Late-onset Portosystemic Encephalopathy in a Patient with Rendu-Osler-Weber Disease

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Picture 1.

Picture 2.

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A 78-year-old man with a history of Rendu-Osler-Weber disease (ROW) characterized by recurrent epistaxis and skin telangiectasia developed disturbed consciousness. The serum ammonia level was elevated at 224 $\mu\text{g/dL}$, and brain T1-weighted magnetic resonance imaging (MRI) showed hyperintensity of the globus pallidus (Picture 1). A contrast-enhanced abdominal CT image obtained in the early arterial phase disclosed early venous drainage (Picture 2, arrows) due to arteriosystemic shunting, while a right anterior oblique projection of three-dimensional abdominal CT angiography demonstrated a large hepatic artery aneurysm (Picture 2, star) with small areas of telangiectasia (Picture 2, arrowheads). The patient was therefore diagnosed with portosystemic encephalopathy (PE), likely resulting from increased intrahepatic portosystemic shunting (PSS) associated with aging. The intravenous administration of branched

amino acids subsequently ameliorated his symptoms. The occurrence of PE in patients with ROW is rare (1). Symmetrical hyperintensity of the globus pallidus on T1-weighted images indicates the accumulation of manganese due to PSS (2). Therefore, MRI findings may be predictors of the onset of PE in ROW patients.

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References

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