

Figure e-1. Macroscopic appearance of Case 1 (A) The left hemibrain appears normal. (B) A coronal section did not reveal any abnormalities. (C-D) Transverse sections of the midbrain (C) and pons (D) showed decreased pigmentation in the substantia nigra (arrow in C) and locus ceruleus (arrow in D).

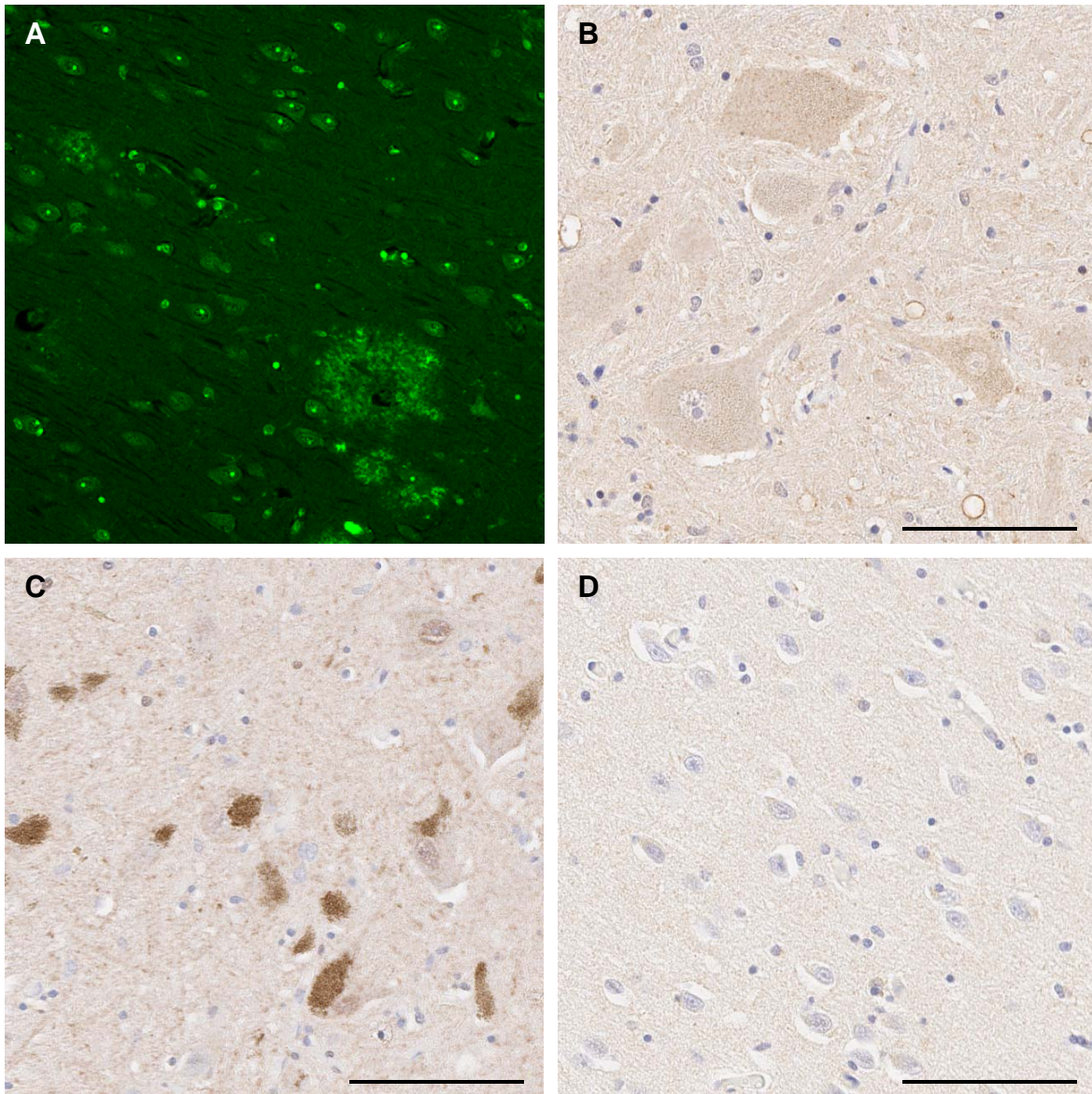


Figure e-2. Histologic examination of additional pathologic proteins associated with neurodegenerative disease (A) Thioflavin-S fluorescent microscopy showed mild senile changes of Alzheimer type. (B-D) Immunohistochemical staining did not detect SOD1 inclusions within the spinal cord (B), or α -synuclein within the midbrain (C), FUS inclusions within the amygdala (D). Bar represents 100 μ m.

ALS	ALS/FTLD		FTLD
SOD1			
VAPB	TARDBP		
OPTN	FUS/TLS	C9ORF72	
	ANG	VCP	GRN
		DCN1	CHMP2B

Figure e-3. Genes associated with adult onset, autosomal dominant ALS and FTLD may have overlapping clinical features