Table 1. Summary of clinicopathological data.

ABC score indicates the score provide for Aβ deposition (A), for Braak stage of neurofibrillary degeneration (B), and neuritic plaque score (C)(28). PSP: progressive supranuclear palsy; MCI: mild cognitive impairment.

	Case-1	**Case-2**	**Case-3**
Age at death (years)	83	72	91
Sex	male	female	female
Duration of illness (years)	20	5	20
Clinical diagnosis	PSP+ MCI	PSP+dementia	Unclassified dementia
Symptoms at early stage	Imbalance and gait disorder	Incoordination	Not available
Symptoms at late stage	Cognitive decline Supranuclear gaze palsy 2 grand mal seizures	Cognitive decline Supranuclear gaze palsy Parkinsonism	Cognitive decline
Motor-Dementia interval	15 years	0 years	0 years
Family history	no	no	yes
ApoE	E2/E3	E3/E3	E2/E3
H1/H2 haplotype	H1/H1	H1/H1	H2/H2
MAPT mutation	none	none	none
Brain weight	1378	1147	1066
ABC score	A0B2C0	A1B1C0	A3B3C3
pTDP-43	no	no	yes
α-synuclein	no	no	no
Hippocampal sclerosis	no	yes	yes
Other	-	Focal encephalitis in brainstem Lacunar infarcts in Globus pallidus and brainstem	-

Table 2. Anatomical distribution of neuronal (NFT: neurofibrillary tangle; PT: pretangle; SB: spherical body), astroglial (AG), and oligodendroglial (OG; in the adjacent white matter) tau pathologies. – indicates none; + : occasional; ++ moderate number; +++: abundant/many. * indicates thorn shaped astrocytes in the white matter; TA: tufted astrocytes; RA: ramified astrocytes; GFA: granular/fuzzy astrocytes; subthal: subthalamic nucleus

