

Case Report

## Unusual presentation of Wernicke's encephalopathy with hypertension

### *Hipertansiyon ile prezente olan sıradışı Wernicke ensefalopati olgusu*

Merve Hilal Dolu<sup>1</sup>, Filiz Mıhçı<sup>1</sup>, Haydar Ali Taşdemir<sup>1</sup>, Ömer Faruk Aydın<sup>2</sup>

<sup>1</sup>Ondokuz Mayıs University, Pediatric Neurology, Samsun, Türkiye

<sup>2</sup>Samsun Liv Hospital, Pediatric Neurology, Samsun, Türkiye

#### ABSTRACT

Wernicke's encephalopathy is an acute neuropsychiatric syndrome caused by thiamine (vitamin B1) deficiency. The classical triad of Wernicke's encephalopathy is mental confusion, oculomotor dysfunction, and ataxia. It is generally associated with alcoholism and prolonged parenteral nutrition without vitamin supplementation. Wernicke's encephalopathy must be treated urgently to prevent death and neurological morbidity. In this report, we present an unusual pediatric case of Wernicke's encephalopathy in which the patient initially presented with hypertension. Intravenous thiamine of 100 mg daily treatment was started, after which she became normotensive. This is the first reported case of Wernicke's encephalopathy presenting with hypertension and resolved after thiamine treatment.

**Keywords:** *Wernicke's encephalopathy, thiamine deficiency, hypertension, psychotic*

#### ÖZET

Wernicke ensefalopatisi, tiamin (B1 vitamini) eksikliğine bağlı akut bir nöropsikiyatrik sendromdur. Wernicke ensefalopatisinin klasik triadı mental konfüzyon, okulomotor disfonksiyon ve ataksidir. Genellikle alkolizm ve ayrıca herhangi bir vitamin takviyesi olmaksızın uzun süreli parenteral beslenme ile ilişkilidir. Ölüm ve nörolojik morbiditeyi önlemek için acilen tedavi edilmelidir. Bu raporda, başlangıçta hipertansiyon ile başvuran bir pediatrik sıradışı Wernicke Ensefalopatisi olgusunu sunuyoruz. Hastamıza IV tiamin 100 mg günlük tedavi başlanmış, bu tedaviden sonra psikozu düzelmiş ve normotansif hale gelmiştir. Hipertansiyon ile başvuran ve Wernicke ensefalopatisinde tiamin tedavisi sonrası düzelen ilk olgudur.

**Keywords:** *Wernicke ensefalopatisi, tiamin eksikliği, hipertansiyon, psikotik*

#### INTRODUCTION

Wernicke's encephalopathy is an acute neuropsychiatric syndrome caused by thiamine (vitamin B1) deficiency that requires emergency treatment to prevent death and neurological morbidity. The classical triad of Wernicke's encephalopathy is mental confusion, oculomotor dysfunction, and ataxia (1-4). It is generally asso-

ciated with alcoholism, anorexia nervosa, hyperemesis gravidarum, and also prolonged parenteral nutrition without vitamin supplementation in malignancies and after gastrointestinal surgery (5). MRI studies typically show an increased T2 signal that is bilaterally symmetrical in the paraventricular regions of the thalamus, the

Received: 04.01.2023 · Accepted: 01.03.2023 · Published: 30.04.2023

Correspondence / Yazışma: Merve Hilal Dolu · Ondokuz Mayıs Üniversitesi, Pediatrik Nöroloji Bölümü, Samsun, Türkiye · dr.mervehilal@hotmail.com

Cite this article as: Dolu MH, Mihci F, Tasdemir HA, Aydın OF. Unusual presentation of Wernicke's encephalopathy with hypertension. *Pediatr Acad Case Rep.* 2023;2(2):60-62.

hypothalamus, mamillary bodies, the periaqueductal region, and the floor of the fourth ventricle and the midline cerebellum (6).

Here, we report an unusual pediatric case of Wernicke's encephalopathy in which the patient presented with hypertension. She was initially treated with intravenous thiamine at 100 mg daily, which was raised to 500 mg three times a day. After this treatment, the patient completely recovered, with no symptoms and normal radiological findings.

