Diffusion MRI features of acute encephalopathy due to stopping steroid medication abruptly in congenital adrenal hyperplasia

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Congenital adrenal hyperplasia (CAH) is characterized by adrenal steroid biosynthesis defect. Steroid replacement therapy should be performed regularly in these patients. Adrenal crisis may be present in acute stress due to increased cortisol requirements or in steroid deficiency due to stopping steroid medication abruptly. In patients with acute adrenal insufficiency, severe hypotension or hypovolemic shock occurs typically. Acute encephalopathy can be seen due to hypoxia, hypervolemia, or hypoglycemia. Diffusion restriction can be seen in cortical-subcortical regions of frontal and parieto-occipital lobes and in splenium of corpus callosum. In CAH patients with neurologic symptoms, Diffusion weighted images (DWI) is very important in the diagnosis and follow-up of acute encephalopathy.

Introduction

Congenital adrenal hyperplasia (CAH) is characterized by adrenal steroid biosynthesis defect. Steroid replacement therapy should be performed regularly in these patients. Encephalopathy in CAH can depend on many different etiologic factors such as hypoxia, hypovolemia, hypoglycemia, infection, and hormonal causes. [sup][1] Acute encephalopathy is commonly associated with hypoxia in newborn infants and infection in childhood. [sup][2]

To our knowledge, there are a few reports about brain magnetic resonance imaging (MRI) features of adrenal crisis in children with CAH in the literature. We report diffusion MRI findings of a rare cause of acute encephalopathy, due to stopping steroid medication abruptly in a child with CAH.

Case Report

A 3-year-old boy was admitted to our emergency department because of convulsions and unconsciousness. He had been diagnosed as 21-hydroxylase deficiency at birth. He had been taking oral steroid (hydrocortisone) replacement therapy since then. His parents had stopped steroid therapy without the knowledge of the physician 3 days before the symptoms started. They were admitted to our hospital 6 hours after the onset of the symptoms. He was in coma and had seizures on admission. Laboratory tests were normal except for hypoglycemia. His plasma glucose level was 20 mg/dl and white blood cell (WBC) count was normal. No evidence of serum electrolyte abnormalities was detected. Specific infectious agents were not identified by comprehensive studies. Intravenous hydrocortisone treatment was initiated. Electroencephalography showed focal epileptic activity. Routine brain MRI was performed. Diffusion weighted images (DWI) and susceptibility weighted images (SWI) were also obtained in order to detect ischemia and hemorrhage. T2-weighted

and fluid attenuated inversion recovery (FLAIR) images revealed increased signal intensities in the bilateral frontal and parietooccippital cortical-subcortical regions and a hyperintense lesion in the splenium of the corpus callosum [Figure 1]. Effacement of sulcal spaces was detected compatible with cerebral edema. Hyperintense cortical-subcortical signals and the lesion in the corpus callosum were hyperintense on DWI with reduced apparent diffusion coefficient (ADC) maps [Figure 2]. There were no signal changes suggesting microhemorrhages on SWI. MRI findings were consistent with cytotoxic cerebral edema in bilateral frontal and parieto-occiptal cortical-subcortical regions due to hypoxia and hypoglisemia. We obtained follow-up brain MRI on fifth, twelfth, and twentieth days once the treatment has started. DWI and FLAIR images on day 20 indicated partial resolution of the high intensity lesions. Hyperintense signal changes gradually decreased and completely disappeared in frontal lobes. However, ventricular dilatation, mild cerebral cortical atrophy, T2 and FLAIR hyperintense signal changes in bilateral occipital lobes were detected on the last MRI [Figure 3]. SWI was normal, and DWI showed restricted diffusion in bilateral occipital lobes. The patient exhibited epilepsy as neurological sequelae.{Figure 1}{Figure 2}{Figure 3}

Discussion

CAH is a group of autosomal recessive diseases characterized by adrenal steroid biosynthesis defect. The most common enzyme insufficiency that causes CAH is 21-hydroxylase deficiency. [sup][3] Typical presentation is ambiguous genitalia in females and/or salt-losing crisis in both genders at birth. The diagnosis is conﬁrmed by elevated levels of serum 17-OH-progesterone and testosterone. Hyponatremia, hyperkalemia, and hypoglycemia can also be seen. Steroid replacement therapy should be performed regularly under close medical supervision in CAH. Adrenal crisis may be present in acute stress due to increased cortisol requirements or in steroid deficiency due to stopping steroid medication abruptly. In patients with acute adrenal insufficiency, severe hypotension or hypovolemic shock occurs typically.

Acute encephalopathies are brain disorders caused by many different causes. Frequently acute encephalopathy caused by oxygen deficiency, infectious agents, toxic drugs, or metabolic disorders. Hypovolemia and hypoglycemia can also lead to encephalopathy. Acute encephalopathy is often characterized with neurological abnormalities. It can be reversible or irreversible depending on the reason of the encephalopathy, onset time of therapy, and the location of the brain involvement. [sup][4]

Okumura et al . categorized acute encephalopathy into two groups as diffuse and central-sparing lesions according to the distribution of brain lesions. [sup][4] Cerebral cortex is susceptible to hypoxia. Global ischemia can cause bilateral and symmetric cerebral infarcts in the border zones between major arterial territories. [sup][5] Hypoxic-ischemic encephalopathy is accompanied by cytotoxic cerebral edema. White matter can be involved rarely. In our case, the patient was in coma due to hypovolemic shock, hypoglycemia, and hypoxia, which were caused by steroid deficiency. MRI showed reduced diffusion in the bilateral frontal and parieto-occipital cortical-subcortical watershed border zones and effacement of sulcal spaces compatible with cytotoxic cerebral edema. There was also a transient lesion in the

splenium of corpus callosum. Various reasons such as mild encephalopathy with a reversible splenial lesion (MERS), corticoid treatment, or hypoglycemia can cause transient lesions in the splenium. [sup][6],[7]

Okumura et al . reported that clinical symptoms, laboratory data, and outcomes were markedly different between patients who had diffuse involvement and central sparing lesions. [sup][4] They reported that in the presence of central sparing lesions, the areas around the Sylvian fissure did not show diffusion restriction and the lateral parts of the occipital lobes are affected more heavily than the medial parts.

In contrast to patients with central sparing lesions, the outcome for patients with diffuse lesions was very poor. [sup][4] In our patient, areas around the Sylvian fissure were spared and there was more marked restricted diffusion in the lateral parts of the bilateral occipital lobes consistent with central sparing lesions. DWI obtained on the twentieth day demonstrated partial resolution of the lesions and patient showed epilepsy as neurological sequelae.

Laminar cortical necrosis of a patient following an adrenal crisis was reported by Saito Y et al . [sup][8] They described MRI findings in acute and chronic periods. In chronic period, progressive ventricular dilatation and linear high signal intensities on T1-weighted and FLAIR images were detected in the cerebral cortex with neurological sequelae. We did not detect laminar cortical necrosis in the first 20 days, however it may be seen in chronic follow-up images.

As a conclusion, acute adrenal insufficiency, which is characterized by hypotension or hypovolemic shock, may be present in acute stress due to increased corticoid requirements or in steroid deficiency due to stopping steroid medication abruptly. Acute encephalopathy can be seen due to hypoxia, hypervolemia, or hypoglycemia. Diffusion restriction can be seen in cortical-subcortical regions of frontal and parieto-occipital lobes and in splenium of corpus callosum. In CAH patients with neurologic symptoms, DWI is very important in the diagnosis and follow-up of acute encephalopathy.

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