

Conventional Remedy to Lou Gehrig's Disease - Amyotrophic Lateral Sclerosis (ALS): A Rare Clinical Entity

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Abstract

Lou Gehrig's disease, commonly known as amyotrophic lateral sclerosis, is an incredibly rare clinical condition that affects about 5 in 100,000 people annually in India, making it one of the rarest diseases in the entire world. With an average age of 55, this disease typically impacts people between the ages of 40 and 70. Males are more prone to develop the condition. Amyotrophic Lateral Sclerosis (ALS) is a neurodegenerative disorder affecting primarily the motor system, but in which extramotor manifestations are increasingly recognized. The loss of upper and lower motor neurons in the motor cortex, the brain stem nuclei and the anterior horn of the spinal cord gives rise to progressive muscle weakness and wasting. Typically, patients report difficulties swallowing or a shift in voice pitch while speaking due to tongue involvement. Initial ALS symptoms can resemble those of other neuromuscular illnesses and are completely non-specific. In this case report, we described a 44-year-old gentleman who has been diagnosed with Amyotrophic Lateral Sclerosis in accordance to an MRI scan describing symptoms like unable to speak, reduced tongue movements, trouble swallowing, excessive salivation, swelling over the tongue and loss of tongue tone for the past three to four months. The present study discusses the clinical presentation, diagnosis, therapeutic regimen and prognosis. Both allopathic and *ayurvedic* oral medications with local therapeutic regimens like *Jivha Pratisarana* (Rubbing of dry powder on tongue) and *Nasya* (nasal drops of medicated oil) were employed in the patient which have satisfactory results enhancing for betterment of lifestyle.

Keywords: Amyotrophic Lateral Sclerosis, Jivha Pratisarana, Lou Gehrig's Disease, Nasya

1. Introduction

Amyotrophic lateral sclerosis, also referred to as Lou Gehrig's disease, first emerged in a famous baseball player named Lou Gehrig on June 2, 1941¹. The Greek root of the word "amyotrophic" is "a" for no, "myo" for muscle, and "trophic" for sustenance. In other words, this condition causes the muscle to starve, which causes it to atrophy. In ALS, "Lateral" refers to the spinal cord, which contains the nerves that govern the muscles, and "Sclerosis" refers to the scarring or degeneration of that region. As a result, the disease causes scarring or degeneration of the motor neurons that supply muscles, which results in gradual muscle weakness². Therefore, misinterpretation becomes especially common in the early stages of the disease. People with ALS have a life expectancy ranging from 3 to 10 years or more. Upper and lower motor neurons that can be discovered in the spinal cord, cranial nerve nuclei, and motor cortex are lost due to the neurodegenerative illness known as Motor Neuron Disease (MND). ALS, or amyotrophic lateral sclerosis, is one of the most widespread MND types. Muscle rigidity and twitching are early clinical signs. These symptoms keep getting worse with time, weakening the muscles and creating muscular wastage. Later-developing symptoms include respiratory failure, difficulty breathing, speaking, and swallowing, among others. Whenever ALS first manifests, the symptoms typically start with weakness in the arms or legs or in the case of bulbar onset ALS, with difficulties speaking or swallowing³. Non-invasive ventilation is the first line of treatment for respiratory failure, with invasive ventilation being used as a last resort for those with advanced ALS. It is advised to have physiotherapy, occupational therapy, and speech therapy. People with ALS should not lose weight or become malnourished since doing so increases survival and quality of life⁴. In this case, *Ayurveda* has tremendous results for restricting the progression of the disease and to cure symptoms. Since there is currently no treatment for ALS in modern science though management of the condition focuses on symptom control and supportive care.

As just a few examples of this uncommon clinical entity have been documented in the literature, the goal of this study is to analyse the presentation and prognosis of it with a holistic treatment module.

2. Case Report

A 44-year-old male patient visited the OPD complaining of being unable to speak, reduced tongue movements, trouble swallowing, excessive salivation, swelling over the tongue, and loss of tongue tone for the past three to four months. The patient was completely symptom-free and normal before this.

2.1 History of Present Illness

The patient was healthy four months ago as per his opinion and then he felt above mentioned symptoms that progressed in 3-4 days. Later he visited the nearby physician and took treatment but was unable to speak and was having trouble swallowing so visited *Shalakyatantra* OPD of Dr. D. Y. Patil College of Ayurved and RC, Pimpri, Pune.

- Family History: Not significant.
- **Past History**: No history of diabetes, high blood pressure or any addiction.
- General Examination: General condition of the patient was fair; appetite of the patient is normal; Bowel- irregular and unsatisfactory; micturition-normal, sleep- sound, pallor and icterus- absent, cervical lymph nodes- not palpable. Blood pressure-124/78 mm of Hg, Pulse- Regular 84/min.

