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Unusual Radiological Presentation of Gayet-Wernicke Encephalopathy: About A Case Report

Azzahiri I[1*], Benfaddoul O[1], Zouita B[1], Basraoui D[1], Jalal H[1]

[1]Department of Radiology, Mother and Child Hospital, Mohamed VI University Hospital, Cadi Ayyad University, Marrakech, Morocco

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***Corresponding author:** Azzahiri I
Department of Radiology, Mother and child hospital, Mohamed VI university hospital, Cadi Ayyad University, Marrakech, Morocco

Abstract **Case Report**

Gayet-Wernicke encephalopathy is an acute neurological disorder resulting from thiamine deficiency. Its diagnosis can be challenging due to its varying clinical presentations, which can be atypical or nonspecific. Although chronic alcoholism is a common cause, Gayet-Wernicke encephalopathy can also develop in conditions associated with thiamine deficiency, including undernutrition, prolonged fasting, chronic vomiting, and exclusive artificial feeding. Although rare, the condition is reversible if promptly diagnosed and treated. Here, we present a case of Gayet-Wernicke encephalopathy in a pregnant patient who presented with intractable vomiting, and whose radiological findings were atypical, making the diagnosis more challenging.

Keywords: Gayet-Wernicke, encephalopathy, MRI, thiamine, case report.

INTRODUCTION

Gayet-Wernicke encephalopathy is an acute and severe neuropsychiatric syndrome that was first described by Carl Wernicke in 1881. It is a rare disease with symptoms that typically include the classic triad of ataxia, ophthalmoplegia, and confusion. While the condition has historically been strongly associated with alcoholism and neurological signs, it is now recognized to occur in a variety of clinical contexts.(Davies *et al.*, 2011) (Sechi & Serra, 2007)

Despite the diversity of its clinical presentations, early diagnosis and treatment of Gayet-Wernicke encephalopathy remain critical to preventing potentially life-threatening complications. This can be challenging as the condition is primarily diagnosed through clinical assessment, and may be difficult to distinguish from other neurological conditions. Additionally, while low blood thiamine levels are considered a hallmark of the disease, it is possible for patients to present with normal levels, further complicating diagnosis. Magnetic resonance imaging (MRI) can be a useful tool for identifying specific features of Gayet-Wernicke encephalopathy, although it has a relatively low sensitivity of 53% and specificity of 93%.(Sechi & Serra, 2007) (Jain *et al.*, 2020)

In our case report, we describe a patient who presented with prolonged intractable vomiting and subsequently developed Wernicke's encephalopathy. The MRI findings in this case were not classical, underscoring the challenges of diagnosis and emphasizing the importance of considering a broad range of potential clinical presentations in order to accurately identify and treat Gayet-Wernicke encephalopathy.

CASE REPORT

This is a 23-year-old woman who was 16 weeks pregnant and admitted to the maternal intensive care unit due to intractable vomiting and electrolyte imbalances. The onset of symptoms began 2 months ago with postprandial vomiting complicated by unrelenting epigastric pain despite treatment, profound fatigue, and unquantified weight loss. Clinical examination revealed a conscious patient who was stable on a hemodynamic and respiratory level, bilateral exophthalmos, bradypsychia, and a normal neurological examination. During hospitalization, the patient's neurological condition worsened with the appearance of ataxia, nystagmus, and altered consciousness. Blood tests showed hemoglobin at 13.3 g/dL, white blood cell count at 9400, hypokalemia with a potassium level of

1.9 mmol/L, and a low TSHus level of 0.009, while cortisol levels were normal at 192 micro/L.

