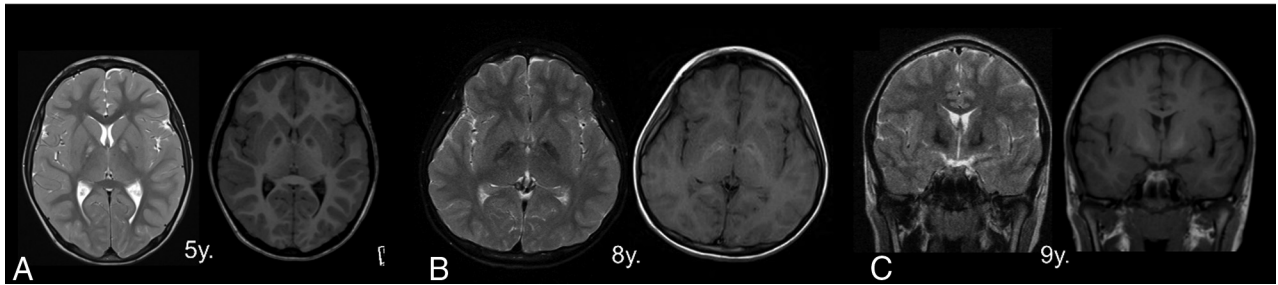
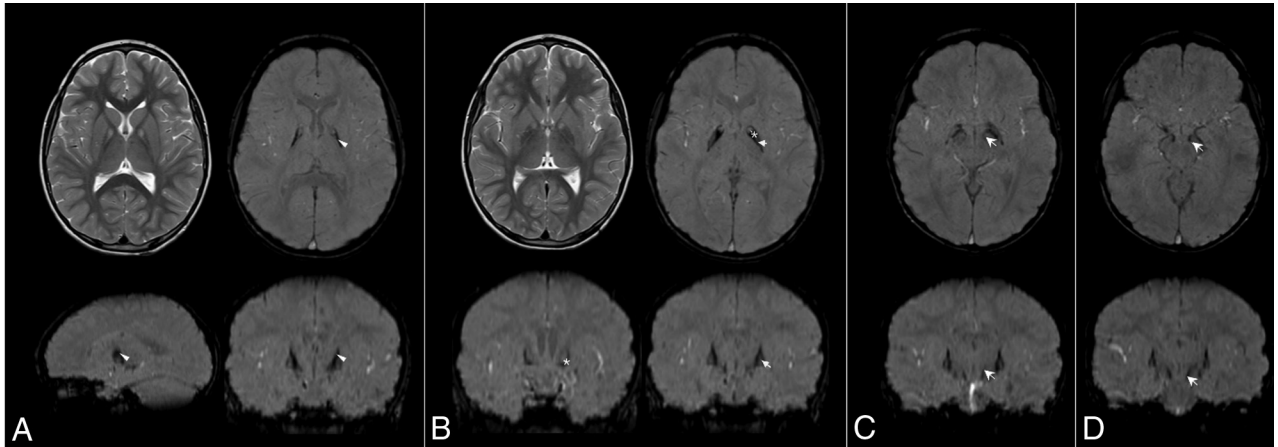


ON-LINE FIG 1. The eye-of-the-tiger sign. Axial T2-weighted images at level I and just above level II of the anterior commissure and coronal images (level III) from 2 patients with PKAN (patient A, 5 years of age; patient B, 9 years of age) and a healthy control (8 years of age) at 3T MR imaging. The isolated T2-hyperintense center in the GP is defined as “hyperintense center” surrounded by isointense signal relative to the adjacent internal capsule (patient A). a indicates the round hyperintense center; b, the surrounding hypointense area.



ON-LINE FIG 2. Signal changes on T1-weighted images. T1 hypointensity is observed isolated in the T2-hyperintense center (A). T1-hyperintensity in the surrounding area is correlated with the T2-hypointensity in the corresponding area on axial and coronal T2-weighted images (B and C).



ON-LINE FIG 3. Patient (5 years of age, 1.5T MR imaging) with isolated globus pallidus hyperintensity on T2-weighted images (A and B). Marked hypointense signals related to iron accumulation are seen in the GP with a marked medial-to-lateral gradient (A, arrowhead; B, arrow and asterisk), the anteromedial aspect of the subthalamic nucleus (C, arrow), and the substantia nigra (D, arrow) on SWI.

On-line Table: Clinical and MRI data of patients with PKAN

Pt.	Sex	Age at MRI (yr)	Duration (yr)	Clinical Features	Subtype	PANK2 Mutations	MRI Field Strength	T2WI Hyperintense Center	T2WI Surrounding Intensity	SWI Hypointensity
1	M	1.9	1.25	Developmental delay	Classic	c.1097delC, c.683T>C	1.5T	Linear streak	Isointense	None
2	F	5	2	Developmental delay, dystonia, retinopathy	Classic	c.1231G>A (homozygous)	1.5T	Round	Isointense	M>>L
3	F	5	3	Developmental delay, dystonia, choreoathetosis	Classic	c.1301C>A (homozygous)	1.5T	Round	Isointense	M>>L
4	F	5	3	Dystonia	Classic	c.1561G>A (heterozygous)	1.5T	Round	Isointense	M>>L
5	F	8	5	Developmental delay, dystonia	Classic	c.1257del, c.1319G>C	1.5T	Round	Hypointense	M>L
6	M	9	ND	Dystonia, ND	Classic	c.1231G>A, c.1253C>T	1.5T	Round	Hypointense	M>L
7	M	16	1	Dystonia	Atypical	c.999A>G, c.1270_1272del	1.5T	Round	Hypointense	M=L
8	F	3	1	Developmental delay, dystonia, spasticity, retinopathy	Classic	c.215_216insA (homozygous)	3T	Linear streak	Isointense	Subtle
9-I	F	1.8	1	Developmental delay, OCD, seizure	Classic	c.658G>T, c.981+3A>G	3T	Suspicious streak	Isointense	None
9-II	F	4	3.2	dystonia, spasticity, retinopathy	Classic		3T	Round	Isointense	M>>L
10	M	4	1.5	Developmental delay, dystonia	Classic	c.524delT, c.1319G>C	3T	Round	Isointense	M>>L
11	F	5	2	Developmental delay, dystonia, retinopathy	Classic	c.1154_1155insT, c.1319G>C	3T	Round	Isointense	M>>L
12	M	9	6	Developmental delay, dystonia	Classic	c.1273_1275delCTT, c.1676C>G	3T	Round	Hypointense	M>L
13	M	17	2	OCD, dystonia	Atypical	c.999A>G, c.1270_1272del	3T	Round	Hypointense	M=L
14	M	27	16	Learning disorders, dystonia, spasticity, dysarthria	Atypical	c.1231G>A, c.1255A>G	7T	Absent	Hypointense	M=L
15	F	29	15	Learning disorders, depression, dystonia, parkinsonism, dysarthria, dysphagia	Atypical	c.1231G>A, c.1255A>G	7T	Absent	Hypointense	M=L

Note:—Pt. indicates patient number; 9-I, initial MRI in Pt. 9; 9-II, follow-up MRI in Pt. 9; OCD, obsessive compulsive disorders; ND, not determined; M, hypointensity in the medial GP; L, hypointensity in the lateral GP; >>, markedly noticeable difference; >, less noticeable difference; =, no difference.