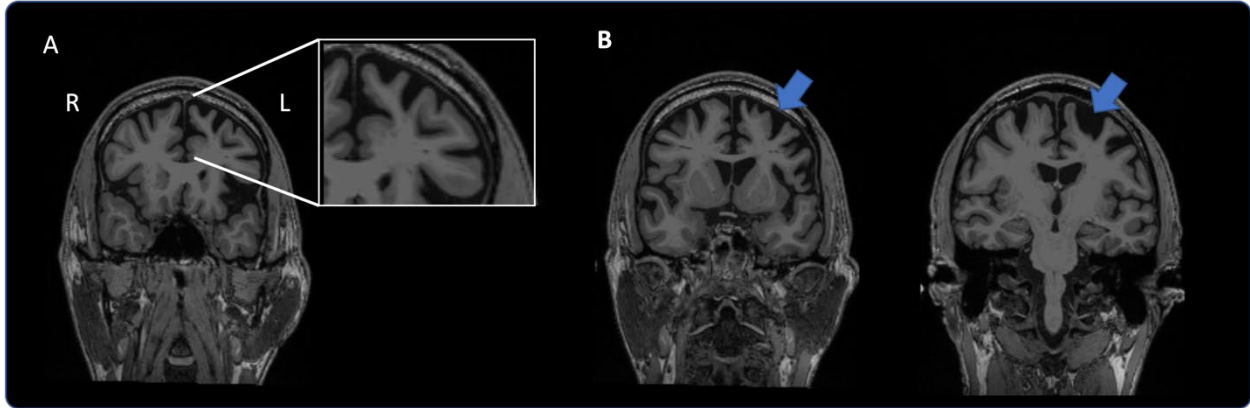
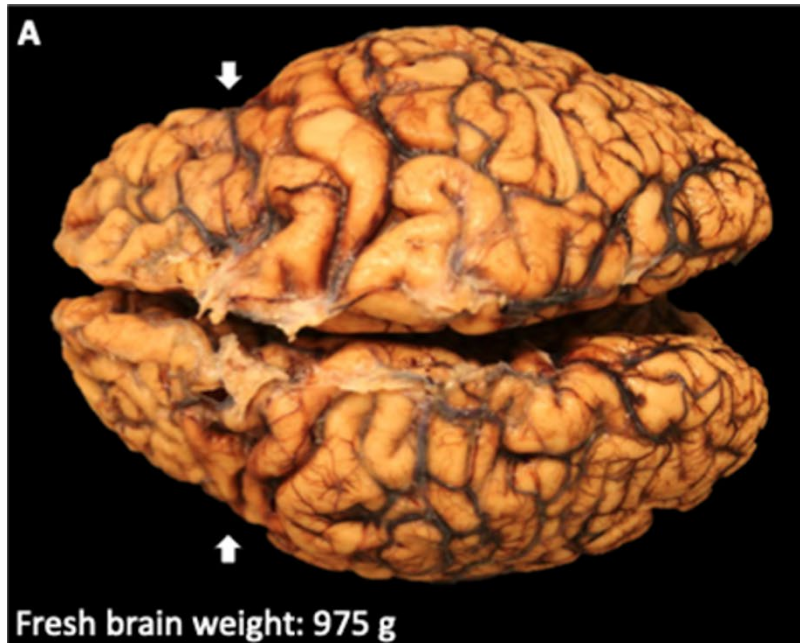


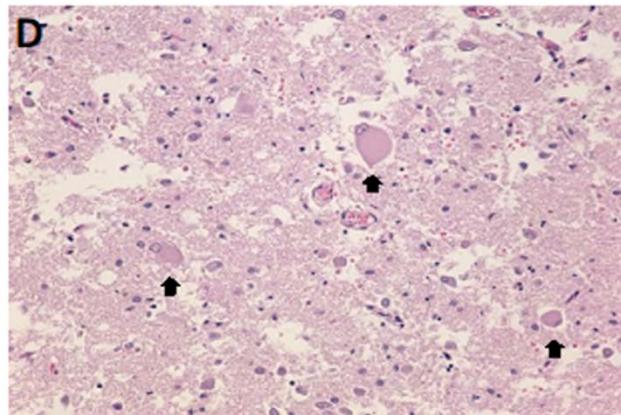
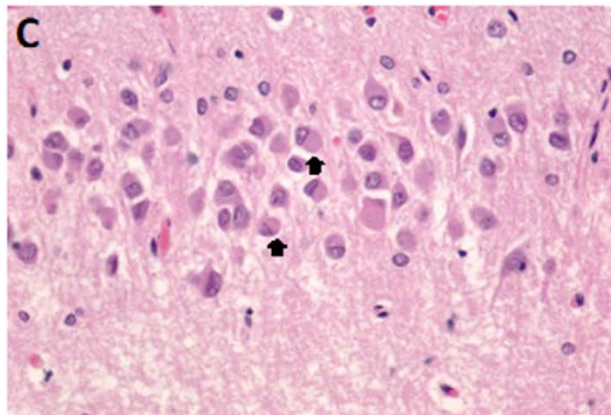
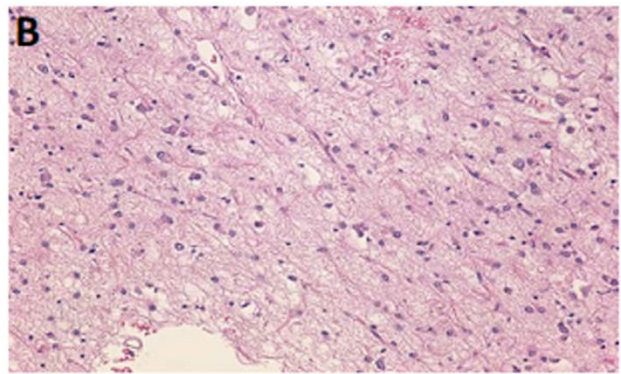
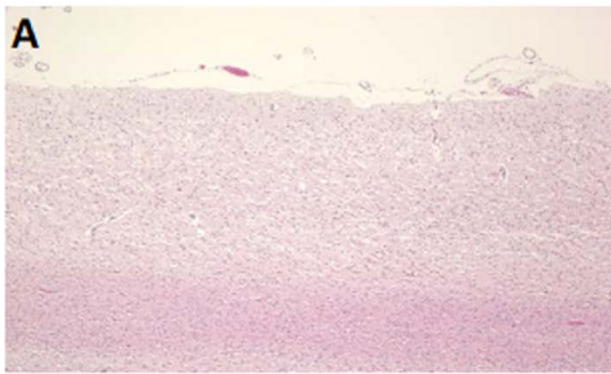
Supplemental Figure 1. Brain MRI, 2 years after symptom onset (age 60). Coronal T1-weighted sequences. *A*: Focal atrophy in left superior frontal gyrus (SFG), anterior region. (Inset magnifies SFG) *B*: Focal atrophy in left SFG; more posterior regions (arrows).



