DOI: https://doi.org/10.53350/pjmhs22166359

**ORIGINAL ARTICLE**

# Frequency of Dementia in Patients with Amyotrophic Lateral Sclerosis

SHOAIB AHMED[1], AMJAD HUSSAIN BALOUCH[2], MUKHTIAR ALI LAKHO[3], MUHAMMAD MUNWAR ALI[4], INAYATULLAH AWAN[5], SAFDAR HUSSAIN ARAIN[6]

[1]*Senior Registrar, Department of Neurology, Pir Abdul Qadir Shah Jeelani Institute of Medical Sciences, Gambat*
[2]*Assistant Professor & Head Department of Psychiatry, Pir Abdul Qadir Shah Jeelani Institute of Medical Sciences, Gambat*
[3]*Senoir Registrar, Department of Neurology, Shaheed Mohtarma Benazir Bhutto Medical University, Larkana*
[4]*Assistant Professor, Department of Neurosurgery, Chandka Medical College/Shaheed Mohtarma Benazir Bhutto Medical University, Larkana*
[5]*Associate Professor, Department of Psychiatry, Ghulam Muhammad Mahar Medical College, Sukkur*
[6]*Associate Professor & Head Department of Neurosurgery, Pir Abdul Qadir Shah Jeelani Institute of Medical Sciences, Gambat*
*Correspondence to: Dr. Shoaib Ahmed, E-mail: dr.shoaibmagsi@gmail.com, Cell: 0331-3328484*

## ABSTRACT

**Objective:** To determine the frequency of dementia in patients with amyotrophic lateral sclerosis.
**Study Design:** Cross-sectional study
**Place and Duration of Study:** Department of Neurology, Chandka Medical College Hospital, Larkana Pakistan from 14thJanuary 2019 to 13thJuly 2019.
**Methodology:** One hundred and ten patients were enrolled. Diagnosis was made after patient examination as proposed by memory impairment greater than 1 cognitive-disturbance.
**Results:** The mean age was 54.36±11.12 years and mean duration of disease was 18.46±7.13 months. Sixty seven (61%) were males and 43 (39%) were females. Dementia was found noted in 11 (10%) patients while 99 (90%) was found to benormal.
**Conclusion:** The frequency of dementia in patients with amyotrophic lateral sclerosis was found to considerable.
**Key words:** Amyotrophic lateral sclerosis, Epidemiology, Dementia, Incidence

## INTRODUCTION

Amyotrophic lateral sclerosis is a neuro-degenerative disorder that has many prominent clinical manifestations including muscle weakness, rapid loss of cortical and spinal neurons, wasting, respiratory failure which even leads to death. In 10% and 90% of the patients transmitted genetically and sporadically, respectively.[1] Few cognitive changes are also noticed in ALS patients.[2]

Even distribution of amyotrophic lateral sclerosis has been observed worldwide. Its prevalence is 5.4. 3.4 and 5.0 per 100,000 individuals in Europe, North America and Brazil, respectively.[3,4] It usually appears in late adulthood. Frontal and temporal lobes get involves in ALD which leads to personality and behavioral changes, language dysfunction and frontal executive deficits.[5] It is now recognized as 2nd most affecting disease in 65 years of age people.[6] Researches have proved that, it usually affects in 5th of 7th decades of life.[7]

In a study, it was found that, Dementia was seen in 12.6% of ALS patients.[8] In another study, It was found that, frequency of dementia was 23% in ALS patients.[9] In another study, dementia was 4.8% in ALS patients.[10] The results of this study was helped us to understand the burden of disease in our population and it was opened windows for newer research protocols and will set the priorities of the patient's treatment.

## MATERIALS AND METHODS

This cross-sectional study was carried out at Department of Neurology, Chandka Medical College Hospital, Larkana from 14th January 2019 to 13th July 2019 and 110 ALS patients were enrolled. All patients of bothgenders, age between 18-70years, diagnosed with ALS for 6 months were included. Patients with history of neurological disorders affecting cognition (majorstroke, severe head injuries, mental retardation), drug-dependence, severe mental illness and use of high-dose psychoactive medications were excluded. Demographic information including name, age and gender was also recorded. Diagnosis were made after patient examination as proposed by memory impairment greater than 1 cognitive-disturbance which includes apraxia, language disturbance and agnosia with considerable reduction in social and behavioral function. Data was entered and analyzed using SPSS-23.

## RESULTS

The mean age was 54.36±11.12 with CI 52.28-56.44years and mean duration of disease was 18.46±7.13 with CI 17.19-19.73 months (Table 1). There were 67 (61%) were males and 43 (39%) were females. Dementia was found in 11 (10%) patients while 99 (90%) was found to be normal (Table 2).

