NEURO

Openventio PUBLISHERS

ISSN 2377-1607 Open Journal http://dx.doi.org/10.17140/NOJ-1-104

Case Report

*Corresponding author
José Pablo Suárez-Llanos, MD
Endocrinology Department of Hospital Universitario Nuestra Señora de la Candelaria (HUNSC)
Ctra. Del Rosario nº145, Santa Cruz de Tenerife 38010, Spain
E-mail: psuarezllanos@gmail.com

Volume 1 : Issue 1
Article Ref. #: 1000NOJ1104

Article History
Received: September 30th, 2014
Accepted: November 8th, 2014
Published: November 10th, 2014

Citation
Suárez-Llanos JP, Gómez-Álvarez B, Lallena-Arteaga E, et al. Wernicke encephalopathy and other vitamin deficiencies in a patient with home parenteral nutrition. *Neuro Open J.* 2014; 1(1): 16-19. doi: 10.17140/NOJ-1-104

Copyright
©2014 Suárez-Llanos JP. This is an open access article distributed under the Creative Commons Attribution 4.0 International License (CC BY 4.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Wernicke Encephalopathy and Other Vitamin Deficiencies in a Patient with Home Parenteral Nutrition

José Pablo Suárez-Llanos[1*], Beatriz Gómez-Álvarez[1], Elisa Lallena-Arteaga[2], Estefanía González-Melo[1], Nieves Caracena-Castellano[1], María Pilar Díaz-Ruiz[3] and María Luz Padilla-Salazar[4]

[1]Endocrinology Department, Hospital Universitario Nuestra Señora de la Candelaria (HUNSC), Santa Cruz de Tenerife, Spain

[2]Neurology Department, HUNSC, Santa Cruz de Tenerife, Spain

[3]Pharmacy Department, HUNSC, Santa Cruz de Tenerife, Spain

[4]General Medicine Department, HUNSC, Santa Cruz de Tenerife, Spain

ABSTRACT

Wernicke Encephalopathy (WE) is an acute and serious secondary neurological syndrome caused by a deficit of thiamine or vitamin B1. Although it rarely presents with the classic triad, the most frequent presenting symptoms are: mental confusion, ocular-motor dysfunction and ataxia. Laboratory and image studies can be useful, but diagnosis is fundamentally clinical, and if this syndrome is suspected treatment must be started as soon as possible. The most frequent cause in our setting is chronic alcoholism; however, there are increasing numbers of reported cases secondary to parenteral nutrition not correctly supplemented with vitamins. Here we present a case of WE secondary to parenteral nutrition not correctly supplemented with vitamins. This patient presented the classic clinical triad and was therefore diagnosed and treated rapidly, responding to treatment in an effective manner. Thus, the clinical presentation corresponded to a deficit of other vitamins associated with the same vitamin complex iv that was not administered, including a regenerative anemia and eczema craquele. This case emphasizes the need to supplement parenteral nutrition with thiamine and other vitamins. At the same time, it stresses the importance of suspecting this syndrome in all clinically compatible patients with non-vitamin supplemented parenteral nutrition.

KEYWORDS: Wernicke Encephalopathy (WE); Home parenteral nutrition; Thiamine deficiency.

ABBREVIATIONS: Wernicke Encephalopathy (WE); Radiotherapy (RT); Home Parenteral Nutrition (HPN); Computerized Tomography (CT); Magnetic Resonance Imaging (MRI).

CASE REPORT

We present the case of a woman 55 years of age, who presented at the hospital emergency room with change of gait, confusion and double vision over the previous two weeks. Her personal history showed a cervical adenocarcinoma ten years previously which required surgical treatment in addition to chemo- and radiotherapy, which was in remission at the time. As a side effect of the radiotherapy treatment she presented severe actinic enteritis which caused intolerance to oral and enteral feeding, and for this reason she had received total Home Parenteral Nutrition (HPN) for two and a half years.

