MRI and SRM findings in haemolytic uraemic syndrome with central nervous system involvement

Published on 09.09.2010

DOI: 10.1594/EURORAD/CASE.8736
ISSN: 1563-4086
Section: Paediatric radiology
Case Type: Clinical Cases
Patient: 10 months, male

Clinical History:

A 10-month-old boy presented with a 4 day history of diarrhoea and acute dehydration. Two days later he was admitted in our hospital with seizures and progressive alteration of consciousness. Cerebral MRI and MRS were performed.

Imaging Findings:

A 10-month-old boy was admitted to the paediatric intensive care unit with a 4 day history of diarrhoea and acute dehydration. Two days later he presented with seizures and alteration of consciousness. MRI showed multiple ischaemic and haemorrhagic lesions in the frontal, parietal and occipital lobes, and basal ganglia, with symmetrical distribution. The right cerebellar peduncle involvement was discreet. Both grey and white matter was involved. Diffusion weighted images demonstrated reduced diffusion with low ADC, and MRS showed lipid peak and NAA disappearance, which was caused by necrosis and cellular death.

Further investigation revealed renal failure and thrombopenia, and microbiology confirmed E. coli gastrointestinal infection.

A diagnosis of haemolytic uraemic syndrome (HUS) induced by E. coli gastroenteritis was made, and encephalopathy secondary to verotoxin-producing coli was the final diagnosis.

Nine months later, the patient presented with muscular hypotonia and psychomotor delay. MRI showed cortical necrosis and diffuse brain atrophy, with regression of basal ganglia and cerebellar peduncle abnormalities.

Discussion:

HUS is a multisystem disease, which is characterised by uraemia, thrombocytopenia and haemolytic anaemia. It is the most common cause of acute renal failure in children, and is primarily seen in those between 1 and 4 years of age. The disease typically starts after a gastrointestinal infection with diarrhoea [1].

Histological studies have shown that verotoxin produced by the bacteria binds to the endothelium of small organ vessels and leads to thrombotic vessel occlusion. Besides the kidneys, the central nervous system (CNS) is involved in 20–50% of cases.

The pathogenesis of CNS involvement is debated. The encephalopathy is secondary to metabolic changes (hyponatraemia, hydration disorders) and hypertension or the neurological damage is directly induced by the verotoxin affecting the endothelium of small brain vessels leading to infarction and bleeding [2,3].

In patients with HUS and clinical signs of major neurological complications (seizures, alteration of consciousness,
hemiparesis, visual disturbances and brainstem symptoms), imaging studies of the brain are performed to document the severity and nature of pathological changes. CT is able to document gross infarction, bleeding and generalised oedema, whereas subtle changes are much better demonstrated with MRI. There are several reports of the MRI findings in patients with HUS and neurological complications. Although various imaging findings have been described, most of the studies found basal ganglia lesions to be characteristic. Other findings, including territorial infarction or diffuse white-matter changes similar to posterior leucoencephalopathy, reflect complications rather than specific changes of the disease [1,4].

The abnormalities consist of oedema and sometimes subacute haemorrhagic infarcts with increased signal in T1 and T2 weighted images [2,5].

Although involvement of the basal ganglia is not specific for HUS and is also seen in various other conditions, including severe hypoxia, intoxication and infectious diseases, it supports the theory of a direct or receptor-mediated verotoxin-induced injury [3,4].

Our case confirms previous investigations regarding the distribution of pathological changes. We found signal intensity changes in the dorsolateral portion of the lentiform nucleus, which is characteristic finding.

These signal changes extend into the surrounding white-matter tracts of the internal and external capsule, and the thalamus.

The present case demonstrates that reduced apparent diffusion coefficient values on DWI could be the imaging findings that most reliably identify irreversible brain lesions, adding a parameter with early prognostic value [5].

**Differential Diagnosis List**: Haemolytic uraemic syndrome with brain involvement

**Final Diagnosis**: Haemolytic uraemic syndrome with brain involvement

**References**:


Figure 1

Description: Sagittal and axial T1 weighted images showing increased signal intensity of haemorrhagic lesions. Origin:
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Description: MRI axial T2 weighted images demonstrate high signal in frontal, parietal and occipital lobes, in the central semiovale, the right cerebellar peduncle, thalami caudate nuclei and lentiform nuclei. Origin:
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Figure 3

Description: Axial T2* weighted images demonstrate well cortical and sub-cortical haemorrhage. Origin:
Figure 4

Description: Single voxel in the ischaemic left occipital lesion. Origin:
Figure 5

Description: Diffusion images showing high signal intensity of lesions. ADC map showed low ADC caused by cytotoxic oedema. Origin:

b

Description: Diffusion images showing high signal intensity of lesions. ADC map showed low ADC caused by cytotoxic oedema. Origin:
Description: Single voxel MRS at a short TE (35ms) in (a) and long time (144) in (b). There is a very high lipid peak (arrow) and decreased NAA (arrowhead). Origin:
Description: Single voxel MRS at a short TE (35ms) in (a) and long time (144) in (b). There is a very high lipid peak (arrow) and decreased NAA (arrowhead). Origin:
**Description:** Axial T2 Flair images showing
- Regression of thalamus, basal ganglia and peduncle abnormalities
- Extensive cortical necrosis with brain atrophy with enlarged supratentorial ventricles and widening of subarachnoid spaces

**Origin:**

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Figure 7

a
Description: Magnetic resonance imaging–angiography is normal. Origin: