FAITS CLINIQUES

Uncommon bilateral optic neuropathy in Wernicke's encephalopathy complicating gravidarum hyperemesis

Neuropathie optique bilatérale révélant une encéphalopathie de Wernicke au cours d'un hyperemesis gravidarum.

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Introduction

Wernicke encephalopathy (WE) is a rare and serious neurological disorder that results from vitamin B1 (Thiamin) deficiency. In the absence of treatment, Korsakov syndrome may ensue, and mortality may occur in nearly 30% of cases [1]. WE is often associated with alcoholism, malnutrition, or gastrointestinal diseases with malabsorption [2]. The association of «gravidarum hyperemesis» and WE was first described in 1939, and its incidence seems to be underestimated [1]. Hyperemesis gravidarum complicates 0.5 to 2% of pregnancies [3] and it is defined by profuse vomiting of the first trimester of pregnancy leading to weight loss, extracellular dehydration, and metabolic alkalosis with hypokalemia. Ocular signs, altered consciousness, and ataxia allow the positive diagnosis. This triad is however complete in only 30% [3]. The oculomotor disorders such as oculomotor palsy and nystagmus were the most reported ophthalmological signs. Optic neuropathy was rarely reported in WE. this serious complication may cause a profound visual acuity impairment. In this paper, we reported an uncommon case of bilateral anterior optic neuropathy with retinal bleeding in a pregnant woman with « gravidarum hyperemesis » that revealed the WE.

Case report

A 24-year-old pregnant woman was hospitalized in the gynecology department at week 15 of gestation for intractable vomiting (hyperemesis gravidarum) with acute stage B pancreatitis. After four days, the patient presented a brutal decrease of visual acuity of both eyes. The ophthalmological examination found a visual acuity limited to light perception bilaterally and altered relative afferent pupillary defects. Fundus examination showed a bilateral stage 2 papillary edema, a right optic disc hemorrhage,and a perifoveolar hemorrhage in the left eye (**Figure 1**). There was no diplopia or nystagmus. Metabolic encephalopathy was suspected and brain magnetic resonance imaging (MRI) was performed. It showed bilateral and symmetrical hyper intense lesions on T2-weighted and FLAIR sequences in periaqueductal gray matter, thalamus, and mammillary bodies, which confirmed WE (**Figure 2**). During the paraclinical workup, the patient developed ataxia. Thus, we put her on intravenous thiamine supplementation (200 mg every 8 hours for 3 days) maintained with 100 mg orally for two weeks.
Improvement occurred on day two of treatment, the visual acuity decreased to 10/10, papillary edema disappeared within two weeks (**Figure 3**), and ataxia disappeared within the first week of treatment.

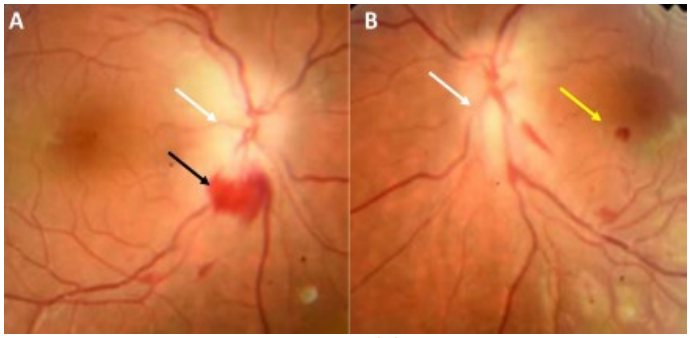

Figure 1. Fundus examination findings (A) Right eye fundus: stage 2 papillary edema (wight arrows) and optic disc hemorrhage (red arrow), (B) Left eye fundus: stage 2 papillary edema (wight arrows) and perifoveolar hemorrhage (yellow arrow)

Discussion

WE is an acute neurological syndrome resulting from a deficiency of thiamine due to excessive alcohol intake or gastrectomy [2]. It is also seen with prolonged fasting causing malnutrition, prolonged vomiting, gastrointestinal neoplasia, anorexia nervosa, malabsorption syndrome, bariatric surgery for morbid obesity, hemodialysis, and peritoneal dialysis [1]. In «gravidarum hyperemesis», profuse vomiting leads to weight loss, extracellular dehydration and metabolic alkalosis with hypokalemia. WE occurs because of a low uptake or loss of thiamine, increased demands of pregnancy, and depleted thiamine stores [4]. Thiamine deficiency leads to cerebral lesions within 2 to 3 weeks (petechial hemorrhagic lesions, edema, atrophy, or neuronal destruction) explaining the clinical manifestations [5].
The diagnosis of WE relies on a classic triad grouping ophthalmological disorders (93%), confusion with temporo-spatial disorientation (80%), and ataxia (76%) [2].
The most common ocular abnormalities are nystagmus, oculomotor palsy, and rarely complete ophthalmoplegia. They result from lesions of the pontine tegmentum including the abducens and oculomotor nuclei [6]. Optic nerve involvement is underreported in literature [7], optic disc edema was found in only 4% of reported cases. Vision loss is another uncommon finding in WE, it is due to optic neuropathy. Vision loss is typically severe, bilateral may lead to loss of light perception [8]. Retinal hemorrhages are also unusual and were seen in only 2% of cases [8].
Optic nerve involvement is probably due to toxicity resulting in accumulation of toxic intermediate metabolic products that disrupt the cellular homeostasis. The retinal hemorrhages are