## CASE REPORT

A 12-year-old girl operated on for a perforated appendix and widespread intra-abdominal abscess and was sent to the pediatric neurology department for double vision, ataxic gait, and hypertension. She had undergone gastric surgery three times and, because oral intake was generally not possible, had been administered peripheral total parenteral nutrition (TPN) for 1.5 months. She was not administered any vitamin supplements and complained of progressive blurred and double vision with ataxic gait.

In the neurological examination, bilateral papilledema, retinal hemorrhages, vertical nystagmus, and sixth nerve palsy in the left eye were determined. Systemic blood pressure was 160/100 mmHg. MRI studies typically showed an increased signal in T2/flair scans that was bilaterally symmetrical in the periventricular regions of the thalamus (Fig. 1).



**Figure 1.** Increased signal bilaterally symmetrical in periventricular regions of thalamus

Intracranial pressure syndrome was initially suspected because of bilateral papilledema and hypertension. The lumbar puncture showed a normal opening cerebrospinal fluid pressure of 17 cm H<sub>2</sub>O, so increased intracranial pressure syndrome was ruled out, and we investigated other possible reasons for hypertension. The vasculitis markers, echocardiography, renal ultrasonography and renal Doppler were all normal. The symptomology and neuroimaging signs led us to diagnose Wernicke's encephalopathy after thiamine deficiency was confirmed. The patient's blood thiamine level

was 17.1 µg/L (25–75 µg/L), and 100 mg of intravenous thiamine replacement was started.

By treatment day 5, the symptoms had improved slightly, with the ataxic gait and nystagmus gone. The patient became normotensive and no longer required antihypertensive medication. However, psychotic symptoms were exhibited. Therefore, we increased the amount of thiamine replacement to 500 mg three times a day for two weeks. At the end of this period, by which the blood thiamine level was 170 µg/L (25-75 µg/L), the patient was no longer showing psychotic symptoms. After treatment, the MRI findings also became normal. The patient was discharged with an oral thiamine supplement. The patient's consent was obtained in this case study.

## DISCUSSION

Wernicke's encephalopathy was first described by Dr. Carl Wernicke in 1881 (1). This disease is caused by thiamine deficiency, primarily among alcoholic adults, although it also occurs in impaired nutritional states treated for a period with TPN (2,5,7). The disorder results from thiamine deficiency; the biologically active form of thiamine pyrophosphate is an essential coenzyme in several biochemical pathways in the brain (5,8). There is limited tissue storage of thiamine, and levels are maintained only through continuous dietary intake.

Subclinical thiamine deficiency symptoms may be nonspecific, such as frequent headaches, fatigue, irritability, abdominal discomfort, and a decline in the growth rate of children (9). Severe and acute thiamine deficiency presents with Wernicke's encephalopathy, characterized by mental status changes, ocular abnormalities, and motor problems, such as gait disturbance and ataxia. However, this classic triad is apparent in as few as 10% of patients (4). Other uncommon symptoms are stupor, hypotension, tachycardia, hypothermia, bilateral visual disturbance, papilledema, and epileptic seizures (5). Hypotension is a rare symptom of Wernicke's encephalopathy, as reported in some adult cases (10,11).

In the case reported here, it was speculated that prolonged hypertension could be due to dysautonomia resulting from thiamine deficiency (10). After intravenous thiamine treatment at 100 mg daily for five days, the hypertension was resolved, along with the other neurological symptoms. However, psychotic symptoms were exhibited, so we raised the thiamine dose to 500 mg three times a day for two weeks, after which the psychotic symptoms were suppressed, and MRI results were normal.

Thiamine deficiency is a treatable disease with a usually good prognosis. In Wernicke's encephalopathy, thiamine replacement should be initiated as soon as suspected since morbidity or mortality may increase because of delayed treatment (12). Patients require intravenous administration of at least intravenous 100 mg of thiamine daily for several days. However, there is still no guideline for pediatric treatment protocol in Wernicke's encephalopathy. A dosage of 50–100 mg daily or 1.8 mg thiamine per 1,000 kcal has been suggested, and 100 mg daily thiamine replacement is routinely recommended in pediatric patients (13). High doses of thiamine therapy have been reported in adult patients (14,15). However, only one pediatric case of Wernicke's encephalopathy has been reported involving high-dosage thiamine treatment (16).