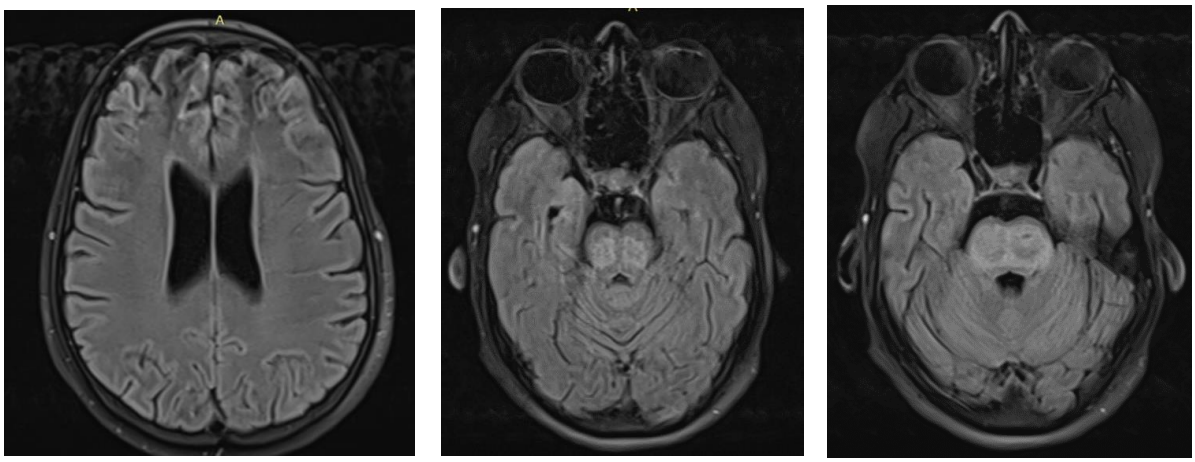

Figure 1: FLAIR sequence: brain MRI showing signal anomalies in Flair hypersignal visible at the frontal cortical level associated with mesencephalic signal anomalies, involving the protuberance, mesencephalic tegmentum, the locus Niger and sparing the red nuclei (arow)

An MRI was performed to investigate the cause of the worsening neurological condition. Due to the risk of general anesthesia on the mother and fetus, a minimal protocol MRI was performed without gadolinium injection. The MRI showed signal abnormalities visible in bilateral frontal and temporal cortical areas in hyperintense T2/Flair and diffusion (Fig 1). It also showed signal abnormalities visible in the mesencephalic area, affecting the pons, mesencephalic tegmentum, locus niger, and sparing the red nuclei, in hypointense T1, hyperintense T2/Flair, and diffusion (Fig 2-3). Based on these findings, there was a strong probability of Gayet Wernicke encephalopathy given the hyperemesis gravidarum and gradually developing neurological symptoms. However, the radiological presentation was atypical due to the centropontine signal abnormalities and those observed in the mesencephalic tegmentum and locus niger. Correlating with the clinical data, the diagnosis of Gayet Wernicke encephalopathy was the first diagnosis suggested, but central pontine myelinolysis was also considered. Due to the patient's worsening neurological condition, the decision was made to administer vitamin B1 without waiting for the biological results. The patient's clinical condition improved with normalization of neurological status after 10 days of treatment.

