revealed excessive noise in the voiced signal, this was not apparent during the perceptual analysis. The acoustic measures were most likely affected by aperiodicity in the patient's voice. In addition, variability in acoustic measures is often found in disordered voices, and one repetition of voice production can in no way be representative of any vocal measure. The acoustic measures reported in table 1 were calculated from an average of 5 repetitions.

A flexible laryngeal exam by an otolaryngologist in March of 1999 revealed a presby-type larynx with good vocal fold mobility and absence of lesions. A laryngeal exam with a 70° rigid scope completed in August of 1999 by a speech-language pathologist and ENT Physician Assistant revealed significant bowing of the true cords with moderate-severe medial compression of the ventricular folds as well as ventricular phonation during all phonatory tasks.

Post-Evaluation Therapy

Due to bowing and volume difficulties, the patient was initially enrolled in the Lee Silverman Voice Treatment (LSVT) program in August. She attended voice therapy four times a week for one-hour sessions. Treatment sessions focused on three variables: (1) maximum duration of sustained phonation at a loud phonation level, (2) maximum fundamental frequency range, and (3) maximum functional speech loudness drills. Appropriate speech breathing patterns were also addressed and incorporated into the daily exercises for each variable.

Initial attempts at LSVT tasks resulted in a further decrease in voice quality and the number of voice breaks due to ventricular compression. Moreover, JC was unhappy with her voice being produced at louder volumes. To compensate, the patient was taught voice focus with a resonant therapy approach in order to relieve some of the laryngeal tension responsible for the ventricular compression. In this behavioral approach to therapy, the patient was taught to focus the voice in the face region by shaping a nasal hum into oral productions of vowels and consonants, maintaining the sensation that was felt when the hum was present.

During successful productions, the voice focus resulted in a decrease in ventricular phonation, with very low volume and significant breathiness. The LSVT was resumed with no measurable success. Because attempts at increasing loudness resulted in recurrence of severe ven-

Table 1: Acoustic variables of J.C's voice at baseline and during the course of treatment.

Month	Fo	Jitter	Shimmer	N/H	vFo	vAm
July	123 Hz	4.06	4.14%	0.48	22.32%	28.00%
Sep.	132 Hz	6.06%	21.32%	.707	31.08%	42.99%
Oct.	141 Hz	4.47%	19.53%	.457	24.52%	41.60%
Nov.	147 Hz	4.28%	16.39%	.466	11.43%	33.33%
Dec.	161 Hz	6.50%	17.36%	.747	13.47%	40.25%

tricular phonation, this treatment strategy was discontinued after 2-weeks.

Following attempts at LSVT, the patient enrolled for one-and-a-half hour voice therapy sessions each week targeting voice focus combined with a regime of isometric vocal exercises. Glottal fry techniques were also incorporated by shaping the sound into vowels and consonants while maintaining the relaxed laryngeal posture needed for the production of the fry. Voice focus and glottal fry resulted in reduction of ventricular compression less than 50% of the time. When ventricular compression was absent, the voice was almost auditorily imperceptible. During isometric exercises (e.g., push-pull) and when instructed to speak in a louder volume, ventricular compression increased and quality of voice decreased.

Therapy continued once a week for approximately $4^1/_2$ months. Focus on voice was gradually phased out and more attention was given to the progressive dysphagia, and then articulation. Articulation treatment included a daily therapy program of oral motor motility and strengthening activities, while dysphagia treatment focused on compensatory swallowing strategies to prevent aspiration. The latter activities included the use of a supra-glottic swallow maneuver, postural changes, texture changes, oral bolus control exercises, reduction of bolus size, and education regarding thickening liquids.

Post-Evaluation Voice, Speech, and Swallowing Deterioration

Despite attempts at LSVT and traditional voice therapy, the patient's voice did not improve. Over the course of $4^1/_2$ months her voice continued to deteriorate, as did her swallowing and articulation abilities. Table 1 shows the acoustic parameters of the patient's voice at initial evaluation and during therapy.

