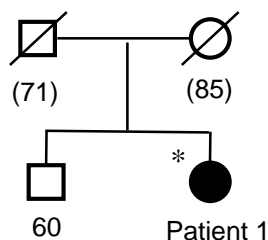**Supplementary table e-1.**

Comparison of genetic, clinical and MRI features between the present family and reported family with deletion upstream of *LMNB1*

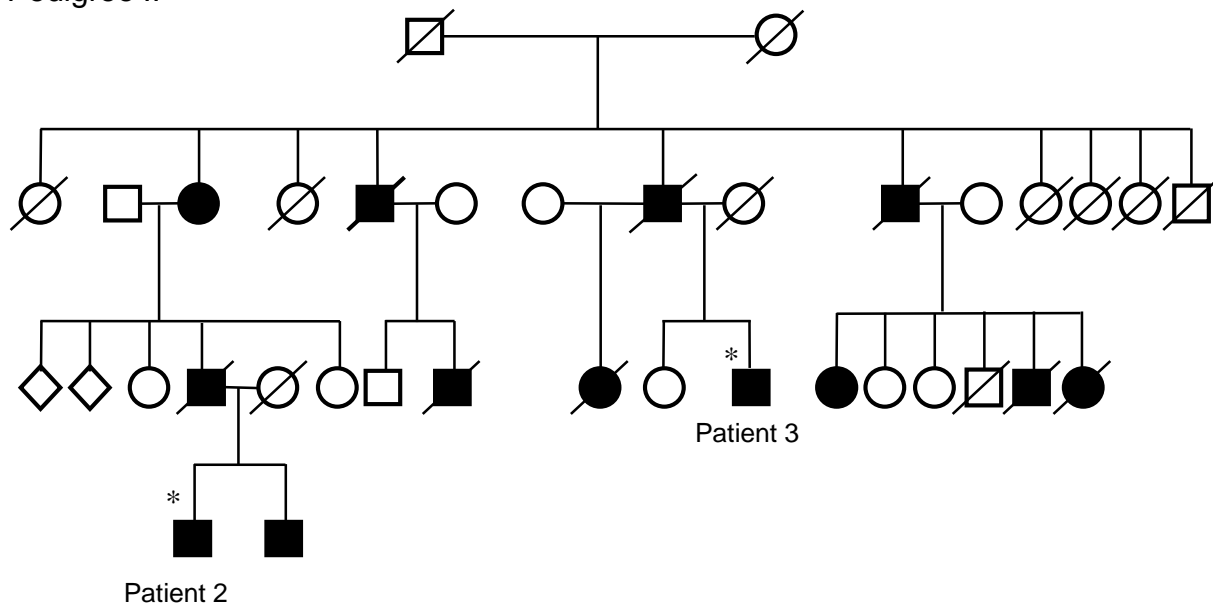|	Present family	Reported family[8]
Genetic feature		
Range of the deletion (kb)	249	660
Clinical features		
Age at onset (y)	43, 34 (mean = 38.5)	41-53 (mean = 47.2)
Onset with autonomic disturbance	2/2	0/22
Pyramidal signs	2/2	12/22
Ataxia	2/2	0/22
Cognitive impairment	2/2	N.A.
MRI features		
Bilateral white matter lesions	+	+
MCP signs	+	−
Anterior temporal lesions	+	−

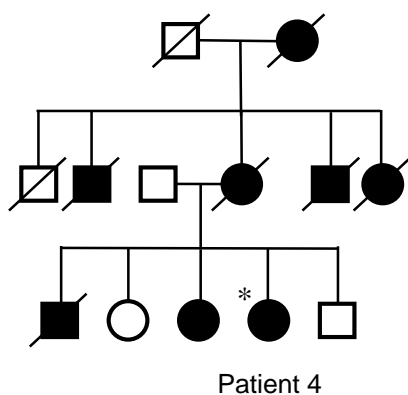N.A., data not available; MCP, middle cerebellar peduncle

