CASE REPORT

Hepatocerebral degeneration — a case report

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Abstract

Acquired hepatocerebral degeneration is a subtype of chronic recurrent hepatic encephalopathy and is characterised by movement disorders and increased signal intensity in the basal ganglia on T1-weighted MRI images.

Case report

A 49-year-old female patient was admitted to a local hospital casualty department after having had a grandmal seizure. Clinically she was found to have hepatomegaly, anaemia and stigmata of chronic alcohol abuse. Biochemical analysis showed deranged liver enzymes and haematological analysis showed anaemia but a normal clotting profile.

Radiological investigations included unenhanced MRI and CT scans of the brain, and an abdominal ultrasound.

On the MRI scan the T1-weighted scans showed high signal intensity in the lenticular nuclei bilaterally and symmetrically, primarily within the globus pallidus (Fig. 1), as well as in the midbrain with sparing of the red nuclei (Fig. 2). T2-weighted scans showed subtle hypointensity in the same areas (Fig. 3). An unenhanced CT scan of the brain revealed no basal ganglia calcification (Fig. 4).

Ultrasound examination of the abdomen showed a diffuse increase in echogenicity of the liver parenchyma indicating fatty infiltration and mild splenomegaly. There was no evidence of portal hypertension.

The patient had no seizures while
Table I. Causes of increased signal intensity in the basal ganglia on T1-weighted images

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<thead>
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<th>Cause</th>
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<tr>
<td>Calcification</td>
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<tr>
<td>Neurofibromatosis</td>
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<tr>
<td>Parenteral nutrition</td>
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<tr>
<td>Hepatocerebral degeneration</td>
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in hospital and was discharged a week later.

**Discussion**

The causes for increased T1-weighted signal intensity in the basal ganglia on MRI scans of the brain are shown in Table I.

In our patient, calcification as a cause was ruled out on the unenhanced CT scan of the brain and there was no history of neurofibromatosis or previous parenteral nutrition.

Given the history of alcohol abuse with the related biochemical and imaging findings the diagnosis in this case was probably hepatocerebral degeneration.

Acquired hepatocerebral degeneration is an irreversible neuro-degenerative syndrome occurring in association with chronic liver disease such as alcoholic cirrhosis and chronic hepatitis as well as with portosystemic shunts. Positive imaging findings correlate strongly with plasma ammonia levels or brain magnesium levels. MR spectroscopic studies have shown elevated glutamine levels and reduced choline metabolite and myo-inositol levels in patients with chronic hepatic encephalopathy. Due to financial constraints, these tests were not performed in our patient. In addition to the areas in the brain described above, the anterior pituitary, caudate nucleus, quadrigeminal plate and subthalamic regions may also show signal intensity alterations.

Hepatocerebral degeneration is a clinical syndrome and a subtype of chronic recurrent hepatic encephalopathy. In some cases the development of the cerebral dysfunction is rapid, developing over a matter of days resulting in altered consciousness from confusion through stupor, coma and death. In other cases the clinical picture is largely progressive and is characterised by movement disorders, dysarthria, tremor and ataxia. Pathologically, the brain shows enlargement and hyperplasia of astrocytes as well as neuronal abnormalities in the cerebral and cerebellar cortices, basal ganglia and diencephalic nuclei. Necrosis with cavitation, gliosis and myelin breakdown occur at these sites.

Medical treatment is often disappointing, but in selected cases liver transplantation may be curative. The basal ganglia hyperintensity may resolve following liver transplantation.

**References**