It is apparent that Fo increased in a linear manner during the course of therapy, despite the fact that less emphasis was placed on voice treatment during the last two months. All other acoustic variables decreased during the first three months, followed by an increase in the last month of therapy. Again, acoustic variables are likely to have been influenced by aperiodicity in the acoustic signal, and the reliability of these data should be considered with that in mind. Perceptually, the voice continued to sound pressed and strained during conversational speech. Although there were changes in the acoustic measurements, these changes were not detected via perceptual measures and the voice continued to sound severely dysphonic.

In regards to swallowing, JC continued to complain of mild swallowing problems during the course of therapy. She was taught compensatory strategies such as thickening thin liquids and foods, cyclic ingestion, bolus and posture positioning, and the supraglottic swallow. In late November of 1999, JC was instructed to alter her diet to mechanical soft, after which she reported less difficulty transporting the bolus posteriorly and initiating a swallow. In January of 2000, JC had to alter her diet once more to mostly puree with some mechanical soft. A second modified barium swallow in January of 2000 revealed aspiration on thin liquids, suggesting increased impairment in the laryngeal protective mechanism during the swallow. However, thickening of liquids and using the chin tuck prevented aspiration. Delayed oral transit and reflex time as well as pharyngeal residue were also observed. Recommendations that were made included puree diet and thick honey consistency liquids using a chin tuck technique during all swallows.

Not surprisingly, J.C.'s dysarthria progressed rapidly. Objective sentence intelligibility measures via listener transcription were taken each month. The patient's dysarthria progressed rapidly, especially in the last two months of therapy. Traditional oral-motor articulation therapy was conducted weekly, and was augmented by the patient completing oral-motor exercise routines daily. Table 2 displays intelligibility, starting with baseline measures that were acquired two months prior to the in-

Table 2: Sentence intelligibility measures of J.C. before and during treatment.

Month	Intelligibility
July (Baseline)	96%
September	90%
October	83%
November	79%
December	68%
January	10%

itiation of therapy, and ending with measures taken at the conclusion of therapy.

Initial intelligibility was high considering the severe voice disturbance. Intelligibility decreased in a linear fashion during the course of treatment, and at a very rapid rate. By late January of 2000 ($1^1/_2$ months after last therapy session), JC's sentence intelligibility was 10%. The patient was using written communication at that time.

Discussion

It is important for the otolaryngologist and speech-language pathologist to be able to identify signs of neurological disease, especially in patients who have multi-focal laryngeal involvement (e.g., dysphonia and dysphagia). The patient described in the current case report was referred to the otolaryngologist by the neurologist who initially diagnosed the patient with MSA, indicating balance irregularities along with bulbar signs. Within two months the speech-language pathologist referred the patient back to the neurologist requesting an evaluation for ALS. This evaluation led subsequently to the expected diagnosis of ALS.

Generalizations regarding dysphonia, dysarthria, and dysphagia in ALS can in no way be made from analysis of one patient. The current case report is only one example of how rapidly progressive forms of ALS are difficult to manage behaviorally. Voice and articulation therapy was ineffective for this patient. Lack of improvement after behavioral therapy has been reported previously in the literature [17]. The speed of progressive deterioration often results in the inability to judge the efficacy of behavioral treatments for patients with ALS [18]. There are limited resources available in the literature that document treatment efficacy in ALS with either fast or slow progressive patterns.

Management of dysphagia did have a greater benefit. While the patient needed to alter the texture of her diet for bolus control purposes, compensatory swallowing strategies and changes in eating habits (e.g., smaller portions, increased mastication time) resulted in her maintaining per oral status and adequate nutritional intake through January of 2000. At that time her speech intelligibility was only 10%. It has been reported that the progressive pattern of dysphagia will follow that of speech in patients with ALS [18]. In J.C.'s case, the two patterns of progression did not follow similar courses. Swallowing abilities continued to be more effective than speech abilities throughout the process of motor deterioration.

Competing Interests

None declared.

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