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【 PICTURES IN CLINICAL MEDICINE 】

T2 Star-weighted MRI of Beta-propeller Protein-associated Neurodegeneration

Kengo Maeda[1,2], Yutaka Yamamoto[1], Katsuhisa Akiyama[2] and Takao Saotome[2]

Key words: SENDA, BPAN, T2 star, MRI, *WDR45*

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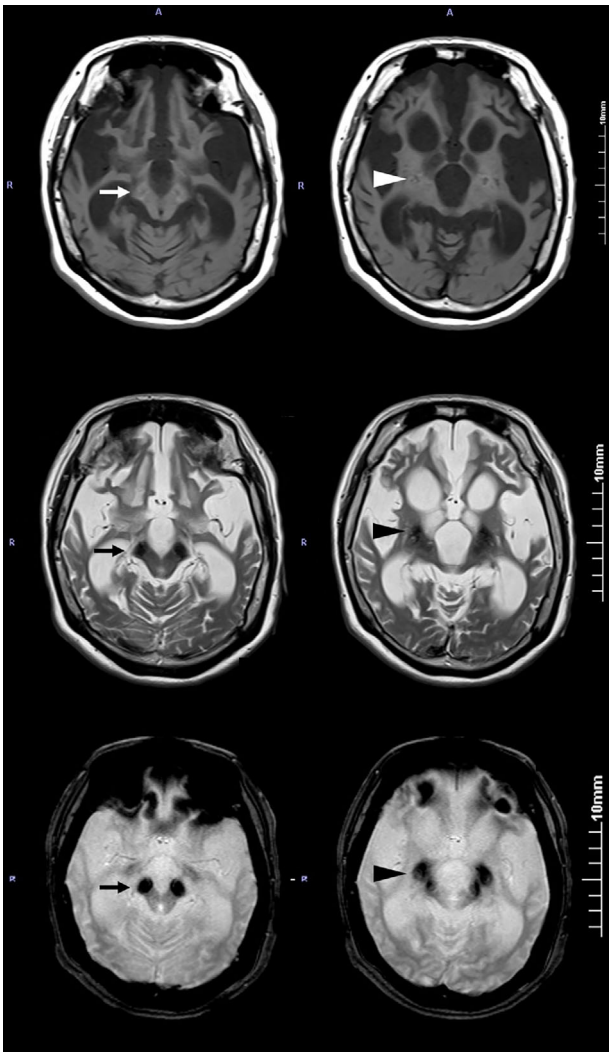

Picture.

The patient was a 41-year-old woman, who had been previously reported to have beta-propeller protein-associated neurodegeneration (BPAN) caused by autophagy gene *WDR45* mutation (1). She had been born normally, however, she developed convulsions nine months after birth. She could speak only a few words. She had dystonia, rigidity, and tremors. Her cognitive function started to decline at 19 years of age. She became bedridden by 25 years of age. BPAN is one type of neurodegeneration with brain iron accumulation. Iron deposition is found at the bilateral globus pallidus and substantia nigra. T1-weighted magnetic resonance imaging (MRI) showed hyperintensity with central hypointensity in these lesions (Picture, upper panels, arrow: substantia nigra, arrowhead: globus pallidus). The substantia nigra and globus pallidus are physiologically hypointense on T2-weigthed MRI. In BPAN, these lesions are more hypointense (Picture, middle panels). T2 star-weighted MRI (Picture, lower panels) shows these lesions much more clearly than usual T2-weighted MRI. T2 star-weighted MRI is superior in detecting iron deposition and therefore is useful for diagnosing BPAN as well as the occurrence of occult microbleeds.

The authors state that they have no Conflict of Interest (COI).

Reference

1. Saitsu H, Nishimura T, Muramatsu K, et al. *De novo* mutations in the autophagy gene *WDR45* cause static encephalopathy of childhood with neurodegeneration in adulthood. Nat Genet **45**: 445-450, 2013.

[1]Department of Neurology, National Hospital Organization Higashi-ohmi General Medical Center, Japan and [2]Department of Internal Medicine, Vories Memorial Hospital, Japan
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Correspondence to Dr. Kengo Maeda, maeda@vories.or.jp
