cortex, which can be accompanied by myoclonus and progressive dementia[1,3].

MRI studies have come to play an ever more important role in the evaluation of patients with neurological diseases[4–7]. On MRI, the sporadic and inherited forms of CJD usually present areas of high signal intensity in T2-weighted and FLAIR sequences, with restricted diffusion, in the cerebral cortex and the basal ganglia, especially the striatum, in a focal or diffuse, symmetric or asymmetric form, sparing the region around the rolandic cortex and the thalami[3]. Classic signs such as the pulvinar sign and the "hockey stick" sign are typical of the variant form and are characterized respectively by hyperintense signals in T2-weighted and FLAIR sequences of the posterior and posteromedial thalami[8,9].

In HvCJD, there is invariably involvement of the parietooccipital cortex, including the primary visual cortex, characterized on MRI by hyperintense signals in T2-weighted and FLAIR sequences, together with restricted diffusion, typically with preservation of the subcortical white matter and of the basal ganglia. It is noteworthy that restricted diffusion can precede the clinical manifestations of CJD[3].

In HvCJD, the electroencephalogram typically shows acute, periodic triphasic waves, predominantly in the posterior areas[10]. Analysis of the cerebrospinal fluid can reveal elevated 14-3-3 protein levels[3]. Histopathological analysis is the gold standard diagnostic method, showing marked neuronal loss, spongiform changes, intense astrogliosis and immunoreactivity to the abnormal pathogenic isoform of the prion protein[11]. The prognosis is bleak, and death usually occurs within one year[2,9].

It is important to make the differential diagnosis of HvCJD. The main differential diagnoses are frontotemporal dementia, status epilepticus, hypoxic-ischemic encephalopathy, severe hypoglycemia, immune-mediated autoimmune encephalopathy, posterior cortical atrophy, and hyperammonemia[3]. Although rare, HvCJD should be borne in mind in the differential diagnosis of visuospatial deficits, especially when MRI shows areas of high signal intensity in T2-weighted and FLAIR sequences, together with restricted diffusion, in the cortical region of the occipital lobes.

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Radiological findings in the liver of a patient with Rendu-Osler-Weber syndrome

Dear Editor,

A 57-year-old male patient with Rendu-Osler-Weber syndrome presented to the emergency department with a 24-h history of lumbar pain. A computed tomography scan of the abdomen showed liver alterations typical of the syndrome (telangiectasias, shunts, and arteriovenous malformations), which is also known as hereditary hemorrhagic telangiectasia. The examination showed opacification of the hepatic veins in the early arterial phase—a consequence of the arteriovenous shunts (Figure 1A). We observed heterogeneous opacification of the portal vein during the portal phase, with more pronounced enhancement in the intrahepatic branches—a result of portal venous shunt—as well as numerous prominent vessels near the hepatic hilum, corresponding to an arteriovenous malformation (Figure 1B). We also observed a confluent vascular mass, measuring 1.4 cm, located in segment II (Figure 1C). In addition, there were extensive areas of altered perfusion in the hepatic parenchyma, in a mosaic pattern, as well as increased caliber of the hepatic artery at its emergence from the superior mesenteric artery, which was also ectatic (Figure 1D).

Imaging exams have played an important role in the study of liver diseases[1–5]. Hereditary hemorrhagic telangiectasia is a dominant autosomal disease with a prevalence of 10–20 cases per 100,000 population[6]. It is a rare systemic fibrovascular dysplasia that makes the walls of blood vessels more vulnerable to trauma and spontaneous ruptures[7]. It affects multiple organs and systems, being characterized mainly by the presence of telangiectasias or vascular shunts in the liver, lungs, kidneys, central nervous system, or skin[8,9]. In adults, it typically manifests as recurrent epistaxis, mucocutaneous telangiectasias, digestive tract hemorrhage, and hemoptysis[9,10]. Telangiectasias appear gradually, the most common sites being the lips, tongue, palate, fingers, and face. The diagnosis of the syndrome is based on the presence of three of the four diagnostic criteria[8]: mucocutaneous telangiectasias, recurrent spontaneous epistaxis, visceral arteriovenous malformations, and a positive family history.

