A 6-year-old boy from Maharashtra, India, presented with subacute-onset generalized dystonia after a febrile illness; his sister manifested insidious-onset multifocal dystonia (Video 1). T2-weighted MRI showed striatal hyperintensities (Figure 1). Clinical exome sequencing detected a novel homozygous variant (c.127 G > T; G43C) in the \textit{PRKRA} gene which encodes PACT, a stress-response protein. The variant was confirmed by Sanger sequencing (eFigure 1, links.lww.com/WNL/C111). DYT-PRKRA is a childhood-onset progressive dystonia.\textsuperscript{1} Some patients have exacerbation with fever and evidence of striatal degeneration in neuroimaging.\textsuperscript{2} PACT-dependent activation of protein kinase R (PKR) leads to apoptosis.\textsuperscript{2} Perhaps, the G43C PACT triggered abnormal activation of PKR and intensified neuronal apoptosis during febrile illness.

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References

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Suvorit Subhas Bhowmick, Sarbani Raha and Amita Bohora
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