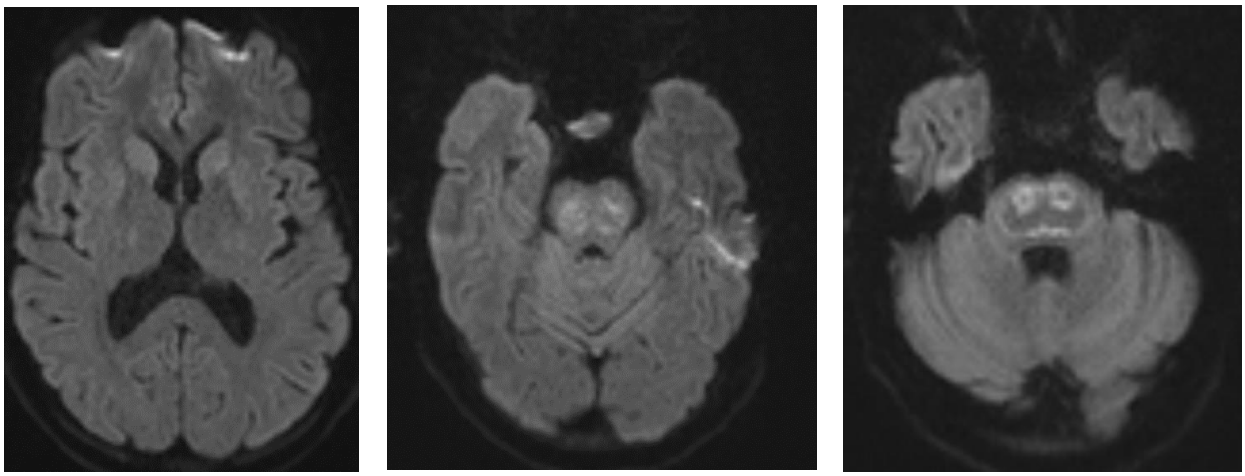

Figure 2: Diffusion sequence: brain MRI showing signal anomalies in Flair hypersignal visible at the frontal cortical level associated with mesencephalic signal anomalies, involving the protuberance, mesencephalic tegmentum, the locus Niger and sparing the red nuclei (arow)

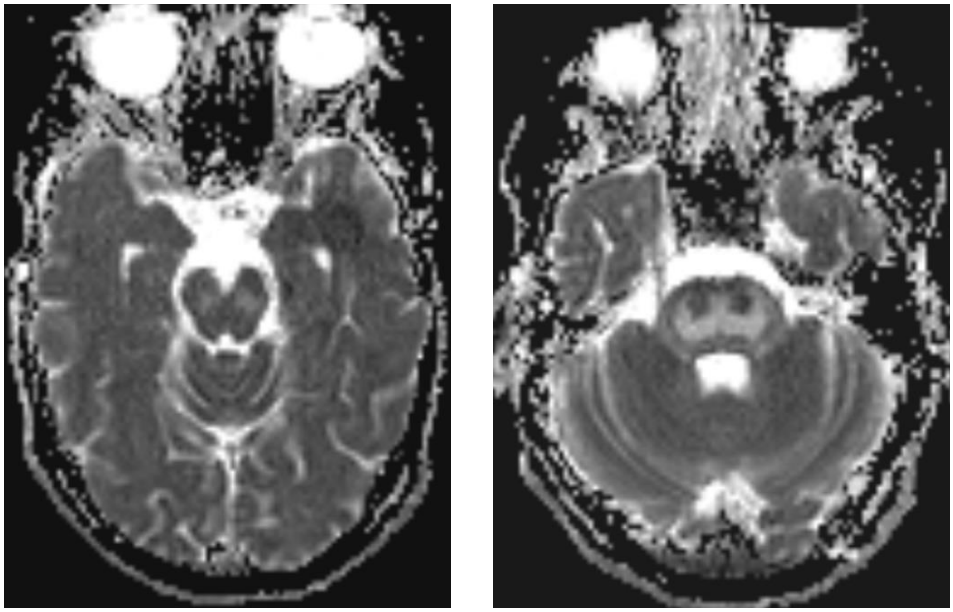

Figure 3: Diffusion sequence: diffusion sequence with ADC: Absence of ADC restriction in the flair and diffusion hypersignal areas

DISCUSSION

Gayet-Wernicke encephalopathy is a rare, acute, and severe neuropsychiatric disorder that arises as a result of a deficiency in vitamin B1 (thiamine). The condition was first described in 1881 by Carl Wernicke in a male alcoholic and a female patient with incoercible vomiting(Oudman *et al.,* 2019), and it is estimated to have an autoptic prevalence of 0.8-2.8%. This neurological disorder predominantly affects men, and it is often observed in the context of chronic alcoholism if dietary thiamine intake is insufficient or in severe undernutrition. Other causes include prolonged vomiting, prolonged parenteral feeding, hunger strike, anorexia nervosa, gastrointestinal surgery, cancer, chemotherapy, and several other factors.(Davies *et al.*, 2011) (EL Badri *et al.,* 2021)

Thiamine deficiency leads to brain lesions that can range from petechial hemorrhagic lesions and edema to atrophy and destruction of neurons, and these lesions may occur within 2 to 3 weeks. While confusion is the most common symptom in about 82% of cases, only 16.5% of patients present with the classic triad of ataxia, ophthalmoplegia, and confusion, while approximately 19% of patients have none of the classic symptoms. Other symptoms can include oculomotor disturbances, gait ataxia, and polyneuropathy, among others.