2.2 Systemic Examination

CVS - S1S2 heard

RS - Air entry bilaterally equal; no additional respiratory sound heard.

CNS - On examination, the patient was fully oriented and responding to all the commands, but lack of speech was noted. Drolling present, Sensory reflexes were normal.

2.2.1 Examination of Rogi (patient) according to Ayurveda

- 1. Prakriti: Pitta-kapha
- 2. Vaya: Madhyama
- 3. Bala: Madhyama
- 4. Agni: Madhyama
- 5. Koshtha: Krura
- 6. Abhyavarana Shakti: Madhyama
- 7. Jarana Shakti: Avara
- 8. Nadi: Pittaj

2.2.2 Samprapti Ghataka

- Dosha- Vata, Kapha
- Dushya- Rasa, Mansa, Majja
- Srotas- Rasavha, Majjavaha
- Adhishthana- Utmang
- Samutthana- Amashaya smuttha
- Vyaktisthana- Jivha
- Rogamarga- Madhyama
- Agni- Manda

3. Local Examination of Tongue

The swelling was present on the tongue. The folds of the tongue mimicked a swelling. The patient was only



Figure 1. Patient presented to us with complaints of unable to speak with decreased tongue movements.

able to move his tongue antero-posteriorly. There were no upward or downward movements of the tongue. Indirect Laryngoscopy showed normal Vocal Cords and their movement (Figure 1).

4. Assessment Criteria

Table 1.	Assessment	criteria	(Before	treatment)
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Sr. No.	Parameter	Score (Before treatment)
1.	Speech	Grade 3 (Speech is often hard to understand)
2.	Salivation	Grade 2 (Moderate drooling, drool reaches the lips and the chin)
3.	Swallowing	Grade 3 (Tolerates fluid only)
4.	Swelling (Tongue)	Grade 1 (Swelling present)

Table 2. Speech parameter gradation

Sr. No.	Speech Parameter	Gradation
1.	Speech is always easy to understand	0
2.	Speech is occasionally hard to understand	1
3.	Speech is often hard to understand	2
4.	Speech is hard to understand most or all of the time	3

Table 3. Salivation parameter gradation¹⁶

Sr. No.	Salivation Parameter	Gradation
1.	Dry (Never drools)	0
2.	Mild drooling, only lips wet	1
3.	Moderate drooling, drool reaches the lips and the chin	2
4.	4. Severe drooling, drool drips off the chin and onto clothes	

 Table 4.
 Swallowing parameter gradation¹⁷

Sr. No.	Swallowing parameter	Gradation
1.	No change over baseline	0
2.	Mild difficulty while swallowing solids	1
3.	Significant difficulty while swallowing solids, often requires soft diet	2
4.	Tolerates fluids only	3
5.	Requires nasogastric tube feeding	4

Table 5. Swelling parameter gradation

Sr. No.	Swelling Parameter	Gradation
1.	Swelling absent	0
2.	Swelling present	1

5. Investigations

5.1 MRI Tongue (Plain and Contrast)

An MRI of the tongue showed an Ill-defined signal intensity abnormality and swelling involving the posterior third/base of the tongue and both oropharyngeal walls showing abnormal enhancement along with lymphoid hyperplasia of the base of the tongue and fatty infiltration and atrophy of the tongue (Figures 2-4).



Figure 2. Film of MRI of tongue.



Figure 3. MRI tongue report.



Figure 4. MRI tongue report.



Figure 5. Video laryngoscopy image showing normal vocal cords with pooling of saliva.

5.2 Video Laryngoscopy (VLS)

Video Laryngoscopy (VLS) was done for further assessment to visualise the ill-defined mass at the base of the tongue and get its biopsy for further management. Video laryngoscopy did not reveal any lesion at the base of the tongue (Figures 5 and 6).

Finally, an opinion of a Neurophysician was taken based on the MRI of the tongue and his symptoms. He was diagnosed with ALS.

• Diagnosis: Lou Gehrig's Disease-Amyotrophic Lateral Sclerosis (ALS).



Figure 6. Video laryngoscopy image showing lingual part of tongue.

6. Therapeutic Regimen

Internal as well as local therapies were given to the patient as follows:

Table 6 demonstrates the treatment plan that was into consideration, which included intravenous, oral and local treatment.

This was carried out for a total of 30 days, and final assessment was done for evaluation of observation and to draw the conclusion.

