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INSIGHTS INTO SOME ATYPICAL FINDINGS OF A CASE WITH AMYOTROPHIC LATERAL SCLEROSIS: ZOOMING FROM SPEECH LANGUAGE PATHOLOGIST'S PERSPECTIVE

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Abstract

The purpose of this article is to present a single case study on amylotrophic lateral sclerosis. The case will illustrate how speech language pathologists assessed the speech, language, cognition and swallowing profiles in bedside situation. Evidence gathered from the case supports the conventional contention of presence of flaccid spastic form of dysarthria and dysphagia. However, something which makes this case fascinating and rare is the presence of clonus of jaw along with cognitive decline and suspected primary progressive aphasia, which lead to severe communication deficit. More globally, this case is an upgradation of understanding of communication and swallowing profile of Amyotrophic lateral sclerosis among multidisciplinary professionals working across neuro-rehabilitation set ups.

Key Note: This case report focuses on the assessment findings with additional unique features from speech language pathologist's standpoint. It basically highlights the presence of hyperkinetic component and presence of progressive aphasia apart from the known mixed component of dysarthria.

Keywords : Amotrophic Lateral Sclerosis, Dysarthria, Cognition, Primary Progressive Aphasia, Dysphagia.

Introduction: Amyotrophic lateral sclerosis (ALS) is a degenerative neurological disease of unknown etiology in which motor neurons of brain and spinal cord are involved. Speech, language & swallowing problems are frequently noted during the course of ALS. Saunders, Walsh, & Smith [1] estimated that about three fourths of persons with ALS are nonspeaking by the time of their death. There is a dearth of literature with respect to speech, language and cognitive functions among ALS patients. In the present paper, we will report a case of ALS with some unique features as observed by speech language pathologist.

Case Report: We are reporting a forty-five-year-old male case from the neurology ward of Sri Aurobindo Institute of Medical Sciences, Indore, Madhya Pradesh with a complaint of deterioration in speaking, swallowing and locomotion. History reported that the onset of speech and language difficulty was 1 year 6 months back while of swallowing difficulty was 3 months back. He presented with symptoms like atrophy, drooling, shortness of breath and was dependent on caregiver for activities of daily living. The symptoms were gradually progressive and he is currently on nasogastric tube. Magnetic Resonance Imaging study (done using 18 channel 1.5 Tesla system) revealed bilateral symmetrical T2 hyperintense signal involving the cortico-spinal tract in the region of posterior limb of internal capsules, cerebral peduncles and pons. Mild-moderate dilatation of the ventricles was seen with predominant enlargement of bilateral occipital horns. Mild generalized cerebral and cerebellar atrophy was also seen. Electromyography results depicted presence of sharp waves, fibrillation potentials and reduced neurogenic firing pattern with increased motor unit action potentials amplitude and duration. On the basis of the findings, a confirmed diagnosis of Motor neuron disorder specific to ALS was done by the Neurologist.

Detailed bedside assessment was carried by Speech Language pathologist focusing the domains such as cognition, speech, language and swallowing. Written consent was taken from patient's caregiver. Tools used for assessments included Glassgow Coma Scale (GCS)to assess alertness, Montreal Cognitive Assessment- Hindi version (MOCA-H) and Clinical Dementia Rating (CDR) for cognitive evaluation, Hindi Aphasia Screening Indore Test (HASIT)for language evaluation, Frenchay Dysarthria Assessment (FDA), Intelligibility rating scale for motor speech disorder and Dysarthria rating of severity for speech evaluation, and Gugging Swallowing Screen (GUSS) for swallowing assessment.

At the time of evaluation GCS revealed that the individual was alert and aware with the surroundings. Oro- motor examination revealed limited functions, range, speed, strength and accuracy of all the active articulators with presence of brisk jaw jerk reflex, hypoactive gag reflex, atrophy, fasciculations, pseudobulbar affect and dysphagia. There was an additional atypical presence of clonus movement in jaw suggestive of presence of hyperkinetic component. Speech characteristics were reduced loudness, hypernasality, monopitch and strained-strangled quality with minimal verbal output. Speech was markedly unintelligibility since it was not a viable means of communication in any environment, regardless of restrictions in content or attempts at repair and requires augmentative and assistive communication devices. The profile of speech problems has been shown in Figure 1. The person with ALS though passed in indirect swallow test (with postural adjustments) but failed in direct swallow test. He could swallow food of semisolid texture (pudding thick consistencies), however showed involuntary cough and gurgly voice across other consistencies.