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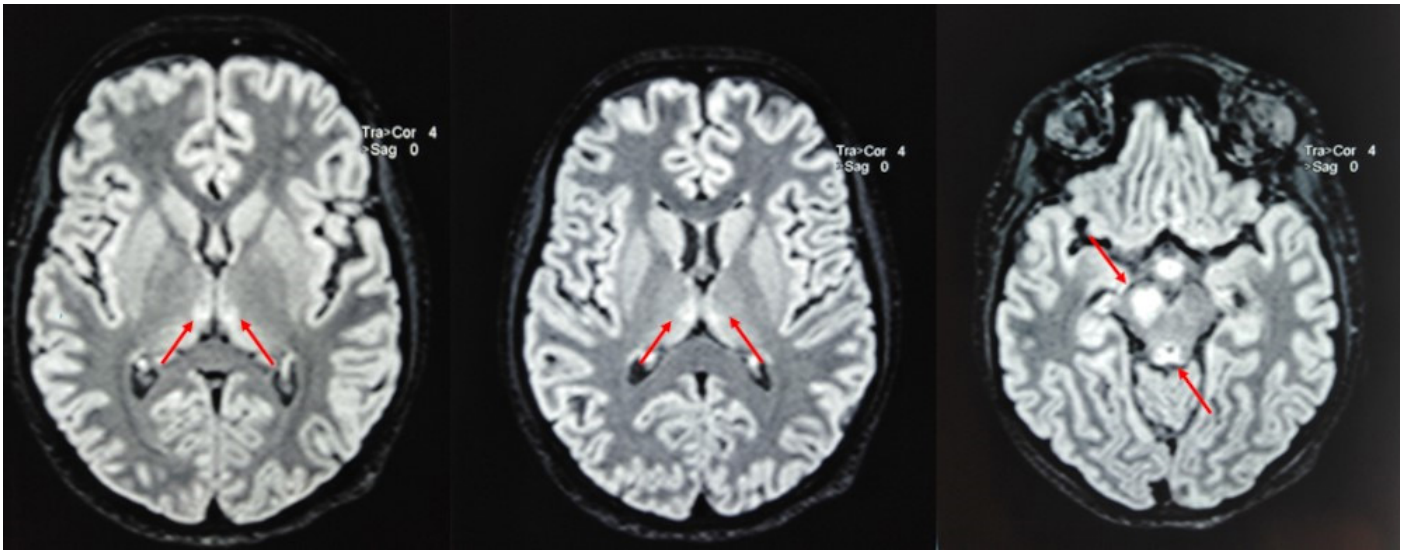

Figure 2. **MRI findings. Bilateral and symmetrical hyper intense lesions on T2-weighted and FLAIR sequences in periaqueductal gray matter, thalamus, and mammillary bodies (arrows)**

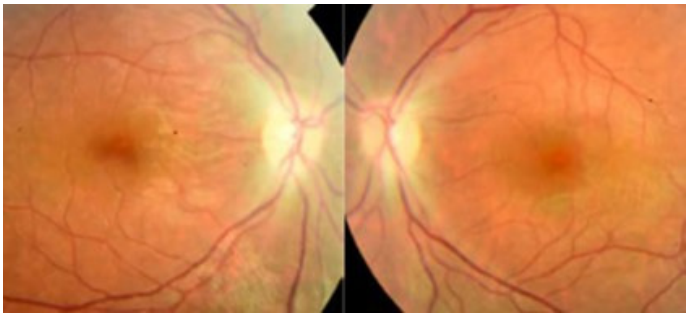

Figure 3. **Fundus aspect after two weeks of treatment. Total disappearance of the bilateral papillary edema**

often peripapillary but vomiting may also contribute to the development of retinal hemorrhages, located in the foveal or parafoveal region [8]. Other uncommon ophthalmic findings in patients with WE, were reported such as altered pupil reactivity, or size, impaired convergence, spasm of the near reflex, and ptosis [8].
Brain MRI had a sensitivity of 53% and a specificity of 93%. It cannot rule out WE but it is the best way to confirm the diagnosis since the blood dosage requires access to specialized laboratories and the results are not there obtained only late [4]. It showed bilateral and symmetrical hyperintense lesions on T2-weigh- ted and FLAIR sequences in periaqueductal gray matter, thalamus, mammillary bodies, and around the third ventricle [9]. The metabolism of the periventricular regions is particularly dependent on Thiamin, which explains radiological findings. Thiamine treatment must not be delayed by investigations results. An intravenous infusion of thiamine (200–500 mg thrice daily IV for 5–7 days) followed by oral thiamine (100 mg thrice daily for 1–2 weeks, and 100 mg/day) thereafter are recommended until there is no further improvement in signs and symptoms.
The reversibility of the disorders and the prognosis depend mainly on the duration of neurological signs before the introduction of treatment. Oculomotor abnormalities respond well to treatment. In most cases, horizontal and vertical gaze palsies and ptosis recover completely within days to weeks. In almost 60 % of cases, horizontal nystagmus can persist for months. Delay or failure of recovery should alert physicians to consider alternative diagnoses. The disc edema resolves and visual function is often preserved as in our case. However, if there is necrosis of ganglion cells or myelinated nerve fibers, there will be permanent vision impairment [8]. In untreated cases, Korsakoff's psychosis, with memory loss (global amnesia) and confabulation occurs [3]. It was described in 80% of cases because of lesions of the hippocampo-mamillo-thalamic circuit, with the predominance of mammillary anomalies [4].
Concerning the fetal prognosis, WE can lead to miscarriage, preterm birth, and intrauterine growth retardation [10]. According to many authors, fetal development is favorable when the treatment was carried out within 24 hours after the onset of neurological disorders [3].

Conclusion

Wernicke's encephalitis is a rare condition that remains underdiagnosed. This is a diagnostic emergency requiring early treatment to prevent complications.

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