Supplementary figure e-1

Pedigree I

Pedigree II

Patient 2

Patient 3

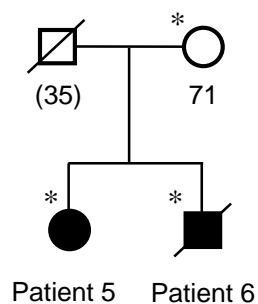Pedigree III

Pedigree IV

Patient 4

Patient 5   Patient 6

Figure legend. Family trees of *LMNB1*-related ADLD patients in this study.
Square indicate male, and circles indicate female. Black symbols indicate affected individuals.
Asterisks indicate subjects whose blood samples were examined for genetic analysis. Number
under the symbols indicates present age. Number in parenthesis indicate age at death.
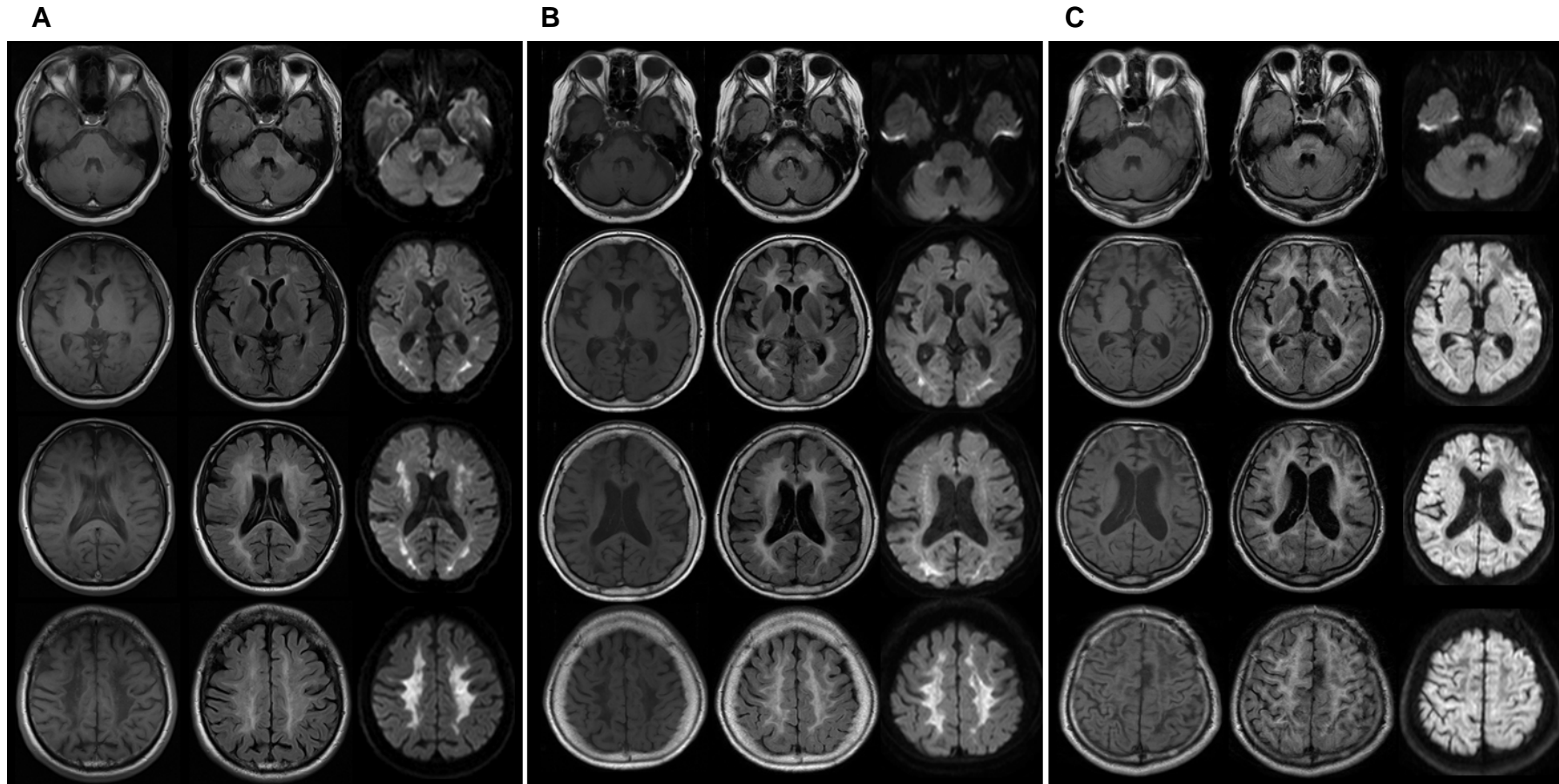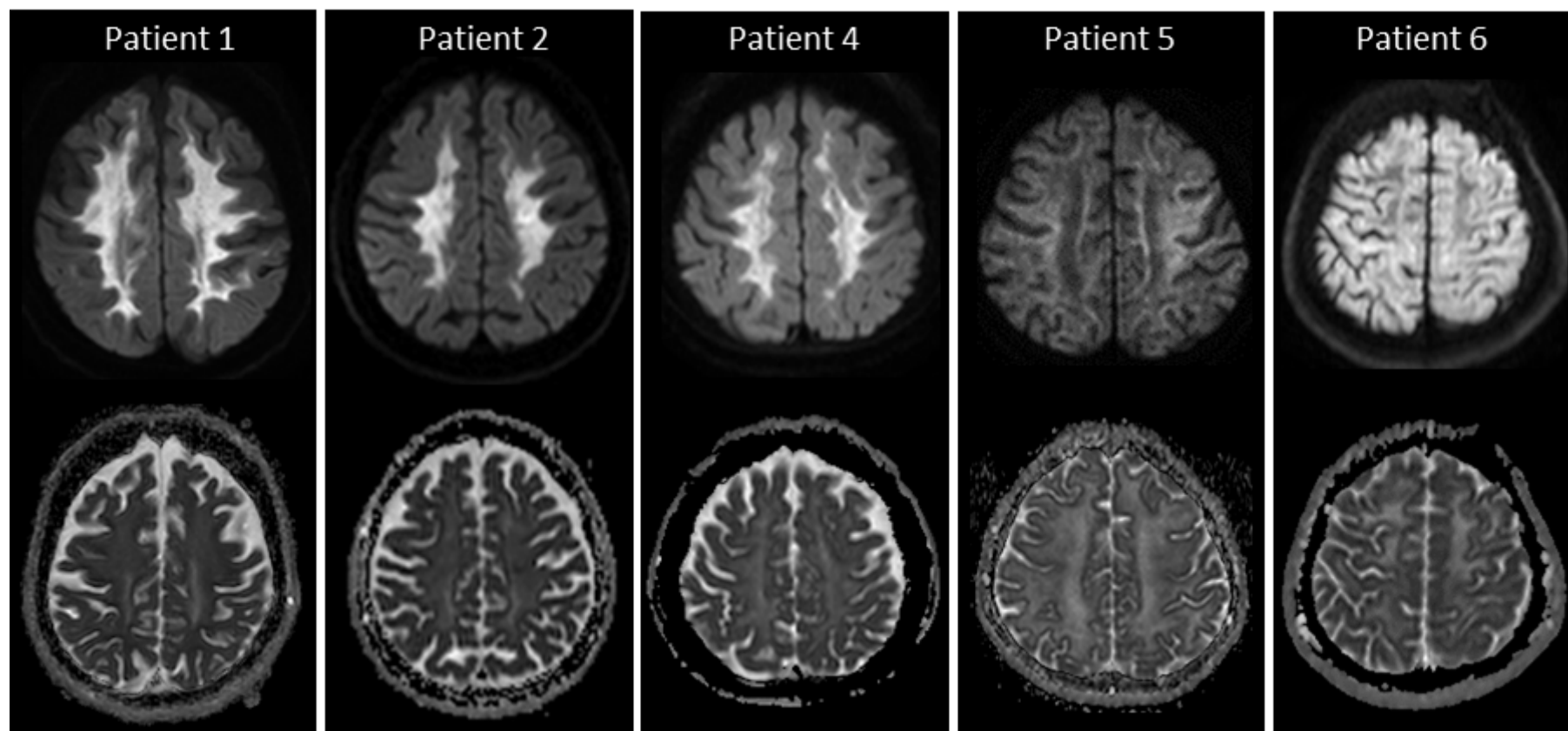
Supplementary figure e-2

Figure legend. MRI findings of *LMNB1*-related ADLD patients.

(A) Findings of MRI with T1WI (*left panel*), FLAIR (*middle panel*), and DWI (*right panel*) of *patient 2* with *LMNB1* duplication at age of 57 years.

(B) Findings of MRI with T1WI (*left panel*), FLAIR (*middle panel*), and DWI (*right panel*) of *patient 4* with *LMNB1* duplication at age of 70 years.

(C) Findings of MRI with T1WI (*left panel*), FLAIR (*middle panel*), and DWI (*right panel*) of *patient 6* with upstream deletion of *LMNB1* at age of 42 years.

Supplementary figure e-3

Supplementary figure 3. ADC map of *LMNB1*-related ADLD patients.

DWI (*upper panel*) and ADC map (*lower panel*) of patients with duplication (*patients 1, 2,* and *4*) and deletion upstream of *LMNB1* (*patients 5* and *6*) were shown. The areas of hyperintensity on DW images show hyper- or iso-intensity in ADC maps.