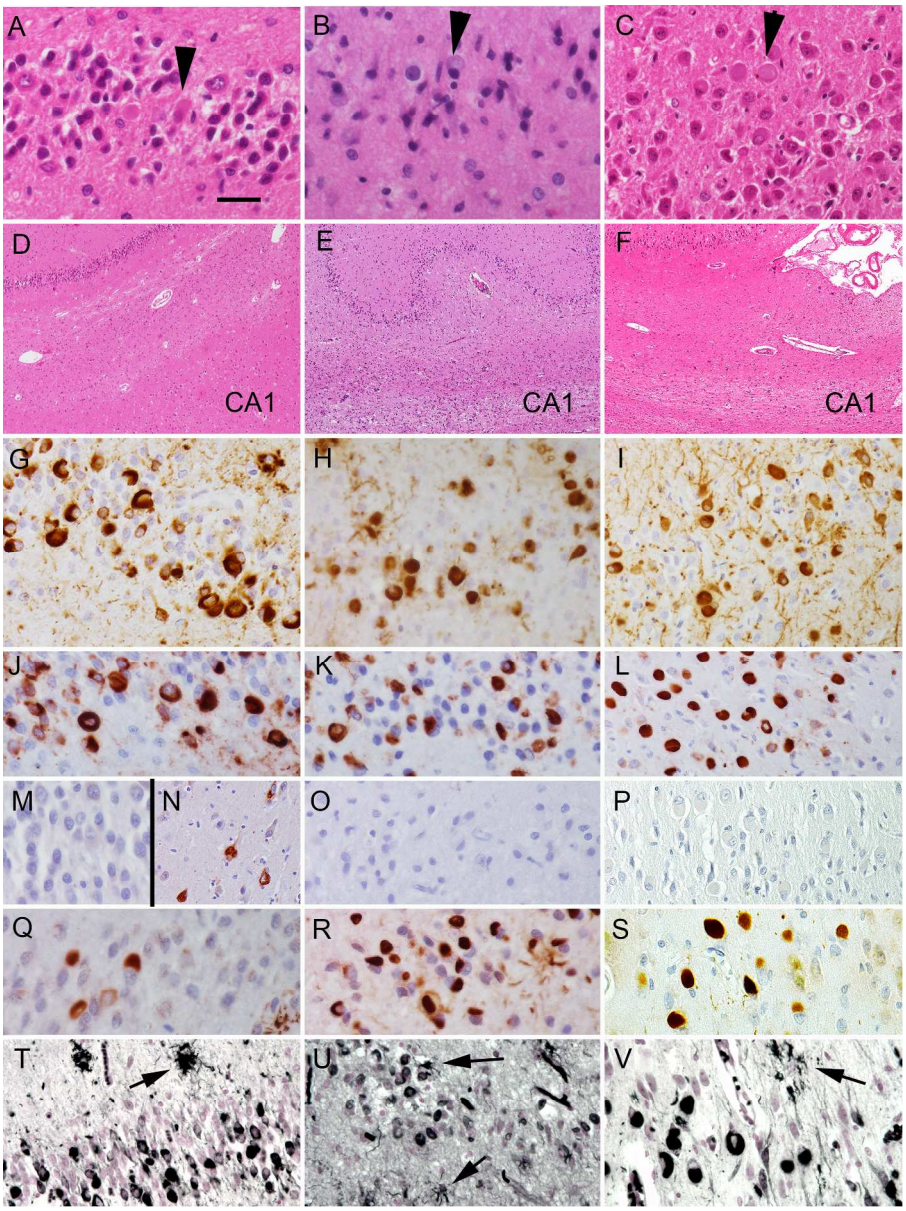
	Case-1				Case-2				Case-3			
Region/Pathology	NFT/PT	SB	AG	OG	NFT/PT	SB	AG	OG	NFT/PT	SB	AG	OG
Frontal	+	-	+^{TA}	+	+	-	+++^{TA}	+	++	-	-	+
Temporal	+	-	-	-	+	-	+++^{TA}	++	+++	-	-	+
Parietal	-	-	-	-	-	-	+++^{TA}	++	++	-	+*	+
Anterior cingulate	++	-	+^{TA}	+	+	-	+++^{TA}	++	++	-	-	+
Entorhinal	+++	-	+^{TA}	+	+	-	+++^{TA}	+	+++	-	-	-
Hippocampus												
CA1/Subiculum	+++	+	++^{TA}	+++	+	+	+++^{TA}	++	+++	+	+$^{TA/RA}$	+
CA2/3	+++	+	+++^{TA}	+++	+	+	+++^{TA}	-	+++	+	++$^{TA/RA}$	+
CA4	++	-	+++^{TA}	-	+	-	+++^{TA}	-	+	-	++$^{TA/RA}$	-
Dentate gyrus	+	+++	++$^{TA/TSA}$	-	+	++	++^{TA}	-	+	+++	+$^{TA/RA}$	-
Amygdala	+++	-	+++$^{TA/GFA}$	+++	+++	+	+++^{TA}	++	+++	-	++^{GFA}	+
Striatum	++	-	+++^{TA}	+	+	-	+++^{TA}	+	+	-	-	-
Globus pallidus	+	-	++^{TA}	+++	++	-	+^{TA}	+++	-	-	-	-
Thalamus+Subthal	++	-	-	+	++	-	+++^{TA}	+++	++	-	-	-
Substantia nigra	++	-	-	-	+++	-	+^{TA}	+++	+	-	-	-
Locus coeruleus	+++	-	-	-	+	-	-	+	++	-	-	-
Pontine base	-	-	-	+	+	-	-	+++	-	-	-	+
Inferior olives	+++	-	-	-	+	-	-	+	-	-	-	-
Hypoglossal nucleus	-	-	-	-	-	-	-	-	-	-	-	-
Dorsal vagus nucleus	++	-	-	-	-	-	-	-	-	-	-	-
Dentate nucleus	++	-	-	-	+++	-	-	+	-	-	-	-

Figure legends

Figure 1. Basophilic inclusions in the dentate gyrus (A-C; indicated by arrowheads) and variable cell loss in the CA1 subregion (D-F) in the three cases (A,D: case 1; B, E: case 2; C, F: case3) reported in this study. Immunostaining for PHF-1 (G-I), 4R tau (J-L), 3R tau (M-P), p62 (Q-S) and for Gallyas silver staining (T-V) in cases 1 (G, J, M, N, Q, T), 2 (H, K, O, R, U) and 3 (I, L, Q, P, S, V) demonstrating the spherical inclusions in the dentate gyrus. Immunostaining for 3R tau in the CA1 subregion (N) demonstrates neurofibrillary tangles in case 1 corresponding to the immunoblot results. Note the argyrophilic astrocytes in T-V (arrows). The bar in A represents 25 μm for A-C, G-V and 100 μm for D-F.