In the context of a low emotional state for personal reasons, the patient neglected the correct management of her parenteral nutrition, having suspended her vitamin supplements (Table 1) for 20-30 days. At the end of this time she presented at the emergency room complain

ISSN 2377-1607 Open Journal http://dx.doi.org/10.17140/NOJ-1-104

Table 1: Composition of the vitamin complex administered to the patient	
Retinol Palmitate (Vitamin A)	3500 IU
Cholecalciferol (Vitamin D3)	220 IU
Alfa Toco pherol (Vitamin E)	11,2 IU
Ascorbid acid (Vitamin C)	125 mg
Nicotinamide (Vitamin B3)	46 mg
Pantothenic acid (Vitamin B5)	16,15 mg
Pyridoxine (Vitamin B6)	5,5 mg
Riboflavin (Vitamin B2)	5,67 mg
Thiamine (Vitamin B1)	3,51 mg
Folinic Acid	414 mcg
D-Biotin	69 mcg
Cyanocobalamin (Vitamin B12)	6 mcg

ing of asthenia, progressive instability of gait (to the point where she could no longer ambulate without aid) for one week prior to presentation, accompanied by a loss of coordination in the superior extremities which made management of the central line, and double vision in all positions of gaze.

On physical examination the patient was disorientated in time and space, without affecting language or speech. Extrinsic ocular motility was altered, presenting paresis of both external rectus muscles and one horizontal rotatory nystagmus, constantly in all positions of gaze. From the motor aspect, muscular balance was normal (motor strength 5/5), as were the osteotendinous reflexes, with the exception of the achilles which were bilaterally absent. The cutaneous plantar reflexes were flexors. The sensitivity study revealed a mild distal hypopalesthesia in the inferior extremities, without other noteworthy changes. The patient also presented ataxia of the trunk which made seating difficult and bipedal ambulation impossible without aid.

Regarding additional tests that were carried out, the analytic study detected the presence of thrombocytopenia (102,000 platelets/ml) and normocytic normochromic anemia (hemoglobin 10,3 g/dl, mean corpuscular volume 87,5 fl), in addition to hypoalbuminemia of 2,53 g/dl, indicating moderate protein deficiency. The remainder of the additional tests, including radiography of the thorax, abdomen and a CT scan of the cranium obtained results within normal parameters.

Considering the clinical history and the described findings on physical examination, Wernicke syndrome was suspected, and for this reason treatment was started with 300 mg of thiamine iv every 8 hours, subsequently continuing with 300 mg iv/day of the vitamin for one week, while simultaneously reintroducing parenteral nutrition with vitamins and minerals incorporated. It should be taken into account that since the patient attended on the weekend, it was not possible to carry out a Magnetic Resonance Imaging (MRI) scan or any non-urgent analysis in her referring hospital, and for this reason thiamine levels previous to iv administration could not be determined.

On detection of low levels of folic acid and the further suspicion of deficiency in other B-group vitamins, a blood smear was taken which indicated serious reticulocytosis. Following transfusion with a concentrate of erythrocytes and 100 mg of folic acid iv administered in two doses, the anemia improved, also showing a great quantity of reticulocytes in the smear over the two days of treatment. Analysis of vitamin levels in the blood, including complex B, were normal (Vitamin B1: 10,81 µg/l, Vitamin B6: 13 nmol/l, Vitamin B12: 897 pg/ml, folic acid: 15,30 ng/ml), after three days of the treatment described previously. Half way through the hospital stay the patient developed non-pruritic dry eczema on the right thigh, corresponding to eczema craquele (xerodermic), probably caused by nutritional deficiency.

The patient recovered equally well from the confusional syndrome as from the ophthalmoparesis in less than 24 hours, continuing with a certain degree of ataxia and instability of gait which improved with rehabilitation, although they did not recover adequate ambulation. The eczema was also resolved with topical treatment.