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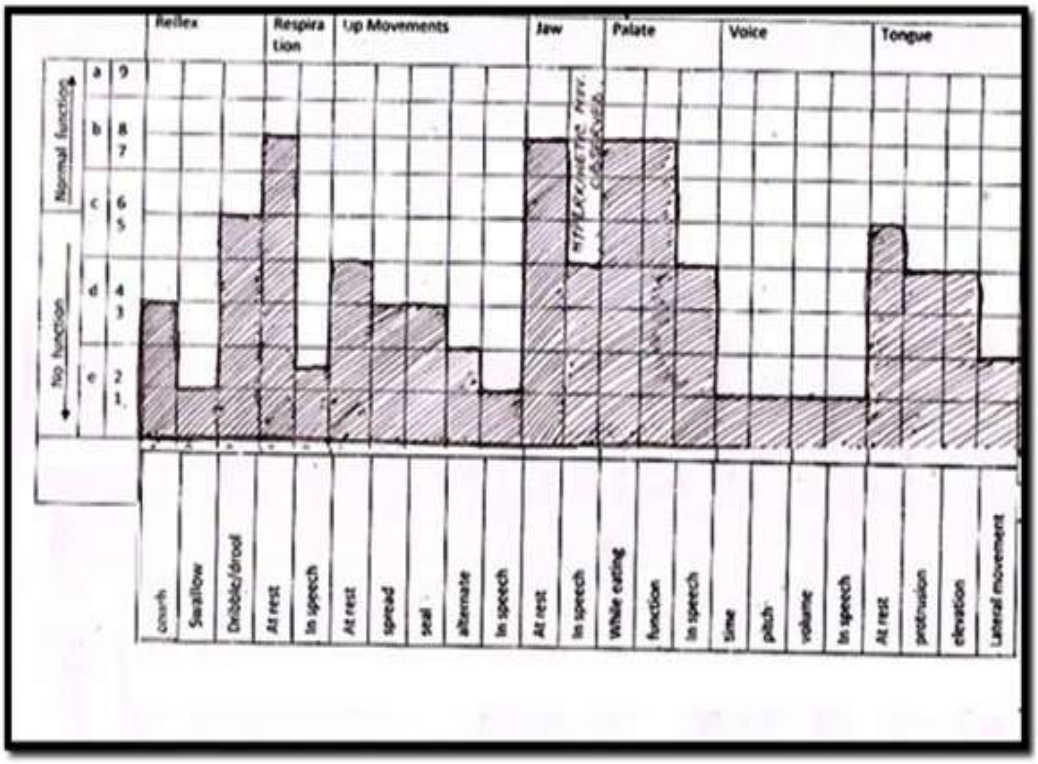

Figure 1: Dysarthria Profile of the patient with ALS.

Assessment of linguistic domains was carried out. No verbal responses could be elicited across repetition, naming and verbal fluency domains. Auditory comprehension was comparatively better preserved as demonstrated by comprehension of simple commands through verbal and written modalities. Reading and writing domains were also affected. The screening through HASIT indicated towards severe non-fluent aphasia. Cognitive screening done through MOCA-H suggests that the patient is lacking in attention, executive function, visuo-construction skills, abstraction, delayed recall and language areas. CDR suggests presence of moderate dementia. The scores of the tests are represented in Figure 2. The samples of patient's cube copying, clock drawing and writing skill, is shown in Figure 3.

Sr. no.	Test administered	Scores obtained	Maximum score	Interpretation
a.	Glassgow Coma Scale	15	15	Patient is conscious and alert.
b.	Montreal Cognitive Assessment- Hindi	04	30	Neuro-cognitive deficit
c.	HASIT	07	30	Linguistic deficit associated with non-fluent subtype.
d.	Speech Intelligibility	01	10	Markedly affected
e.	Dysarthria severity	05	05	Severe dysarthria
f.	Gugging Swallowing Screen	07	20	Severe oro pharyngeal dysphagia with high risk of aspiration

Figure 2: Results of formal tests administered in the patient.