Table 1: Descriptive statistics of age and duration of disease (n=110)

| Variable | Mean±SD |
|---|---|
| Age (years) | 54.36±11.12 |
| Duration of disease (months) | 18.46±7.13 |

Table 2: Demographic information of the patients (n=110)

| Variable | No. | % |
|---|---|---|
| Gender | | |
| Males | 67 | 61.0 |
| Females | 43 | 39.0 |
| Dementia | | |
| Yes | 11 | 10.0 |
| No | 99 | 90.0 |

## DISCUSSION

Amyotrophic lateral sclerosis is the common motor neurodegenerative disorder that usually affects in late adulthood. It causes loss of nerves that plays significant role in connecting brain to muscles that results in voluntary upper and lower muscle weakening throughout brain to spinal cord.[11,12] Other degenerative diseases either only affect upper motor-neuron or lower motor-neuron.[13] Most of the patients showed cognitive/behavioral problems and dementia.[14,15]

The incidence of ALS in familial and sporadic FTD or FTD in ALS has not been extensively studied. A recent review of 151 patients with ALS showed increased risk of Parkinson disease (PD) and dementia in relatives based on clinical history.[16]

Dementia appeared to be higher in ALS groups than to the control group.[13,14] Likewise, PD chances were higher in patients who had ALS in families than to the normal family member group. However, these patients with ALS did not have neuropsychological testing to determine whether they had any evidence of dementia.

Chow et al[17] reported that 15% of all cases either developed ALS or had a first degree relative with ALS. Rosengren et al[18] found that14% of patients with FTD carried a diagnosis of definite ALS whereas another study, van Swieten[19] reported 36% had features of this condition. Majoor-Krakauer et al[16] showed that 16.7% of ALS out which 60% males and 40% females. Our study indicated that 67 (61%) were male and 43 (39%) were female with ALS patients.

## CONCLUSION

Dementia incidence in amyotrophic lateral sclerosis patients was found to considerable. Amyotrophic lateral sclerosis patients with elder age are significantly higher risk of dementia.

## REFERENCES


1. Chiò A, Calvo A, Mazzini L, Cantello R, Mora G, Moglia C, at al. Extensive genetics of ALS A population-based study in Italy. Neurology 2012;79(19):1983-9.
2. Kenna KP, McLaughlin RL, Byrne S. Delineating the genetic heterogeneity of ALS using targeted high-throughput sequencing. J Med Genet 2013;50:776-83.
3. Chio A, Logroscino G, Traynor BJ. Global epidemiology of amyotrophic lateral sclerosis: a systematic review of the published literature. Neuroepidemiology 2013;41:118-30.
4. Linden-Junior E, Becker J, Schestatsky P, Rotta FT, Marrone CD, Gomes I. Prevalence of amyotrophic lateral sclerosis in the city of Porto Alegre, in Southern Brazil. Arq Neuropsiquiatr 2013;71:959-62.
5. Neary D, Snowden JS, Gustafson L. Frontotemporal lobar degeneration: a consensus on clinical diagnostic criteria. Neurology 2008;51:1546-54.
6. Harvey RJ, Skelton-Robinson M, Rossor MN. The prevalence and causes of dementia in people under the age of 65 years. J Neurol Neurosurg Psychiatry 2003;74:1206-09.
7. Gislason TB, Ostling S, Borjesson-Hanson A. Effect of diagnostic criteria on prevalence of frontotemporal dementia in the elderly. Alzheimers Dement 2015;11:425-33.
8. Montuschi A, Iazzolino B, Calvo A, Moglia C, Lopiano L, Restagno G, at al. Cognitive correlates in amyotrophic lateral sclerosis: a population-based study in Italy. J Neurol Neurosurg Psychiatry 2014.
9. Rippon GA, Scarmeas N, Gordon PH, Murphy PL, Albert SM, Mitsumoto H, at al. An observational study of cognitive impairment in amyotrophic lateral sclerosis. Arch Neurol 2006;63(3):345-52.
10. Oh SI, Park A, Kim HJ, Oh KW, Choi H, Kwon MJ, et al. Spectrum of cognitive impairment in Korean ALS patients without known genetic mutations. PloS one 2014;9(2):871-3.
11. Robberecht W, Philips T. The changing scene of amyotrophic lateral sclerosis. Nat Rev Neurosci 2013; 14: 248-264.
12. Turner MR, Hardiman O, Benatar M, Brooks BR, Chio A, et al. Controversies and priorities in amyotrophic lateral sclerosis. The Lancet Neurology 2013; 12: 310-22.
13. Lomen-Hoerth C, Murphy J, Langmore S, Kramer JH, Olney RK, et al. Are amyotrophic lateral sclerosis patients cognitively normal? Neurology 2003;60:1094-7.
14. Phukan J, Pender NP, Hardiman O. Cognitive impairment inamyotrophic lateral sclerosis. Lancet Neurol 2007; 6: 994-1003.
15. Ringholz GM, Appel SH, Bradshaw M, Cooke NA, Mosnik DM, et al. Prevalence and patterns of cognitive impairment in sporadic ALS. Neurology 2005; 65: 586-90.
16. Majoor-Krakauer D, Ottman R, Johnson WG, Rowland LP.Familial aggregation of amyotrophic lateral sclerosis, dementia,and Parkinson's disease: evidence of shared genetic susceptibility. Neurology 1994;44:1872-7.
17. Chow TW, Miller BL, Hayashi VN, et al. Inheritance of frontotemporaldementia. Arch Neurol 1999;56:817-22.
18. Rosengren LE, Karlsson JE, Karlsson JO, Persson LI, Wikkelsø C. Patients with amyotrophic lateral sclerosis and other neurodegenerative diseases have increased levels of neurofilament protein in CSF. J Nneurochem 1996;67(5):2013-8.
19. van Swieten JC, van den Hout JHW, van Ketel BA, Hijdra A, Wokke JHJ, van Gijn J. Periventricularlesions in the white matter on magnetic resonance imaging inthe elderly. Brain 1991; 114, 761-74.