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【 LETTERS TO THE EDITOR 】

Brain Magnetic Resonance Imaging Abnormalities in Patients with Hereditary Hemorrhagic Telangiectasia

Key words: hereditary hemorrhagic telangiectasia, manganese

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To the Editor I read with interest the article, "Hereditary Hemorrhagic Telangiectasia Induced Portosystemic Encephalopathy: A Case Report and Literature Review," by Kawabata et al. (1). While cerebral arteriovenous malformations (AVMs) (2) and basal ganglia signal abnormalities on magnetic resonance imaging (MRI) (2, 3) have been reported in patients with hereditary hemorrhagic telangiectasia (HHT), the article does not refer to these findings.

According to the paper by Parvinian et al. (2), the overall prevalence of cerebral AVMs in patients with HHT is 10%, and that of basal ganglia hyperintensity on T1-weighted MRI is 23-38%. Regarding the findings for basal ganglia hyperintensity, T1-weighted MRI shows bilateral symmetric hyperintensity at the globus pallidus and substantia nigra in such cases (3). This finding has been attributed to intracranial deposition of manganese in hepatic arteriovenous shunting (2, 3). The presence of basal ganglia hyperintensity demonstrated a significant association with increased age, the presence of hepatic AVMs, hepatic telangiectasias, nasal telangiectasias, increased peak cardiac output, hepatic failure, and elevated peak serum alkaline phosphatase levels and elevated total bilirubin levels (2). No significant association was noted with sex, the genetic mutation status, or parkinsonism (2). Basal ganglia hyperintensity may be a clue suggesting a diagnosis of hepatic encephalopathy, including portosystemic encephalopathy, in clinical practice (2-4).

The author states that he has no Conflict of Interest (COI).

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