MR imaging findings in alcoholic and non-alcoholic acute Wernicke's encephalopathy

Poster No.: C-1117
Congress: ECR 2013
Type: Educational Exhibit
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Keywords: Neuroradiology brain, MR, MR-Diffusion/Perfusion, Diagnostic procedure, Education, Acute
DOI: 10.1594/ecr2013/C-1117

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Learning objectives

Wernicke's encephalopathy (WE) is an uncommon but severe neurological syndrome, caused by thiamine (vitamin B1) deficiency. It is characterized by the sudden onset of altered consciousness, ophthalmoplegia and ataxia; however this classic clinical triad is present in only a minority of patients, making this condition often misdiagnosed and, consequently, life-threatening. Its prognosis depends on prompt early intravenous administration of thiamine. MR imaging is an essential tool to get the right diagnosis, especially when clinical presentation is incomplete. The aim of this exhibit is to describe the spectrum of MR imaging findings in alcoholic and nonalcoholic acute Wernicke's encephalopathy based on a series of 6 patients.
Background

WERNICKE'S ENCEPHALOPATHY

Etiopathogenesis

Role of thiamine (vitamin B1)

Thiamine is a water-soluble vitamin involved in the maintenance of membrane integrity and osmotic gradients across cell membranes. It is stored in body tissues, especially in the liver, predominantly as thiamine diphosphate (TDP). TDP also plays an important role in the conversion of glucose into energy and, in particular, acts as an essential coenzyme in the pentose phosphate pathway and in the Krebs cycle.

A healthy adult requires approximately 1-2 mg of thiamine daily, depending on the carbohydrate intake. Body's reserves of thiamine are only 30-50 mg so any malnutrition condition lasting more than 3-4 weeks can cause complete depletion of the vitamin's stores. In case of thiamine deficiency, intracellular TDP is depleted and the cycles of Krebs and of pentose phosphates cannot metabolise glucose, causing first intracellular then extracellular glutamate accumulation with consequent failure to maintain the normal osmotic gradient across cell membranes. Disregulation of these thiamine dependent metabolic pathways, leading to simultaneous cytotoxic and vasogenic oedema in glial cells and neurons, represents the biochemical mechanism responsible for the signs and symptoms of WE.

Alcoholic Wernicke's encephalopathy

The most common cause of thiamine deficiency is chronic alcohol abuse. Alcoholism is not directly responsible for vitamin B1 deficiency; its effects are related to the complications of liver cirrhosis such as problems to the gastrointestinal tract with low mucosal absorption rate and consequent malnourishment.

Nonalcoholic Wernicke's encephalopathy

Apart from alcohol, a lot of other conditions causing malnutrition and decreased thiamine absorption such as gastrointestinal surgical procedures (including gastric bypass surgery, gastrojejunostomy, gastrectomy, colectomy), hyperemesis gravidarum, chemical therapy, AIDS, anorexia nervosa, fasting, starvation, hemodialysis, uremia, pancreatitis and wrong formula feeding have been reported as predisposing factors.
Pathologic findings

In acute WE the most evident alterations are described at the level of the structures around the third ventricle such as the medial thalami, the periaqueductal grey matter, the mammillary bodies and the tectal plate of the midbrain. All these areas are considered typical sites of involvement. Because of their high oxidative metabolism, it has been suggested that these regions are particularly sensitive to thiamine deficiency.

The dorsal medulla, the vermis and the paravermian regions of the cerebellum, the corpus callosum and the fronto-parietal cortex are recognized as less commonly involved areas. Involvement of these less typical regions is more frequent in nonalcoholic WE.

Pathologic findings comprise demyelination, alterations of the blood-brain barrier (BBB) with vasogenic oedema, necrosis, vascular proliferation and astroglial and microglial proliferation with petechial hemorrhages.

Clinical manifestations

Clinically WE is characterized by the sudden onset of ocular disorders (nystagmus, diplopia, ophthalmoplegia, decreased bilateral visual acuity), equilibrium disorders (vestibular paralysis, ataxia) and cognitive impairment (apathy, spatial disorientation, altered consciousness).

However, this classical triad can be seen in just one-third of patients. For this reason, it has been proposed that a suspicion of Wernicke's encephalopathy should be based on two of the following four conditions: malnutrition, oculomotor abnormalities, cerebellar dysfunction and an altered mental state.

