

T2-hyperintensity in the internal globus pallidus in Machado-Joseph disease

Hiperintensidade em T2 em globo pálido interno na doença de Machado-Joseph

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A 69-year-old male patient with Machado-Joseph disease (MJD) presented with a mild cerebellar ataxia, global areflexia, and nystagmus. Magnetic resonance imaging showed cerebellar atrophy; brainstem atrophy, mainly pontine, and a linear abnormal bilateral hyperintense along the medial aspect of the globus pallidus internus on T2-weighted sequence and fluid-attenuated inversion recovery (FLAIR) (– **Figure 1**). This radiographic finding implies degeneration of the lenticular fasciculus.¹ The hyperintensity may be associated with degeneration of the subthalamic fascicles or the nigrostriatal dopaminergic fibers.¹ This finding is not pathognomonic of MJD, although it has been described in subjects with other types of spinocerebellar ataxias and in healthy elderly people.²

Authors' Contributions

ATM: conceptualization, investigation, methodology, project administration, resources, supervision, writing – original draft, writing – review & editing; JEDA: conceptualization, writing – original draft, writing – review &

editing; LVA: conceptualization, writing – original draft, writing – review & editing; RNLFL, GLF, AMTN: conceptualization, writing – review & editing; HAGT: conceptualization, project administration, supervision, writing – review & editing.

Conflict of Interest

The authors have no conflict of interest to declare.

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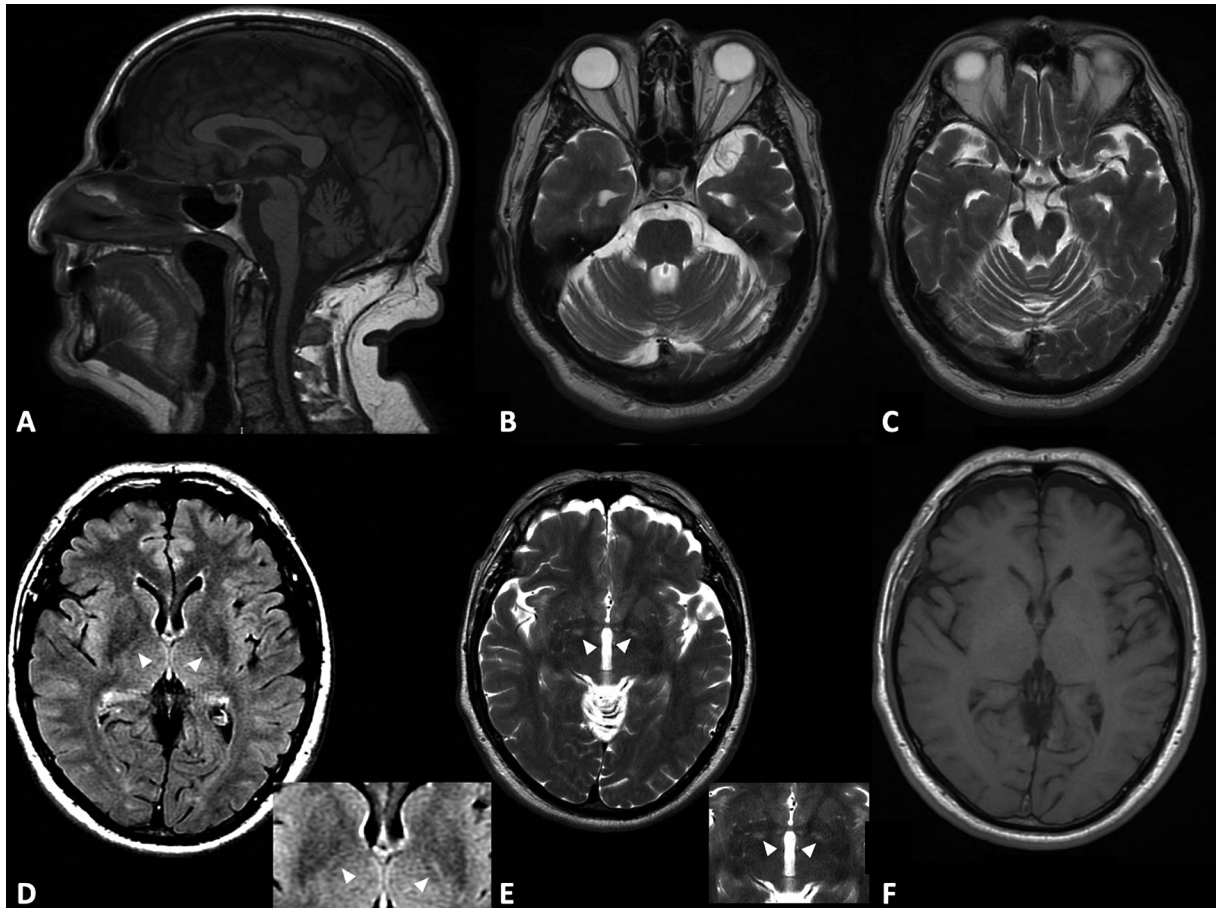


Figure 1 T2 and FLAIR hyperintensity in a patient with Machado-Joseph disease (SCA3). Cerebellar atrophy is shown in sagittal T1 (A), and axial T2 (B and C). Hyperintensity in the medial aspect of the internal globus pallidus is shown in the FLAIR (D) and T2 (E), but in T1 (F) there is no sign of abnormality.