DISCUSSION

HPN is indicated in patients with an inability use the digestive tract, as occurred in our patient, who presents chronic intestinal insufficiency due to actinic enteritis, indicating standard parenteral nutrition of 1,700 kcal, to which is added daily a complex of minerals and multivitamins adjusted to their needs. Poor praxis or adherence to the treatment can give rise to a state of calorific-protein or micronutrient malnutrition, causing the typical symptoms of each deficiency.[1,2]

Wernicke syndrome is a neurological complication, relatively common, stemming from a deficit of thiamine or vitamin B1 and which can lead to an extremely negative outcome with high mortality if not correctly identified and treated.[3] Thiamine is a water soluble vitamin which is involved in energy-producing enzymatic cascades during glucose metabolism. With reduced activity of these enzymes, it produces inhibition of normal glucose metabolism, causing damage to certain areas of the brain.[4] Wernicke syndrome is characterized clinically by the triad of mental confusion, ocular-motor dysfunction and ataxia. Nevertheless, the presence of this triad is more of an exception than a rule and the conjunction of the three symptoms is in a third of patients. Confusion is the most frequent presenting symptom, followed by ataxia and ocular problems.[5] Our patient presented the classic triad, which greatly facilitated diagnosis.

Although the most frequent cause in our setting is chronic alcoholism, Wernicke Encephalopathy (WE) can be

caused by any other medical condition which produces malnutrition.[6] This includes prolonged administration of parenteral nutrition without the correct vitamin supplements, as occurred in our case.[2] While laboratory and image studies can be useful, EW diagnosis is fundamentally clinical and often it is the clinical response to treatment that confirms the suspected diagnosis.[7] The sensitivity and specificity of laboratory tests in symptomatic patients is uncertain. There usefulness is questionable due to the fact that a finding of normal thiamine does not exclude a diagnosis of WE and that in the majority of healthcare centers it is not possible to obtain results immediately, requiring treatment to be administered empirically on suspicion of the syndrome.[8] In this case, confronted with the suspicion of WE and the impossibility of requesting thiamine levels in advance, establishment of opportune treatment proceeded, obtaining said levels, which were normal, at a later stage.

Imaging studies are also unnecessary in all suspected WE patients and the initiation of treatment should not be delayed.[9] However, imaging diagnosis can be useful in providing evidence of WE and help eliminate possible alternative diagnoses. CT scan can show small, symmetrical signs of low density in the diencephalon, midbrain and periventricular regions which are enhanced after the administration of contrast. Nevertheless, this is considered an insensitive test because, as in our case, a negative result does not exclude WE.[10] MRI is the image study of choice for diagnosis, but although sensitivity is somewhat better, it is estimated to be just 53% with the highest specificity (93%).[11] The most common findings include areas of increased T2 and diminished T1 signs, localized in the third ventricle, periventricular region, medial thalamus and mammillary bodies, in addition to an alteration in the diffusion signal in said zones.[12] The abnormal T2 sign, however, can disappear in the first 48 hours after treatment with thiamine.[13] In our case, given that by the time it was possible to undertake an MRI more than 48 hours had elapsed, and that the patient was responding well to treatment, the test was not carried out.

In all those patients with clinically suspected WE, thiamine should immediately be administered intravenously prior to administration of glucose. Although different therapeutic options exist, it is accepted that the treatment should consist of 250-500 mg three times per day for the first 3-5 days, and if there is an improvement, 250 mg iv once per day for the next 3-5 days, moving finally to 100 mg orally en those patients who tolerate it.[9,14] Although sometimes incomplete, in general the response to treatment is rapid, with ophthalmoparesis being the first symptom tor resolve, followed by a partial or total improvement of confusional syndrome, with a certain degree of ataxia persisting in up to 50% of cases,[15] as occurred with our patient. She recovered as well from the confusional syndrome as from the diplopia in less than 24 hours, remaining with a certain degree of residual ataxia and gait instability.

There are various published cases in the literature which are linked the use of parenteral nutrition to WE.[2,16,17] Given that the majority of patients for whom TPN is indicated have no other way of acquiring said vitamin, its deficiency is due to the lack of supplementation of the same in the parenteral nutrition bag. The recommended dose of thiamine for a healthy adult is 1,4 mg per day or 0,5 mg per 1000 kcal consumed. It is calculated that the reserve of thiamine in the body is approximately 30 g, from which we can calculate that the symptoms will appear after 18-20 days of exogenous non-availability of the vitamin.[18]

CONCLUSION

In conclusion, we consider the supplementation of thiamine and other vitamins in patients with TPN to be an absolute necessity. It is important to come to the suspicion of this syndrome in those patients when they present compatible symptoms, with the objective of establishing treatment with thiamine as soon as possible. Likewise, with thiamine included in a vitamin complex, it is common to find other deficiencies which must also be ruled out.

