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Answers to Medical Quiz

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Ans. Wernicke's Encephalopathy. The MRI study shows symmetrical hyperintense signal in the medial thalami and periaqueductal gray matter as well as in floor of fourth ventricle. This pattern is considered typical for Wernicke's encephalopathy [WE].

DISCUSSION

Wernicke's encephalopathy is an acute life threatening neurological disorder caused by deficiency of vitamin B1 (Thiamine). The disease is found mostly in malnourished alcoholics but also occurs in other conditions such as hyperemesis gravidarum, systemic neoplasms, gastrointestinal surgery, chronic dialysis, prolonged intravenous feeding, anorexia nervosa and acquired immunodeficiency syndrome[1]. The classic symptoms are opthalmoplegia, nystagmus, ataxia, confusion and apathy[1]. Prompt therapy with thiamine rapidly reverses the symptoms. Approximately 84% of those who survive develop Korsacoff's psychosis, characterized by memory disturbances and confabulation[2].

The mechanisms underlying the pathogenesis of the lesions observed in WE are not understood completely. Thiamine is needed for glucose metabolism, neurotransmitter synthesis and maintenance of an osmotic gradient in cell membranes. Deficiency of thiamine results in reduced osmotic gradients and swelling of the intercellular spaces, especially in the periventricular regions, which have a high rate of thiamine mediated metabolism[3,4]. Neuropathologic changes of WE are found in the medial thalamus, the hypothalamus, the periaqueductal tissues of the mid brain, the grey matter of the floor of the fourth ventricle and in the mamillary bodies[3,4]. Histopathological changes in these sites include edema, petechial hemorrhages, demyelination and reactive astrocytosis. In the chronic form, (Korsacoff's psychosis) the mamillary bodies atrophy[2,4]. Even with thiamine treatment, mortality in WE is 10-20%[1].

The sensitivity and specificity of MR imaging in the diagnosis of this disease are 53% and 93% respectively[5]. MRI shows typical symmetric hyper intense areas in the paraventricular region of the thalamus and periaqueductal parts of the mid brain on T2 weighted images, more conspicuous on FLAIR images[5,6]. Following contrast administration, there may be enhancement in these areas as well as in the mamillary bodies[7]. Atrophy of the mamillary bodies is considered to be a sign of chronic WE[5]. Follow-up MRI in cases successfully treated with thiamine show reduction or disappearance of the hyperintensities and dilatation of third ventricle and aqueduct[4]. Normal MRI does not exclude the diagnosis of WE[5]. In alcoholics with WE, CT shows cortical,

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frontal lobe and cerebellum shrinkage in addition to low density areas in the paraventricular region of the thalamus. Low density areas in CT are uncommon with reported incidence of only 13%[5]. Thus MRI is more useful in the diagnosis of WE. Several tests, including red blood cell [RBC] transketolase activity assay, thiamine pyrophosphate [TPP], stimulation of RBC transketolase (the TPP effect), and 24-hour urinary excretion of thiamine can be used to assess thiamine status. Of these, RBC transketolase activity, with or without TPP challenge, is the most accurate assessment tool, but, is not widely available[8]. Rapid reversal of symptoms on administration of thiamine is in fact an accepted mode of confirming the diagnosis[1,5].

Bilateral paramedian thalamic infarction due to occlusion of a central unpaired thalamic perforating artery, an anatomic variant, can produce symmetrical hyper intensities in medial thalami but hyper intensities would not be observed in the other areas as observed in this patient[9].

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