

Table SI. Summary of patients from whom CSF was obtained.

| Patient | Age | Gender |
|----------------|------------|---------------|
| Patient 1 | 67 | Male |
| Patient 2 | 77 | Male |
| Patient 3 | 69 | Female |
| Patient 4 | 67 | Female |
| Patient 5 | 74 | Female |
| Control 1 | 64 | Male |
| Control 2 | 43 | Male |
| Control 3 | 68 | Female |

Table SII. Summary of patients from whom brain samples were obtained.

| Patient | Zone | Age | Gender |
|----------------|---|------------|---------------|
| Patient 6 | FC C O WM O GM | 65 | Female |
| Patient 7 | FC GM FC WM C O WM+GM | 44 | Male |
| Patient 8 | FC WM FC GM C WM C GM O WM+GM | 72 | Female |
| Patient 9 | FE | 53 | Male |
| Patient 10 | FE | 56 | Male |
| Patient 11 | FE | 56 | Female |
| Control 4 | FE | 77 | Female |
| Control 5 | FE | 63 | Male |
| Control 6 | FE | 62 | Female |
| Control 7 | FC | 58 | Female |

FE : External frontal cortex ,FC: Frontal cortex, C: Cerebellum, O : Occipital cortex , WM: white matter , GM: Grey matter

Table SIII. Human proteins which are common to the three ALS patients, that are not present in control brain.

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| 3'(2'),5'-bisphosphate nucleotidase 1 |
| 6-phosphofructo-2-kinase/fructose-2, 6-bisphosphatase |
| Actin-related protein 2/3 complex subunit 5 |
| Acyl-coenzyme A thioesterase 9 |
| Acylphosphatase-1 |
| ADP-ribosylation factor GTPase-activating protein 1 |
| Amine oxidase |
| Aminopeptidase B |
| Annexin A3 |
| Apolipoprotein E |
| ATPase ASNA1 |
| ATP-dependent (S)-NAD(P)H-hydrate dehydratase |
| Beta-adrenergic receptor kinase 1 |
| Bifunctional coenzyme A synthase |
| Calcium/calmodulin-dependent protein kinase I |
| Calpain small subunit 1 |
| Calsyntenin-1 |
| Centrosomal protein of 170 kDa protein B |
| Clavesin-2 |
| Cytochrome b-c1 complex subunit 7 |
| Cytochrome c oxidase subunit 6B1 |
| Cytoplasmic dynein 1 light intermediate chain 1 |
| Cytoplasmic protein NCK2 |
| Developmentally-regulated GTP-binding protein 1 |
| Dipeptidyl-peptidase 6 |
| DPYSL3 protein |
| F-actin-capping protein subunit beta |
| Fibroblast growth factor 12 |
| Flavin reductase (NADPH) |
| Glutamate receptor 2 |
| Glutamic acid decarboxylase |
| Glutathione reductase |
| Glycerophosphodiester phosphodiesterase |
| Glycogen debranching enzyme |
| Guanylate cyclase soluble subunit beta-1 |
| Haptoglobin |
| Hemopexin |
| Histone-binding protein RBBP4 |
| HLA-B associated transcript 3, isoform CRA_ |
| Hyaluronan and proteoglycan link protein 4 |
| Inositol polyphosphate-4-phosphatase |
| Isochorismatase domain-containing protein 2 |
| Keratin, type I cytoskeletal 17 |
| Kinesin-like protein KIF3A |
| Liprin-alpha-3 |
| L-xylulose reductase |
| Lysine--tRNA ligase |
| MAGUK p55 subfamily member 6 |
| MAPRE1 protein (Fragment) |
| Methylcrotonoyl-CoA carboxylase beta chain |
| MOSC domain-containing protein 2, mitochondrial |

