

A Case of Dysphagia due to Motor Neuron Disease: An Uncommon Cause

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Abstract

Dysphagia is a commonly encountered symptom in ENT clinical practice. Dysphagia due to the neurological cause is known. However, dysphagia due to amyotrophic lateral sclerosis (ALS) is relatively uncommon in ENT practice. ALS is a neurodegenerative disease characterized by progressive muscular paralysis. In motor neuron disease there is a sequence of progressive dysphagia and dysarthria, which may be associated with gradual spasticity in the weakened and atrophic limbs, affecting the gait and manual dexterity. The management of ALS is multidisciplinary, supportive and palliative. Non-invasive ventilation prolongs survival and improves quality of life. We report a case of a 57-year-old male who presented with progressive dysphagia and dysarthria associated with cramps in legs since 2 years. Keeping the possibility of neurological dysfunction as the cause we treated this case with baclofen satisfactorily.

Keywords: Amyotrophic lateral sclerosis, Baclofen, Deglutition disorder, Dysarthria, Motor neuron disease

INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is the most common adult-onset neurodegenerative disorder of the motor neuron system. Males are affected more than females, with an M:F ratio about 1.5:1, although recent data suggests that the gender ratio may be approaching equality.¹⁻⁴ The mean age of onset for sporadic ALS (SALS) varies between 55 and 65 years with a median age of onset of 64 years.^{5,6} Only 5% of cases have an onset before the age of 30 years,⁶ although juvenile sporadic onset cases are increasing.⁷

It is characterized by affection of upper motor neuron (UMN) and lower motor neuron (LMN) in the primary motor cortex, brainstem and spinal cord.⁸ Approximately, two-thirds of patients with typical ALS have a spinal form of the disease (limb onset) and present with symptoms related to focal muscle weakness and wasting, where the symptoms may start either distally or proximally in the

upper and lower limbs. Gradually, spasticity may develop in the weakened atrophic limbs, affecting manual dexterity and gait. Patients with bulbar onset ALS usually present with dysarthria and dysphagia for solid or liquids with simultaneous limbs symptoms. Paralysis is progressive and leads to death due to respiratory failure in ALS cases. The diagnosis is based on history, clinical examination, electromyography, and exclusion of "ALS-mimics" (e.g. cervical spondylotic myelopathies, multifocal motor neuropathy) by appropriate investigations. Signs of UMN and LMN damage not explained by any other disease process are suggestive of ALS. The management of ALS is supportive, palliative, and multidisciplinary. Non-invasive ventilation may prolong survival and improve quality-of-life.

CASE REPORT

A 57-year-old Indian male presented with progressive dysphagia, dysarthria and cramps in legs since 2 years, nasal twang since 1½ years and hoarseness of voice, difficulty while chewing food since 2 months. Patient was not a known case of diabetes and hypertension. Clinically patient had spastic slurred speech, intermittent nasal twang and visible fasciculation over tongue. Indirect laryngoscopy examination revealed left vocal cord palsy. Barium swallow study was normal and did not show any obstructive or mass

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lesion. A provisional impression of neurological dysphagia was considered.

On further neurological examination, tongue was spastic and atrophic. There was gross muscle wasting around both shoulder girdles. Fasciculation was present around both shoulder girdles, in biceps, triceps, trapezes, and spinal muscles. Deep tendon reflexes of the upper limb were normal bilaterally whereas the lower limb had brisk reflexes on either side. Plantar reflexes were found to be equivocal. A clinical impression was revised based on above neurological findings as motor neuron disease (MND)–Bulbar type ALS.

On further investigations, electromyography (EMG)-nerve conduction velocity studies showed long duration polyphasic in all tested muscles of limbs and tongue which was suggestive of widespread MND. Possibility of space-occupying lesions or any other pathology was ruled out by computed tomography and magnetic resonance imaging (CT/MRI) imaging.

In view of both routine clinical as well as neurological findings and its slow progressive nature, the final impression was made as bulbar MND-ALS.

In this case it was a disease that started as bulbar involvement with hoarseness and dysphagia progressing to limbs. Medical treatment was started with treatment baclofen 10 mg in divided daily dose initially for 1 week and followed by 20 mg in divided daily dose for 3 months. Physiotherapy and speech therapy was advised along with dietary supplementation. Patient was followed up for 3 months. At the end of 3 months post treatment, patient had an overall improvement like enhancement of oral food intake along with a decrease in spasticity of limbs leading to improved daily activities as well as quality of voice.

DISCUSSION

Among the causes of neurological dysphagia like cerebrovascular accidents, accidental or surgical trauma, multiple sclerosis, space-occupying lesions such as tumors, the MND is relatively rare cause.

MND is of various types viz. classic ALS (combined UMN and LMN involvement), progressive bulbar palsy (predominant bulbar involvement), progressive muscular atrophy (predominant LMN involvement), primary lateral sclerosis (predominant UMN involvement).

