**Neuropsychological assessment**

At the tertiary referral center, a comprehensive neuropsychological assessment was performed at month 24 and month 31 from symptom onset by an experienced neuropsychologist and evaluated: global cognitive functioning with the Mini-Mental State Examination (1); frontal functioning with the Frontal Assessment Battery (2); verbal memory with the digit span forward (3); non-verbal memory with the spatial span forward (3), and Benson’s Figure delayed recall (4); attention and working memory with the attentive matrices (5), the digit span backward (6), and the Modified Card Sorting Test (7); visuospatial abilities with the Benson’s figure copy (4) and the copy of simple figures (8); praxia with orofacial and ideomotor limb apraxia tests (5); social cognition with the Story-based Empathy Task (9); behaviour with the Neuropsychiatric inventory (10) and the Frontal Behavioural inventory (11); disease severity with the clinical dementia rating scales (12, 13), and autonomy with basic (14) and instrumental (15) activities of daily living.

The patient also underwent a comprehensive language testing which evaluated: syntactic comprehension with the Token test (16); confrontation naming and single word comprehension with the CaGi test (17); object knowledge with the Pyramids and Palm Trees Test (18); repetition, reading, and writing with the Aachener Aphasie Test (19). For investigating motor speech and speech fluency, we recorded, respectively, multiple repetitions of single words (not standardized) and speech samples while the patient described the image of the picnic picture subtest of the Western Aphasia Battery (20).

**MRI acquisition**

MRI acquisition was performed at the tertiary referral center. Using a 3.0 T scanner (Ingenia CX, Philips), the following brain MRI sequences were obtained at month 24 and month 31: 3D T1-weighted (TFE) (TR=7 ms; TE=3.2 ms; flip angle=9 [degrees]; 204 contiguous sagittal slices with voxel size=1 x 1 x 1 mm, matrix size=256 x 240, FOV=256x240 mm2); 3D FLAIR (TR=4800 ms; TE=267 ms; TI=1650 ms; ETL=167; NEX=2; 192 contiguous sagittal slices with voxel size=0.89 x 0.89 x 1 mm, matrix size=256 x 256, FOV=256x256 mm2); and 3D T2 (TR=2500 ms; TE=330 ms; ETL=117; NEX=1; 192 contiguous sagittal slices with voxel size=0.89 x 0.89 x 1 mm, matrix size=256 x 258, FOV=256x256 mm2).

**Progranulin sequencing**

At month 24 total genomic DNA was extracted from peripheral blood using NucleoSpin Blood L Extraction Kit (Macherey-Nagel) following the manufacturer instructions. *GRN* (Homo sapiens, ENSG00000030582.18, LRG\_661) whole coding region and the intron-exon junctions were analyzed with a Sanger protocol (21).

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