REFERENCES

1. Palm E, Dotson B. Copper and zinc deficiency in a patient receiving long-term parenteral nutrition during a shortage of parenteral trace element products. *J Parenter Enteral Nutr*. 2014. doi: 10.1177/0148607114549255

2. Francini-Pesenti F, Brocadello F, Manara R, et al. Wernicke's syndrome during parenteral feeding: not an unusual complication. *Nutrition*. 2009; 25(2): 142-146. doi: 10.1016/j.nut.2008.08.003

3. Naidoo DP, Bramdev A, Cooper K. Autopsy prevalence of Wernicke's encephalopathy in alcohol-related disease. *S Afr Med J*. 1996; 86(9): 1110-1112.

4. Hazell AS, Butterworth RF. Update of cell damage mechanisms in thiamine deficiency: focus on oxidative stress, excitotoxicity and inflammation. *Alcohol*. 2009; 44(2): 141-147.

5. 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(4): 341-345.

6. Ogershok PR, Rahman A, Nestor S, et al. Wernicke encephalopathy in nonalcoholic patients. *Am J Med Sci*. 2002; 323(2): 107-111.

7. Sechi G, Serra A. Wernicke's encephalopathy: new clinical settings and recent advances in diagnosis and management. *Lancet Neurol*. 2007; 6(5): 442-455. doi: http://dx.doi.org/10.1016/S1474-4422(07)70104-7

8. Davies SB, Joshua FF, Zagami AS. Wernicke's encephalopathy in a non-alcoholic patient with a normal blood thiamine level. *Med J Aust*. 2011; 194(9): 483-484.

9. Latt N, Dore G. Thiamine in the treatment of Wernicke encephalopathy in patients with alcohol use disorders. *Intern Med J*. 2014; 44(9): 911-915. doi: 10.1111/imj.12522

10. Cerase A, Rubenni E, Rufa A, et al. CT and MRI of Wernicke's encephalopathy. *Radiol Med*. 2011; 116(2): 319-333. doi: 10.1007/s11547-011-0618-x

11. Antunez E, Estruch R, Cardenal C, et al. Usefulness of CT and MR imaging in the diagnosis of acute Wernicke's encephalopathy. *Am J Roentgenol*. 1998; 171(4): 1131-1137.

12. Zuccoli G, Pipitone N. Neuroimaging findings in acute Wernicke's encephalopathy: review of the literature. *Am J Roentgenol*. 2009; 192(2): 501-508. doi: 10.2214/AJR.07.3959

13. Park SH, Kim M, Na DL, et al. Magnetic resonance reflects the pathological evolution of Wernicke encephalopathy. *J Neuroimaging*. 2001; 11(4): 406-411.

14. Thomson AD, Cook CC, Touquet R, et al. The Royal College of Physicians report on alcohol: guidelines for managing Wernicke's encephalopathy in the accident and Emergency Department . *Alcohol*. 2002; 37(6): 513-521. doi: http://dx.doi.org/10.1093/alcalc/37.6.513 513-521

15. Gascón-Bayarri J, Campdelacreua J, García-Carreira MC, et al. Wernicke´s encephalopathy in non-alcoholic patients: a serie of 8 cases. *Neurología*. 2011; 26(9): 540-547. doi: 10.1016/j.nrl.2011.03.001

16. Sequeira Lopes da Silva JT, Almaraz Velarde R, Olgado Ferrero F, et al. Wernicke's encephalopathy induced by total parental nutrition. *Nutr Hosp*. 2010; 25(6): 1034-1036.

17. Francini-Pesenti F, Brocadello F, Famengo S, et al. Wernicke's encephalopathy during parenteral nutrition. *J Parenter Enteral Nutr*. 2007; 31(1): 69-71. doi: 10.1177/014860710703100169

18. Tan GH, Farnell GF, Hensrud DD, et al. Acute Wernicke's encephalopathy attributable to pure dietary thiamine deficiency. *Mayo Clin Proc*. 1994; 69(9): 849-850. doi: http://dx.doi.org/10.1016/S0025-6196(12)61786-8