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

Mejda Bouladi[1,3], Houda Lajmi[2,3], Amal Ben Othmen[1,3], Lamia El Fekih[1,3]

1. Ophthalmology Department. Mongi Slim University Hospital, La Marsa, Tunis, Tunisia.
2. Ophtalmology Department. FSI University Hospital, La Marsa, Tunis, Tunisia.
3. Faculty of Medicine of Tunis, University of Tunis El Manar.

Abstract

Wernicke encephalopathy (WE) is a rare neurological disorder that results from vitamin B1 (Thiamin) deficiency, classically characterized by the triad of ophtalmoplagia, altered consciousness, and ataxia. WE is often associated with alcoholism, malnutrition, or gastrointestinal diseases with malabsorption. The association of «gravidarum hyperemesis» and WE seems to be underestimated. We report a 24-year-old pregnant woman with hyperemesis gravidarum, who presented with decreased visual acuity of both eyes. Fundus examination showed a bilateral stage 2 papillary edema. brain magnetic resonance imaging (MRI) showed bilateral and symmetrical hyper intense lesions on T2-weighted and FLAIR sequences in periaqueductal gray matter, thalamus, and mammillary bodies, which confirmed WE complicated by bilateral optic neuropathy. Her symptoms resolved after thiamine treatment. This case raises of the possibility of optic neuropathy in WE, which is a diagnostic emergency requiring early treatment to prevent complications.

Key words: Wernicke encephalopathy, gravidarum hyperemesis, thiamine deficiency, optic neuropathy, optic disc edema

Résumé

L'encéphalopathie de Wernicke (EW) est un désordre neurologique rare, causé par un déficit en vitamine B1 (thiamine) et caractérisé par une triade diagnostic: des troubles oculomoteurs, une altération de la conscience et une ataxie. L'EW est souvent associée à l'alcoolisme, à la malnutrition ou aux pathologies gastro-intestinales avec malabsorption. Son association aux vomissements gravidiques est sous-estimés. Nous rapportons le cas d'une femme enceinte de 24 ans avec des vomissements gravidiques qui s'est présentée avec une diminution bilatérale de l'acuité visuelle. L'examen du fond d'œil a révélé un œdème papillaire bilatéral stade 2. L'imagerie par résonance magnétique cérébrale a montré des lésions bilatérales et symétriques en hypersignal T2 et FLAIR dans la substance grise périaqueducale, le thalamus et les corps mamillaires, ce qui a confirmé le diagnostic d'EW compliquée d'une neuropathie optique bilatérale. Les signes cliniques ont disparu après un traitement par la thiamine. Ce cas soulève la possibilité d'une neuropathie optique dans l'EW, qui est une urgence diagnostique nécessitant un traitement précoce pour prévenir les complications.

Mots clés: Encéphalopathie de Wernicke, vomissemnt gravidique, carence en thiamine, neuropathie optique, œdème papillaire

Correspondance
Bouladi Mejda
Ophthalmology department, Mongi Slim University Hospital, La Marsa, Tunis, Tunisia
Email: mejda.bouladi@fmt.utm.tn

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 which is a chronic neurological condition that primarily affects memory and cognitive function 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, 3].

Hyperemesis gravidarum complicates 0.5 to 2% of pregnancies [4] 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% [4]. 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.

Aim: 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. Direct pupillary reflex was reduced symmetrically. In both eyes, anterior segment was calm an intraocular pressure was normal. Fundus examination showed a bilateral stage 2 papillary edema, a right optic disc hemorrhage,and a perifoveolar hemorrhage in the left eye (figure 1).

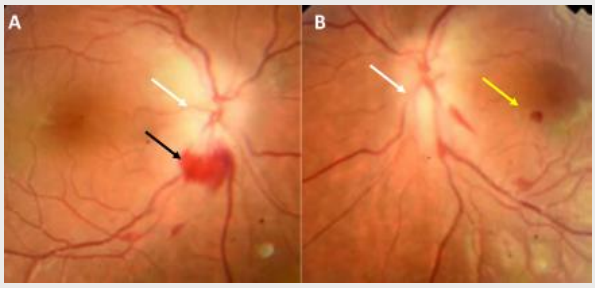

Figure 1. fundus examination findings

(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)

There was no nystagmus or oculomotor palsy. The patient was clinically dehydrated, her blood pressure was normal and she was afebrile. The neurological examination was unremarkable at the first presentation but after 24 hours she developed ataxia.

