

Table 1. Clinical features of the patients with familial or sporadic CANVAS

	Family A-II-2	Family B-II-2	Family C-II-4	Patient 1
Sex	M	M	M	M
Age at onset	48	77	64	55
Age at examination	60	89	70	72
Inheritance	AR	AR	AR	S
Cerebellar ataxia	+	+	+	+
Neuropathy	+	NA	+	+
Vestibular failure	+	+	+	+
Chronic cough	+	NA	+	+
Upper limb reflexes	normal	NA	exaggerated	normal
Lower limb reflexes	normal	NA	exaggerated	decreased
Babinski signs	-/-	-	+/+	-/-
Sleep apnea	+	-	-	-
MRI findings	Cerebellar atrophy	Cerebellar atrophy	Cerebellar atrophy	Cerebellar atrophy
Repeat expansion	AAGGG	AAGGG	AAGGG	ACAGG

M: male, AR: autosomal recessive, S: sporadic, NA: not available