Supplemental Figure 2. Postmortem neuropathology, gross examination. (A) Superior view showing significant frontal atrophy (brain weight: 975 grams) with sharp demarcation of atrophy (arrows). (B) Coronal section of left frontal lobe showing severe cortical atrophy with shrunken gyri, expanded sulci, and yellowish discoloration of the subcortical white matter.



Supplemental Figure 3. Postmortem neuropathology, microscopic findings (hematoxylin & eosin). (A) Low-power image of cerebral cortex showing significant neuronal loss, gliosis, spongiform change, and gliotic white matter. (B) Medium-power image of cerebral cortex showing severe neuronal dropout and gliosis. (C) High-power view of dentate gyrus of hippocampus showing frequent, round, eosinophilic intracytoplasmic inclusions characteristic of Pick bodies (arrows). (D) Low-power view of temporal cortex highlighting ballooned neurons with abundant glassy eosinophilic cytoplasm with eccentric nuclei (arrows).



Supplemental Table 1: Detailed language assessment at presentation (Age 60; 2 years after symptom onset)

Domain	Performance
Expressive Language	
Fluent?	Paucity, word-finding pauses
Grammatical?	Yes
Phrase length?	>6 words
Paraphasic, phonetic errors?	No
Motor speech impairment?	No
Praxis	Normal upper limb, oro-buccal, instrumental and complex praxis
Fluency	
Letter	3 "P" words in one minute FAS task 9: 2F/3A/4S (1 st %)
Category	9 animals, 6 fruits, 5 vegetables in one minute (1 st %)
Design	12-25 th %
Naming	BNT: 60/60 CSB: 63/64
Writing	Provided reasonable written picture description and spontaneous written sentence, with intact spelling and grammar.
Reading	WAB Comprehension of Sentences: 40/40 WAB Reading Commands: 10/10 decoding; 10/10 performance
Repetition	Intact for words, phrases, and sentences with only one word omission on the longest item WAB Repetition: 96/100
Auditory Comprehension	Able to follow short directions. Some difficulty with lengthy commands, especially when the agent completing the action appeared at the end of the sentence (e.g., "point to the pen with the book") WAB Auditory Commands: 62/80
BNT= Boston Naming Task CSB= Cambridge Semantic Battery WAB= Western Aphasia Battery	

Supplemental Table 2: Detailed neurobehavioral status examination in non-language cognitive domains at presentation (Age 60; 2 years after symptom onset)

Domain	Performance
Attention	Digit span (9 th %): <ul style="list-style-type: none"> - Forward: 5 - Backward: 4 Spatial span: <ul style="list-style-type: none"> - Forward: 5 (5th %) - Backward: 4 (9th %)
Orientation	Preserved
Calculation	Quick, accurate, arithmetic performance Serial sevens without error
Executive function	Accurate and well-organized clock draw Impulse Control <ul style="list-style-type: none"> - CPT-II: High level of omissions, variability, and perseverative response (atypical) - Tower Test: short first move time, rule violations, impulsivity (atypical) Abstract Reasoning <ul style="list-style-type: none"> - 20 questions test: concrete approach (9th %)
Memory	CERAD Word List <ul style="list-style-type: none"> - Encoded 3/7/7 over 3 trials, total 17/30 (10th %) - Freely recalled 8/10 after 5-minute delay (66th %) - Recognized 100% with one false positive (21st %) Visual Memory <ul style="list-style-type: none"> - Warrington Face Memory Test (95th %) Verbal Memory <ul style="list-style-type: none"> - Warrington Word Memory Test (25th %)
Visuospatial function	Accurate 3-dimensional cube drawing Top-down organization on complex figure copy was very poor
Global Function	MoCA: 28/30 with points lost for word fluency and repetition DRS: 132/144 GDS: 4 (normal) Pre-morbid intellectual abilities estimated in average range <ul style="list-style-type: none"> - AMNART: 109
AMNART = American version of the National Adult Reading Test CERAD = Consortium to Establish a Registry for Alzheimer's Disease Word List task CPT = Continuous Performance Task DRS = Dementia Rating Scale GDS = Global Deterioration Scale MoCA = Montreal Cognitive Assessment	