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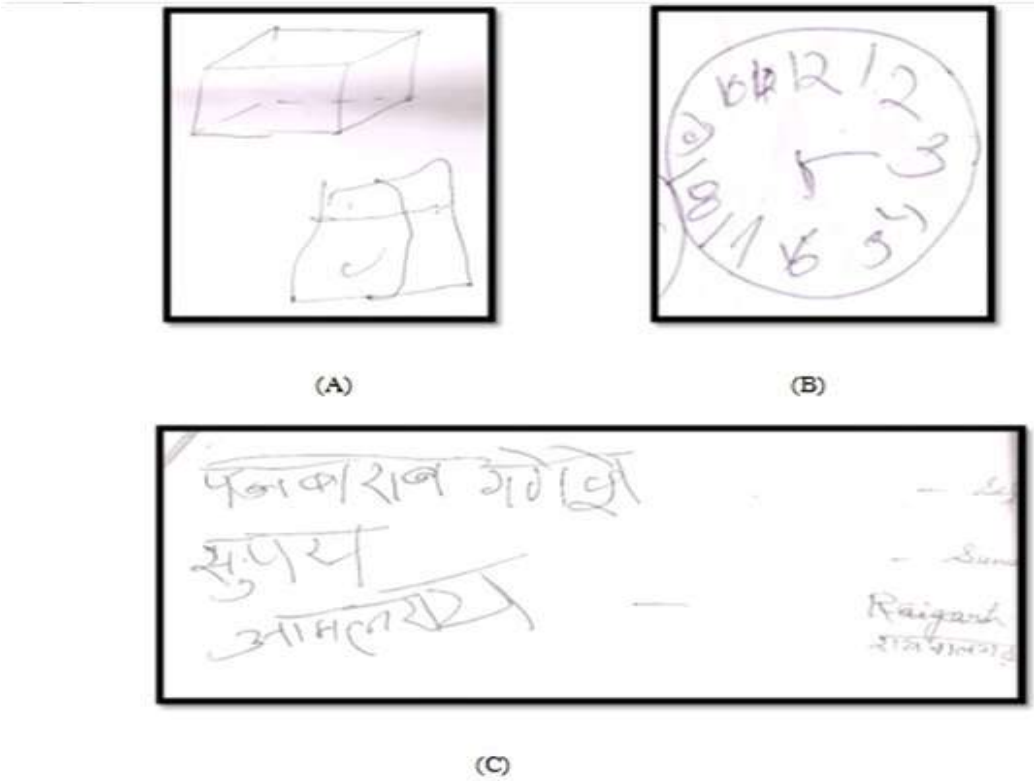

Figure 3: Samples of patient's (A) cube copying, (B) clock drawing and (C) writing skill.

Presence of mixed (flaccid-spastic) dysarthria is known among ALS, but we found a unique feature of presence of jaw clonus, indicative of hyperkinetic component along with flaccid-spastic component. This is rare yet fascinating. Beevor, et al. [2] described a case of ALS with clonus of the lower jaw way back in 1886. Saluja et al [3] reported a 57-year-old male case of ALS with jaw clonus. Sharma, B., et al [4] reported a case with jaw clonus in a patient who presented with vascular dementia and Parkinsonism.

In many studies [5,6] the presence of cognitive dysfunction in ALS have been cited, but the presence of Primary progressive aphasia (PPA) is often underacknowledged in research. A recent study carried out by Tan et al [7] reported that there is a presence of PPA in one third Fronto-Temporal Dementia-ALS patients. In the present case report, we correlated the language impairment (non-fluent type) with the history of deterioration of language which lead us to diagnose the case as probable primary progressive aphasia.

Accumulating the evidences, we finally diagnosed the case as Progressive Aphasic Dementia & Mixed Dysarthria (Flaccid>Spastic-hyperkinetic subtype) attributing to mutism along with severe oro pharyngeal dysphagia with high risk of aspiration, secondary to bulbar onset ALS. Rehabilitation and facilitation of alternative and augmentative communication was something that has to be started to improve his quality of life.

Conclusion: Most persons with ALS face a breakdown in communication during the course of the disease, which makes it challenging for SLP's to evaluate communication skills. A descriptive assessment considering all the possibilities should be done, so that SLPs could meet the communicative needs of persons with ALS. This patient is seemingly a unique lesson and an up gradation of understanding for professionals working in multidisciplinary neuro rehabilitation set up in India.

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