Sr. No.	Therapeutic regimen	Route of drug administration	Dosage	Duration
1.	Inj. Edaravone in 100ml NS	Intravenous	Alternate day	30 days
2.	Tab. Rilutor 50 mg	Orally	1 Tab. twice a day (After food)	15 days
3.	Tab. <i>Agnitundi vati</i> (125mg)	Orally	2 Tab. thrice a day (Before food)	Initial 7 days
4.	Tab. Ekangaveer ras (125mg)	Orally	2 Tab. thrice a day (After food)	28 days
5.	Tab. <i>Gandharvaharitaki vati</i> (500mg)	Orally	2 Tab. HS (After food) with warm water	28 days
7.	Liq. Dhanadanyadi Kashayam	Orally	20ml twice a day (Before food) with ½ glass of water	28 days
8.	Ashtanga Ghruta	Orally	20ml in morning with warm water	14 days
9.	Jivha Pratisarana with Vaak-Shudhikara Choorna	Locally	Once a day in morning	28 days
10.	Pratimarsha Nasya with Panchendriyavardhan Taila	Locally	Once a day, 2 drops in each nostrils	28 days
11.	Speech Therapy	-	Daily	28 days

 Table 6.
 Therapeutic regimen with internal and external therapies

7. Chronology in Observation

Sr.No.	Parameter	Score (Before treatment-0 th day)	Score (After treatment-30 th day)
1.	Speech	Grade 3 (Speech is hard to understand most of time)	Grade 1 (Speech is hard to understand occasionally)
2.	Salivation	Grade 2 (Moderate drooling, drool reaches the lips and the chin)	Grade 1 (Mild drooling, only lips wet)
3.	Swallowing	Grade 3 (Tolerate fluids only)	Grade 2 (Mild difficulty while swallowing solids)
4.	Swelling (Tongue)	Grade 1 (Swelling present)	Grade 0 (Swelling absent)

 Table 7. Observation according to assessment criteria of before and after treatment

8. Graphical Representation of Observation



Figure 7. Graphical representation of observation (before and after treatment).

9. Discussion

In the majority of cases, the cause of Amyotrophic Lateral Sclerosis (ALS) is still unknown. Because the initial signs and symptoms of ALS are so vague, they can resemble those of other neuromuscular illnesses. This makes disease misdiagnosis more common. The diagnosis of ALS is often done clinically and needs proof of a gradual spread of symptoms, which can take some time to establish due to the absence of reliable diagnostic biomarkers. Patients with a bulbar onset were reportedly diagnosed more quickly than those with a Limb onset. The spinal onset of ALS affects between 58% and 82% of patients⁵. Across all nations,

this is constant. Speaking, chewing, and walking are all controlled by the afflicted muscles. Until the capacity to move, speak, eat, and finally breathe is lost, motor neuron loss continues to advance gradually. In the end, ALS results in paralysis and perhaps death, typically from respiratory failure⁵.

9.1 Role of Riluzole and Edaravone in Treating ALS⁶

Two approved medications for the treatment of ALS are riluzole and edaravone. These medications have no effect on the disease's progression or cure. They just contribute to slowing the rate of mortality and morbidity. Oral riluzole has been demonstrated to extend life by three to six months. It has been demonstrated that edaravone, administered via intravenous infusion, slows the deterioration in everyday functioning.

9.2 Role of Ayurvedic Treatment in ALS

ALS can be understood as *anuktavata vyadhi* based on similarity of symptoms as there is no direct correlation in *Ayurvedic* classics. With immunosuppressant or cortico-steroids as only available treatment in modern sciences, *Ayurveda* can be ray of hope. The scope of *Ayurvedic* management and preventive aspects mainly concentrates on improving the quality of life and decrease dependency by patients on others. Hence, here an attempt is made to analyse the disease and its management.

The main *Dosha* contributing to ALS is *Vata*. The biological energy known as *vata* acknowledges and promotes all actions in the body⁷. *Acharya Charaka* explains that the senses and the channels carrying the sensory and motor impulses from the *Shiras* are like the rays from the sun. When the *Gati* of *Vata* is obstructed then the functions of *Vata* impaired and that is liable for causing various *Vatavyadhi Shiras* (Head) is the centre of *Indriyas* (special senses)^{8,9}. Language is more of the function of *Udana Vayu*, if it gets vitiated then loss of speech, and drolling symptoms are noted. *Vatahara*, restoring *udana vayu* pathway and *Bhrihmana Chikitsa* are the chosen treatments in this scenario because *Vata Dosha* is the primary cause of the disease¹⁰.

9.3 Mode of Action

9.3.1 Tab. Agnitundi vati

Amazingly, *Agnimandya*, which was brought on by *Kaphavruddhi*, is handled by *Kalpa*. It performs like a miracle under circumstances like *Vishtabdhajirna*. This *kalpa* eliminates *Shaithilya* from *Amashaya*, activates *Amashaya*, and corrects the *Gati* of *Saman Vayu*. When indigestion-related food is packed into the intestines as undigested faeces, the food obtains the appropriate direction and *vata*-related symptoms like *adhmana*, *aanaha*, and *shoola* go away. The primary effects of this *kalpa* are to reduce *Amavisha vatapradan lakshanas* and to correct *Agnimamdya*, *Ajirna*, and *Visuchika*. *Agnitundi Rasa* strengthens nerves by acting on the nerves that emerge from *Shira*, and *Prushthavansh Rajju* aids in maintaining control of the face, hands, and legs¹¹.