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 (figure 2).

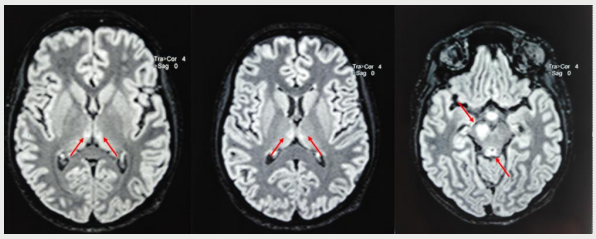

Figure 2. MRI findings

Bilateral and symmetrical hyper intense lesions on T2-weighted and FLAIR sequences in periaqueductal gray matter, thalamus, and mammillary bodies (arrows)

The diagnosis of WE complicated by bilateral optic neuropathy was retained. Thus, we put the patient 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 increased to 10/10, papillary edema disappeared within two weeks (figure 3), and ataxia disappeared within the first week of treatment.

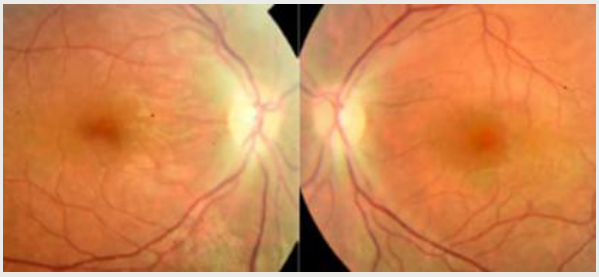

Figure 3. fundus aspect after two weeks of treatment

Total disappearance of the bilateral papillary edema

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 [5]. Thiamine deficiency leads to cerebral lesions within 2 to 3 weeks (petechial hemorrhagic lesions, edema, atrophy, or neuronal destruction) explaining the clinical manifestations [6].

The diagnosis of WE relies on a classic triad grouping ophthalmological disorders (93%), confusion with temporo-spatial disorientation (80%), and ataxia (76%) [2].

Our patient presented with atypical presentation of WE, the complete triad was absent and the ophtalmological disorder was bilateral optic neuropathy. According to

the literature, the most common ocular abnormalities are nystagmus, oculomotor palsy, and rarely complete ophthalmoplegia [7]. They result from lesions of the pontine tegmentum including the abducens and oculomotor nuclei [8]. Optic nerve involvement is underreported in literature [8], optic disc edema was found in only 4% of reported cases [7]. 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 [10]. Retinal hemorrhages are also unusual and were seen in only 2% of cases [10].

Various hypotheses could explain the mechanism of optic disc edema and optic neuropathy in WE. Thiamine deficiency can lead to brain damage and increased intracranial pressure responsible of the optic disc swelling. In addition, increased pressure around the optic nerves could lead to the obstruction of axoplasmic flow. Another possible explanation is that optic neuropathy in WE could be similar to nutritional neuropathies. Finally, Optic nerve involvement is probably due to toxicity resulting in accumulation of toxic intermediate metabolic products that disrupt the cellular homeostasis [7]. The retinal hemorrhages are often peripapillary but vomiting may also contribute to the development of retinal hemorrhages, located in the foveal or parafoveal region [10].

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 [10].

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 [5]. 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 [11]. 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 [10].

In untreated cases, Korsakoff's psychosis, with memory loss (global amnesia) and confabulation occurs [4]. It was described in 80% of cases because of lesions of the hippocampo-mamillo-thalamic circuit, with the predominance of mammillary anomalies [5].

Concerning the fetal prognosis, WE can lead to miscarriage, preterm birth, and intrauterine growth retardation [12]. According to many authors, fetal development is favorable when the treatment was carried out within 24 hours after the onset of neurological disorders [4].

To conclude WE should be considered in pregnant women suffering from frequent vomiting with bilateral optic neuropathy. Rapid administration of intravenous thiamine is crucial in treating this condition, as it can lead to a rapid recovery if identified and treated early.

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