A 75-year-old man was admitted to our hospital because of parkinsonism. Past history included recurrent nasal hemorrhage since around 12 years of age. His father also had recurrent nasal hemorrhage. On admission, he had telangiectasia on the forehead. The routine serum chemistry and complete blood count were unremarkable including liver function tests and hepatitis screening tests. T1-weighted image (T1WI) of brain MRI revealed bilateral hyperintensities involving the substantia nigra and the globus pallidus (Fig. 1). Arterial-phase image of abdominal enhanced CT revealed multiple abnormal vessels throughout the liver. Early filling of right hepatic vein suggested presence of arteriovenous shunts (Fig. 2). The serum ammonia level was 126 μg/dl (normal 12–66 μg/dl) and the whole blood manganese level was 4.1 μg/dl (normal 0.8–2.5 μg/dl). He was diagnosed as hereditary hemorrhagic telangiectasia (HTT). There are only two reports which include the brain MRI of HTT with hepatic involvement (1, 2). We considered the hyperintensities on T1WI to be caused by deposition of manganese due to the intrahepatic arteriovenous shunts.

References

Figure 2. Arterial-phase image of abdominal enhanced CT disclosed multiple abnormal vessels throughout the liver. Early filling of right hepatic vein suggested presence of arteriovenous shunts. 1: right hepatic vein, 2: inferior vena cava.