9.3.2 Tab. Ekangaveer ras

Due to its reputation as the best *vata nashaka* preparation, *ekangaveer ras* has an impact at higher levels. Relief from the effects of *Rasayana* and *vatanashaka ghataka dravyas* of *Ekangaveer ras*, such as *gandhak*, *Rasasindur*, *Kantaloha bhasma*, *Tikshana lohabhasma*, and *Abhrak bhasma*, on symptoms like speech difficulty, facial paralysis and sensory alterations¹².

9.3.3 Tab. Gandharvaharitaki vati

Gandharva Haritaki is a traditional polyherbal *ayurvedic* remedy. The condition is prevalent in *Kati Pradesh*, the *Vata dosha's* primary location. *Mridu virechana* is a line of treatment for *Vatavyadhi*, and this remedy has *Vatahara*, *Vrishya*, and *Snigdha Virechaka* qualities also helps to restore *apana vayu*. It is recommended for the treatment of pain in *Gridhrasi*, *Ardita*, *Sandhivata*, and *Vatavyadhi*¹³.

9.3.4 Liq. Dhanadanyadi Kashayam

A herbal decoction called *Dhanadanayanadi Kashayam* is utilised to restore lost muscle and nerve function as well as to soothe exacerbated *Vata Dosha*¹⁴.

9.3.5 Ashtanga ghruta

Loss of tongue tone and decreased movements were reduced significantly, which may be due to *Shamana* of *Vata Guna* like *Chala* and *Shita* by virtue of *Ashtanga Ghrita*. The majority of *Ashtanga Ghrita's* constituents have *Madhura Vipaka*, *Medhya*, *Rasayana*, and *Vatahara* characteristics. It will assist the management by doing the *Dhatuposhana*. As a preventative and therapeutic agent, *Ashtanga Ghrita* is thus a neurotropic substance¹⁵.

9.3.6 Jivha Pratisarana with Vaak-Shudhikara Choorna

By virtue of Ushna Veerya and Katu Vipaka, the Vakshuddhikara Churna, which uses Vacha as its key ingredient and is also a Kapha Vata hara, Swarya, and Medhya, is helpful in removing Kapha's obstruction of Vata. The medications' medhya property aids in treating the psychological condition. Other substances such as Kushta, Ajamoda, and Jeerka, which contain Ushna Veerya and Katu Vipaka, also function as Vata Kapha Shamaka, which is advantageous in Samprapti *Vighatana*. In addition to acting as *Rasayana* and *Balya*, *Pippali*, *Haritaki*, and *Shunthi* possess *Snigdha*, *Teekshna Ushna*, and *Madhura Vipaka* properties. This makes them advantageous against *Vata* and *Dhatukshaya*. Having *Deepana* and *Pachana* qualities, *Jeeraka*, *Shunthi*, and *Ajamoda* will act as on *Agni* and aid in the appropriate assimilation and pharmacological activities of the medications¹⁶.

9.3.7 Pratimarsha Nasya with Panchendriyavardhan Taila

Marsha Nasya therapy delivered was with Panchendriyavardhana Taila, which is administered through the nasal root and continues up to Sringataka Marma (a crucial point where blood vessels and nerve endings converge in the head region). From Shringataka Marma, the medication travels to various Srotas (Channels) and lessens the vitiation of doshas. The motor functions of the nerves are improved by nasya, which nourishes the nervous system through diffusion as well as by neuronal and vascular pathways. The vessels that supply the Jatru Urdwa Pradesha are nourished and reinforced by Nasya. With the use of Panchendriya Vardhana oil, the obstruction caused by vitiated Vata Dosha in the Murdha (head) can be relieved, and the damaged nerves' normal functional capacity may be reestablished. Additionally, it aids in enhancing blood flow to brain lobes that are linked¹⁷.

9.3.8 Speech Therapy

Speech therapy was given for 30 days to enhance the movements of the tongue.

10. Conclusion

The present case study has shown satisfactory results by relieving signs and symptoms. On the basis of clinical observation, it can be concluded that by our treatment module, ALS having symptoms with tongue involvement shows gratifying results in time span of 30 days. The treatment's objective was to increase survival time and life quality with the prevention of disease progression. There is a further need of this treatment regimen on a greater number of patients to evaluate the exact findings and prove the module accurately.

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