MR imaging findings

Typical and atypical findings

On MR imaging, the pathologic alterations described above are usually seen as bilateral and symmetrical T2w and FLAIR hyperintensities in the thalami, mamillary bodies, tectal plate and periaqueductal area.

Signal intensity alterations in the cerebellum, cerebellar vermis, cranial nerve nuclei, pons, splenium and cerebral cortex represent atypical MRI findings. They are always found in association with the classical neuroradiological presentation. These findings derive from the alteration of osmotic gradients across cell membranes, with simultaneous cytotoxic and vasogenic oedema.
The absence of MR signal alterations, however, does not exclude the diagnosis of WE. Gadolinium administration can be a useful tool to indentify WE cases with negative MRI scan. Contrast-enhanced T1-weighted images point out areas with disrupted blood-brain barrier. Strong enhancement of the mammillary bodies, for instance, can be the only sign of the disease and is more frequent in chronic alcoholics.

**Role of DWI**

In WE the role of diffusion-weighted imaging (DWI) with determination of the apparent diffusion coefficient (ADC) is not yet well defined. The lesions may show hyperintensity on DWI images and reduced, normal, or increased ADC value; moreover, areas with distinct diffusion characteristics can be seen in several different structures simultaneously. This heterogeneity may result from disease severity, acuteness and timing of imaging.

According to the literature, high signal changes on diffusion-weighted images (DWI) with either decreased apparent diffusion coefficient (ADC) represent cytotoxic oedema of the neurons and glial cells whereas areas with normal or increased ADC values indicate the presence of vasogenic oedema.
Imaging findings OR Procedure details

In all patients (three chronic alcohol abusers, a gravida 1 with hyperemesis, a leukemic patient under chemical therapy, a post-bariatric surgery patient) brain MRI (1.5 T scanner Philips Intera) revealed FLAIR and T2-weighted hyperintense symmetric areas at the level of periaqueductal gray matter [Fig. 1] and medial portions of the thalami [Fig. 2-3]. In nonalcoholic patients similar signal-intensity alterations were also observed at the level of the tectal plate (gravida) [Fig. 4], bilateral fronto-parietal cortex (leukemic patient) [Fig. 5] and pons (post-bariatric surgery patient) [Fig. 6]. All the lesions showed high signals on diffusion-weighted images (DWI) with either increased ADC [Fig. 7-8].
Fig. 1: 64-year-old man with a 15-year history of alcohol abuse: axial T2-weighted MR image showing hyperintense signal alteration at the level of periaqueductal gray matter.

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Fig. 2: 43-year-old man with a 10-year history of alcohol abuse: axial T2-weighted MR images showing bilateral and symmetric hyperintense signal alteration at the level of the medial portion of the thalami

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Fig. 3: 50-year-old man with a 7-year history of alcohol abuse: coronal FLAIR MR images showing bilateral and symmetric hyperintense signal alteration at the level of the mamillary bodies (a) and the medial portion of the thalami (b).

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Fig. 4: 33-year-old woman, primipara, in her 16th week of gestation: coronal FLAIR (a) and sagittal T2-weighted (b) MR images showing bilateral and symmetric hyperintense signal alteration at the level of the medial portion of the thalami and of the tectal plate.

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Fig. 5: 45-year-old man, affected by acute lymphoblastic leukemia, under chemical therapy: axial T2-weighted (a) and coronal FLAIR (b) MR images showing bilateral hyperintense signal alteration at the level of the fronto-parietal cortex.

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**Fig. 6:** 52-year-old man who had undergone bariatric surgery: axial T2-weighted MR image showing hyperintense signal alteration at the level of the right portion of the pons.

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Fig. 7: 33-year-old woman, primipara, in her 16th week of gestation: axial diffusion-weighted (b)MR image with ADC map (a) showing bilateral and symmetric high signal with increased ADC at the level of the medial portion of the thalami.

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**Fig. 8:** 45-year-old man, affected by acute lymphoblastic leukemia, under chemical therapy: axial diffusion-weighted (b)MR image with ADC map (a) showing bilateral high signal with increased ADC at the level of the fronto-parietal cortex.

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Conclusion

Our experience conforms with the most recent literature results that consider atypical (signal-intensity alterations in cerebral cortex and pons in the described cases) MR imaging findings more frequent in nonalcoholic patients.
References


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