In conclusion, to our knowledge, this is the first pediatric case of Wernicke's encephalopathy presenting with hypertension and resolved after thiamine treatment. We recommend that a high dose of thiamine (500 mg three times daily) treatment should be in the clinician's mind with incomplete recovery or worsened symptoms in pediatric patients with Wernicke's encephalopathy.

#### Patient Consent Form / Hasta Onam Formu

The parents' of this patient consent was obtained for this study.

#### Conflict of Interest / Çıkar Çatışması

The authors declared no conflicts of interest with respect to authorship and/or publication of the article.

#### Financial Disclosure / Finansal Destek

The authors received no financial support for the research and/or publication of this article.

## REFERENCES

1. Wernicke C. Lehrbuch der Gehirnkrankheiten für Aerzte und Studierende. Vol. 2. Theodor Fischer. Berlin: Kassel, 1881: 229-42.
2. Hazell AS, Butterworth RF. Update of cell damage mechanisms in thiamine deficiency: focus on oxidative stress, excitotoxicity and inflammation. *Alcohol Alcohol* 2009; 44: 141-7.
3. Campbell AC, Russel WR. Wernicke's encephalopathy: the clinical features and their probable relationship to vitamin B deficiency. *Q J Med* 1941; 10: 41-64.
4. Harper CG, Giles M, Finlay-Jones R. Clinical signs in the Wernicke-Korsakoff complex: a retrospective analysis of 131 cases diagnosed at necropsy. *J Neurol Neurosurg Psychiatry* 1986; 49: 341-5.
5. Sechi G, Serra A. Wernicke's encephalopathy: new clinical settings and recent advances in diagnosis and management. *Lancet Neurol* 2007; 6: 442-55.
6. Zuccoli G, Siddiqui N, Bailey A et al. Neuroimaging findings in pediatric Wernicke encephalopathy: a review. *Neuroradiology* 2010; 52: 523-9.
7. Thomson AD, Guerrini I, Bell D, et al. Alcohol-related brain damage: report from a Medical Council on Alcohol Symposium, June 2010. *Alcohol Alcohol* 2012; 47: 84-91.
8. Manzo L, Locatelli C, Candura SM, et al. Nutrition and alcohol neurotoxicity. *Neurotoxicology* 1994; 15: 555-65.
9. Tanphaichitr V. Thiamin. In: Shils ME, Olson JA, Shike M, Ross AC, eds. *Modern Nutrition in Health and Disease*, 9th edn. Baltimore MD: Williams and Wilkins, 1999: 381-89.
10. Maeda H, Kuriyama A, Tanaka T. Prolonged hypotension associated with wernicke's encephalopathy. *Eur J Clin Nutr* 2018; 72: 168-9.
11. Gardiner S, Hartzell T. Thiamine Deficiency: A Cause of Profound Hypotension and Hypothermia After Plastic Surgery. *Aesthetic Surgery Journal* 2015; 35:1.
12. Harper CG, Butterworth RF. Nutritional and metabolic disorders. In: Graham DI, Lantos PL, editors. *Greenfield's neuropathology*. London: Arnold 1997: 601-55.
13. Manzanares W, Hardy G. Thiamine supplementation in the critically ill. *Curr Opin Clin Nutr Metab Care* 2011; 14: 610-7.
14. Caso F, Fiorino A, Falautano M, Leocani L, Martinelli V, Minicucci F, et al. Treatment of Wernicke's encephalopathy with high dose of thiamine in a patient with pyloric sub-stenosis: description of a case. *Neurol Sci* 2010; 31: 859-61.
15. Nishimoto A, Usery J, Winton JC, Twilla1J. High-dose parenteral thiamine in treatment of Wernicke's encephalopathy: Case Series and Review of the Literature. *In Vivo* 2017; 31: 121-4.
16. Park SW, Yi YY, Han JW, et al. Wernicke's encephalopathy in a child with high dose thiamine therapy. *Korean J Pediatr* 2014; 57: 496-9.