T2-hyperintensity of the Middle Cerebellar Peduncles in a Patient with SCA7

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A 41-year-old man with spinocerebellar ataxia type 7 (SCA7) noticed a staggering gait at 21 years of age and deterioration of vision at 25 years of age (1). His past medical history revealed no other diseases. A neurological examination conducted at 41 years of age revealed a bedridden status, mild cognitive decline, diminished vision, limitation of upward gaze with severe slow eye movements, scanning speech, limb and truncal ataxia and rigopsasticity in all extremities with generalized hyperreflexia and pathological reflexes. Brain MRI showed atrophy of the pons, cerebellum and middle cerebellar peduncles (MCP) in addition to T2-hyperintensity (Picture A, B) and fluid attenuated inversion recovery (FLAIR) hypointensity (Picture C, D) of the MCP.

Atrophy is generally more predominant in the pons than in the cerebellum, and T2-hyperintensity of the MCP is rare in patients with SCA7. This case demonstrates that T2-hyperintensity of the MCP, similar to that observed in patients with multiple system atrophy, fragile X-associated tremor/ataxia syndrome, SCA2 and SCA6 (2), can be observed in SCA7 patients in the advanced stage.

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References
