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 pres-
ent 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 thalamus(3). Classic signs such as the pul-
vinar 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 pos-
teromedial thalamus(8,9).

In HvCJD, there is invariably involvement of the parieto-
occipital cortex, including the primary visual cortex, charac-
terized on MRI by hyperintense signals in T2-weighted and FLAIR
sequences, together with restricted diffusion, typically with pres-
ervation 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(11). Histopathological analysis is the gold standard
diagnostic method, showing marked neuronal loss, spongiform
changes, intense astrogliosis and immunoreactivity to the abnor-
mal pathogenic isof orm 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 hy-
poglycemia, immune-mediated autoimmune encephalopathy,
posterior cortical atrophy, and hyperammonemia(3). Although
rare, HvCJD should be borne in mind in the differential diag-
nosis 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 syn-
drome presented to the emergency department with a 24-h his-
tory of lumbar pain. A computed tomography scan of the abdo-
men showed liver alterations typical of the syndrome (telangiec-
tasias, shunts, and arteriovenous malformations), which is also
known as hereditary hemorrhagic telangiectasia. The examina-
tion showed opacification of the hepatic veins in the early arte-
rial 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 exten-
sive 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
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 tel-
angiectasias or vascular shunts in the liver, lungs, kidneys, cen-
tral 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); mucocuta-
neous 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

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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 arterioporal 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); arterioporal (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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