ALS or Charcot disease or Lou Gehrig disease is the most common MNDs. It is a progressive disease of the cortico-bulbar and cortico-spinal tracts. In this form of

the disease, progressive dysphagia affecting the oral and oropharyngeal stage together with dysarthria and anarthria account for the misery.

The incidence of ALS is reported to be between 1.5 and 2.7 per 100,000 population/year.² However, the prevalence ranges from 2.7 to 7.4 per 100,000 population/year.¹ The mean age of onset for SALS varies between 55 and 65 years with a median age of onset of 64 years.^{5,6} Males are affected more than females with an M:F ratio about 1.5:1.^{1,4}

There is no consistent association between a single environmental factor and risk of developing ALS. It was found that only smoking is likely to be associated with ALS, while other risk factors were weakly related.^{9,10}

“Amyotrophy” refers to the atrophy of muscle fibers, which are denervated as their corresponding anterior horn cells degenerate, leading to weakness of affected muscles and visible fasciculation. “Lateral sclerosis” refers to hardening of the anterior and lateral cortico-spinal tracts as motor neurons in these areas degenerate and is replaced by gliosis. Symptoms of ALS include limb muscle weakness, cramps, occasionally fasciculation, disturbances of speech, swallowing, dysarthria, pathological laughter or crying.

UMN dysfunction leads to stiffness, brisk or abnormally spreading tendon reflexes, presence of abnormal reflexes (hyper reflexic jaw jerk, Babinski sign), and loss of dexterity in the presence of normal strength.

LMN dysfunction manifests as muscle twitching (fasciculation), reduction of muscle bulk (atrophy), foot drop, depressed reflexes, breathing difficulties.

Diagnosis

Diagnosis is mainly clinical with combined features of UMN and LMN dysfunction such as weakness, atrophy, fasciculation of muscles occurring in combination with increased tone and hyperreflexia.

In ALS EMG study confirms the diagnosis and helps to exclude other peripheral causes. EMG study shows fibrillation and fasciculation potentials of high amplitude and long duration polyphasic motor units. Nerve conduction study is normal in sensory and abnormal in motor with reduced motor compound muscle action potentials. CT/MRI is used to rule out structural lesions of muscle. Nerve biopsy must be considered if the presentation is atypical, biochemical markers, and genetic studies are also considered.

Complications of ALS include progressive inability to perform activities of daily living, including handling utensils for self-feeding, deterioration of ambulation, aspiration pneumonia, and respiratory insufficiency.

Wheelchair-bound or bedridden patients are likely to have decubitus ulcers and skin infections, deep vein thrombosis and pulmonary emboli.

Treatment

Agents in routine clinical practice include skeletal muscle relaxants viz. baclofen, tizanidine (for UMN), *N*-Methyl-D-aspartate receptor antagonist as dextromethorphan and quinidine (for emotional liability due to the pseudobulbar effect). Riluzole is another drug that has been shown to have a modest effect on prolonging life in ALS patients. Riluzole at 100 mg probably prolongs median survival by 2-3 months when taken for 18-month duration. The drug is generally well tolerated with the most common side effects being asthenia, nausea, gastrointestinal upset and abnormal liver function tests, and therefore liver function should be regularly monitored during therapy.¹¹

Dysphagia is a common symptom of ALS and leads to increased risk of aspiration, malnutrition, weight loss and dehydration. Most guidelines state that supplementary enteral feeding should be considered when body weight falls by >10% of the pre-diagnostic or baseline weight.^{12,13}

The three options available for enteric feeding include percutaneous endoscopic gastrostomy, percutaneous radiologic gastrostomy or radiologically inserted gastrostomy, and nasogastric tube feeding. There is no cure for progressive dysarthria in ALS.

Some symptomatic and compensatory strategies may temporarily improve the patient's communication and have an impact on quality of life. The patient may move from oral communication to written communication, to using an augmentative communication device, or via another person.¹⁴

Restriction of physical activity is not always necessary. Indeed, early in the course of ALS patients are encouraged to continue routine activities. However, patients should not overexert themselves to the point of fatigue or pain. Patients should maintain a regular exercise regimen within individual limits. Patients with slowly progressive disease will be able to tolerate exercise and benefit from it more than patients with rapidly progressive disease.

Prognosis

ALS is a fatal disease. Overall median survival from onset of symptoms for ALS ranges between 2 and 3 years for bulbar onset cases and 3-5 years for limb onset ALS cases.

CONCLUSION

In patients of dysphagia focusing only the local signs alone, may miss the diagnosis of a systemic cause. Therefore, a complete neurological examination is essential in patients having dysphagia associated with neurological disturbances for precise diagnosis and management of neurodegenerative disorder.

Although ALS is incurable, there are treatments that can prolong meaningful quality of life; therefore, diagnosis and its treatment are important to both patient as well as family.

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