Figure 2. Immunostaining for PHF-1 in the CA1 (A-C), CA4 (D-F) subregions, amygdala (G-I), frontal cortex (J-L) and putamen (M-O) in cases 1 (A, D, G, J, M), 2 (B, E, H, K, N) and 3 (C, F, I, L, O). Note the numerous tau positive astrocytes in case 2 in all subregions, while less are seen in case 1 together with neurofibrillary tangles in the CA1 subregion. Case 3 exhibits neurofibrillary tangles and dystrophic neuritic plaques together with astrocytes in CA subregions and amygdala (* in image I indicates white matter in amygdala with thorn shaped astrocytes compatible with ARTAG). The bar in A represents 40 μm for A and 100 μm for B-O.

Figure 3. Biochemical analysis of tau in Triton X-100 insoluble fraction extracted from frozen brain tissues. (A) Total-tau (17025) and (B) phosphorylated-tau (PHF-1) immunoblot showed abundance of tau pathology in Case-1 and Case-2 Hp, but not in Case-1 Pu. Isoform specific immunoblot revealed predominant 4R-tau pathology in both cases (C) consistent with IHC finding. Further, Case-1 Hp was also immune-reactive to 3R-tau antibody (d). Hp = hippocampus, Pu = putamen; Alzheimer disease (AD), corticobasal degeneration (CBD), and Pick`s disease (PiD) control samples are also shown.



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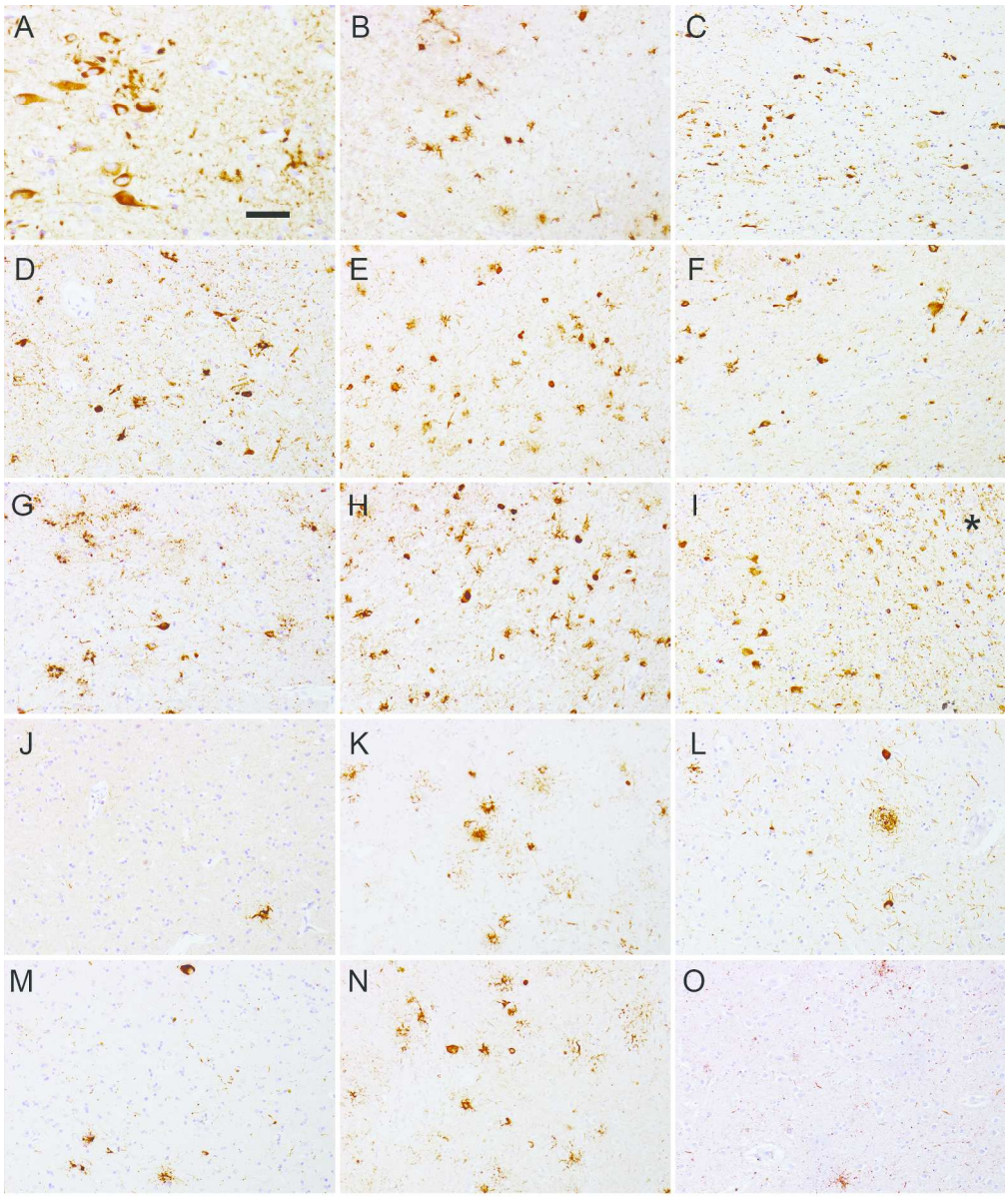