Vitamin B1 is present in the diet, and daily requirements are approximately 1.5 mg/d. Absorption occurs in the proximal small intestine, and after intestinal absorption, thiamine is phosphorylated in the liver to give thiamine pyrophosphate, the active form of vitamin B1 and an essential coenzyme in several biochemical reactions. Thiamine is mainly distributed to the most consuming organs, including the heart, liver, brain, and kidneys. Thiamine is mainly transported across the blood-brain barrier by transporters (active transport) and by a slow, passive process (when the concentration exceeds the active transport threshold).

When Gayet-Wernicke encephalopathy is suspected, magnetic resonance imaging (MRI) is the preferred examination method with a sensitivity of 53% and a specificity of 93%. In its classic form, this condition presents with lesions that appear hypointense in T1 and hyperintense in T2 and Flair. These lesions are characterized by their specific locations, including bilateral and symmetrical involvement of the median thalami and the peri-ventricular regions of the third ventricle in 85% of cases, as well as involvement of the periaqueductal area in 65% of cases and the mammillary bodies in 58% of cases. Other localizations, such as the tectal plate, the elongated marrow, and the cerebellum, are also possible. Gadolinium injection leads to contrast enhancement of the mammillary bodies, tectal plate, thalami, and periaqueductal area.In the late phase, diffuse cerebral atrophy and atrophy of the mammillary bodies and vermis are observed. The restriction of ADC indicates the irreversibility of the lesions.However, normal imaging does not necessarily exclude the diagnosis.(de Oliveira *et al.,* 2019) (Segal *et al.,* 2016)

On magnetic resonance imaging (MRI), the involvement of the peri-ventricular regions and medial thalamus may not be entirely specific for Gayet-Wernicke encephalopathy, as they can also be present in demyelinating pathologies caused by inflammatory or infectious diseases such as Behçet's disease, or central and extra-pontine myelinolysis.

In this case report, given the clinical setting and radiological presentation, extrapontine myelinolysis was the main differential diagnosis we had considered. However, the neurological improvement seen a few days after thiamine administration ruled out this diagnosis.

While one would expect low thiamine levels in Wernicke's encephalopathy, thiamine tests have well-known limitations, and serum thiamine levels are a poor measure of thiamine status. Several laboratories now use high-performance liquid chromatography with fluorescence detection to measure thiamine pyrophosphate in blood, but this assay does not necessarily reflect intracerebral thiamine levels. Additionally, Wernicke's encephalopathy cannot be diagnosed simply by measuring the circulating thiamine level, and hence the priority of brain MRI in complementary investigations.(Davies *et al.*, 2011) (Jain *et al.*, 2020)

Early diagnosis and treatment are crucial, and the mortality rate is 17-20% if not treated adequately. Treatment should be started as soon as the diagnosis is suspected and continued until clinical improvement. Complete remission of symptoms is possible with early diagnosis and adequate treatment. Prevention is achieved by parenteral administration of vitamin B1 to all patients at risk (Sechi & Serra, 2007) (Thomson *et al.*, 2002).

CONCLUSION

Gayet-Wernicke encephalopathy is a rare and severe condition characterized by a triad of ophthalmoplegia, mental confusion, and ataxia, which is highly suggestive in a high-risk situation. The diagnostic reference for Gayet-Wernicke encephalopathy is magnetic resonance imaging, and if necessary and feasible, vitamin levels will be measured. It is important to note that a normal thiamine level does not exclude the diagnosis of Gayet-Wernicke encephalopathy. Early treatment is crucial in order to achieve a favorable outcome and prevent the development of Korsakoff syndrome (Oudman *et al.*, 2019).

DISCLOSURE OF INTEREST

The authors declare that they have no conflicts of interest concerning this article.

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