In Rendu-Osler-Weber syndrome, the liver is the organ most often affected, hepatic involvement being reported in 74% of cases. Hepatic involvement is typically diagnosed 10–20 years after the

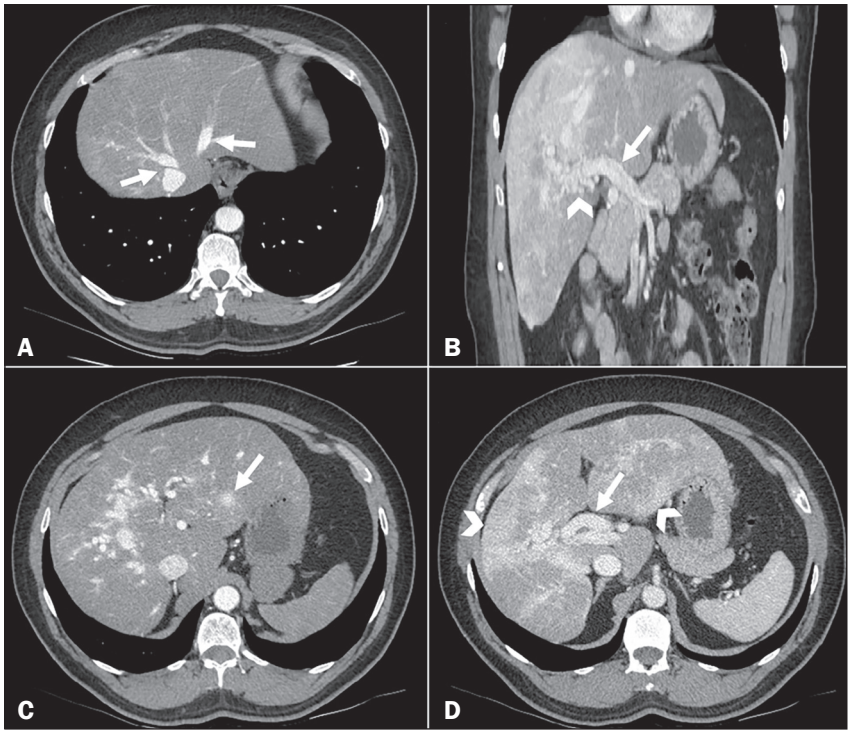

Figure 1. Computed tomography scan of the abdomen in axial slices (**A**, **C**, and **D**) and in a coronal slice (**B**). **A:** Note the opacification of the hepatic veins in the early arterial phase (arrows). **B:** Heterogeneous opacification of the portal vein during the portal phase (arrow), accompanied by numerous ectatic vascular structures surrounding the hepatic hilum, representing an arteriovenous malformation (arrowhead). **C:** Confluent vascular mass, measuring 1.4 cm, in segment II (arrow). **D:** Extensive areas of altered perfusion in the hepatic parenchyma, in a mosaic pattern (arrow heads), together with increased caliber of the hepatic artery (arrow).

appearance of the first telangiectasia. In 65% of cases, the liver shows heterogeneous enhancement in the arterial phase, with a mosaic perfusion pattern, which is characterized by areas of altered perfusion, indicative of arterioportal shunts. Hepatic telangiectasias, found in 63% of cases, can be focal or diffuse and are described as rounded lesions, smaller than 10 mm, that are hypervascular in the arterial phase and, in the portal phase, often exhibit density equal to that of the hepatic parenchyma. When such a lesion is larger than 10 mm, as it is in 25% of patients, it is referred to as a confluent vascular mass, comprising areas of grouped multiple telangiectasias or visible shunts[10,11].

Vascular shunts, which are seen in 65% of cases of Rendu-Osler-Weber syndrome, appear in one of three forms[11]: arteriovenous (from the hepatic artery to the hepatic vein); arterioportal (from the hepatic artery to the portal vein); and portal-venous (from the portal vein to the hepatic vein). Vascular shunts are associated with complications such as congestive heart failure and portal hypertension[12]. In some cases, there are also hepatic vascular malformations, which can cause a right-to-left shunt, resulting in varying degrees of pulmonary hypertension, heart failure, and hepatic encephalopathy[8].

The treatment of Rendu-Osler-Weber syndrome includes measures to control epistaxis, as well as surgical removal, radiotherapy, and embolization of vascular malformations, with an emphasis on endovascular treatment[8].

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