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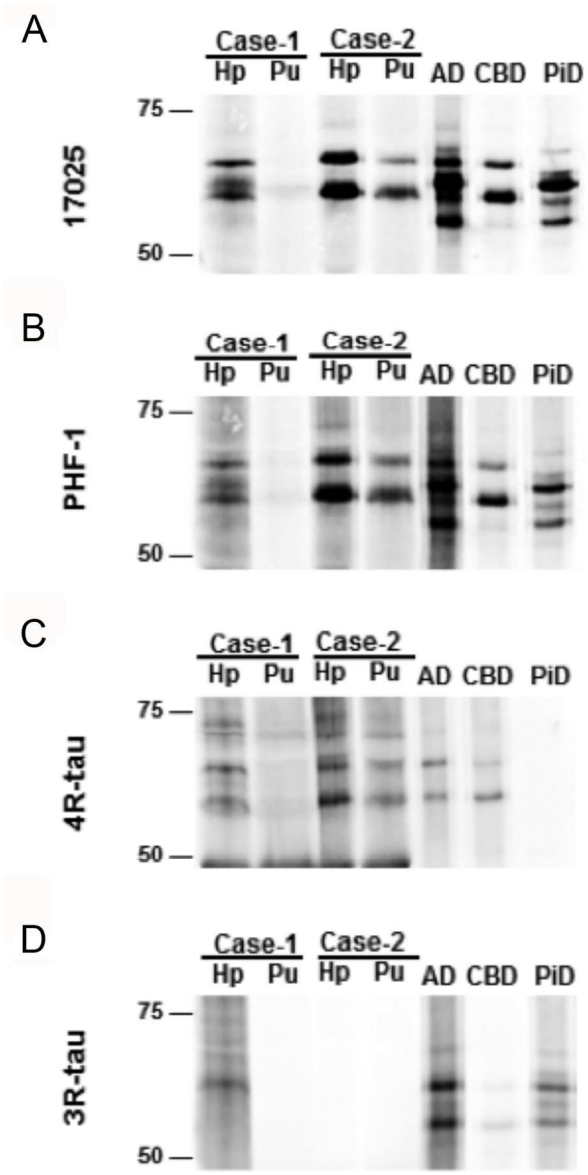

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