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| Myelin proteolipid protein |
| MYO5A variant protein |
| NAD kinase domain-containing protein 1 |
| NADH dehydrogenase |
| Neural cell adhesion |
| Neuronal growth regulator 1 |
| Neuronal pentraxin-1 |
| Nicastrin |
| Nicotinate-nucleotide pyrophosphorylase |
| N-terminal EF-hand calcium-binding protein 2 |
| Nucleoside diphosphate kinase (Fragment) |
| Phosphatase and actin regulator 1 |
| Phosphofurin acidic cluster sorting protein 1 |
| Phospholipase D3 |
| Phosphorylase |
| Plasma membrane calcium-transporting ATPase 3 |
| Plexin-A1 |
| PRKCA-binding protein |
| Programmed cell death protein 6 |
| Proline synthase co-transcribed bacterial homolog protein |
| Proprotein convertase subtilisin/kexin type 1 |
| Protein kinase C gamma type |
| Protein NDRG4 |
| Protein SCA1 |
| Protein TFG |
| Purine nucleoside phosphorylase |
| RAB11B protein |
| Rap1 GTPase-activating protein 2 |
| Retinol dehydrogenase 11 |
| Rho guanine nucleotide exchange factor 7 |
| Sepiapterin reductase |
| Septin-5 |
| Sideroflexin-5 |
| Spectrin beta chain, non-erythrocytic 4 |
| Sterol-4-alpha-carboxylate 3-dehydrogenase |
| Succinate-semialdehyde dehydrogenase |
| Synapsin III |
| Synaptogyrin-3 |
| SYNPO protein |
| TNF receptor-associated protein 1 variant (Fragment) |
| Trimethyllysine dioxygenase |
| TUBB protein |
| Type I inositol 1,4,5-trisphosphate 5-phosphatase |
| Vacuolar protein sorting-associated protein 26B |

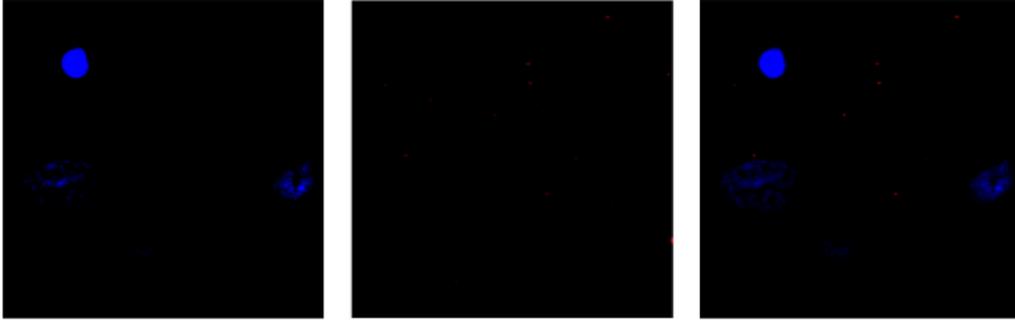
Figure S1. Immunohistochemistry analysis of brain sections from the frontal cortex of control individuals. Brain sections (frontal cortex) from control individuals 4, 5 and 6 were observed with a confocal laser scanning microscope. DAPI appears in blue and anti-C. glabrata is shown in red. The different panels in the figure are indicated. Scale bar: 10 μm .

Dapi

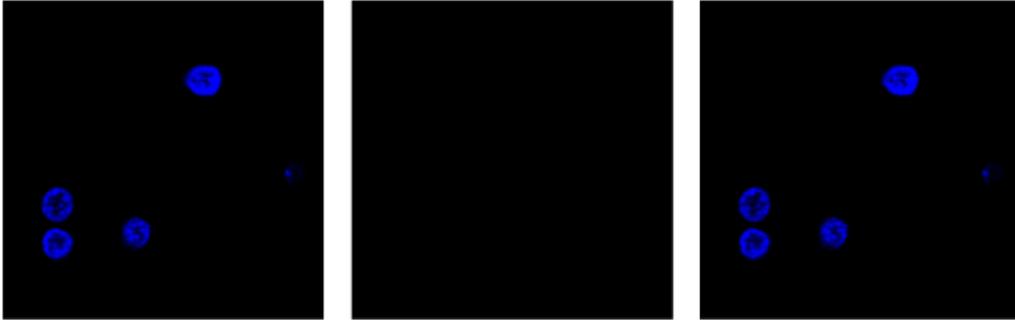
anti-C. glabrata

Merge

Control 4



Control 5



Control 6



Figure S2. Analysis of proteins by SDS-PAGE from brain samples. Proteins from homogenized brain samples (frontal cortex) were separated by SDS-PAGE (12.5%) and stained with Coomassie blue.

