ECRONICON OPEN ACCESS

EC NEUROLOGY
Case Report

Presentation of a Case of Dry Beri-Beri in our Hospital and the Electromyographic-EMG-ENG Correlation

A Molina León* and C Tudor Franco

Clinical Neurophysiology Service, University Hospital of Santa Lucia, Cartagena, Murcia, Spain

***Corresponding Author:** A Molina León, Clinical Neurophysiology Service, University Hospital of Santa Lucia, Cartagena, Murcia, Spain.

Received: November 03, 2021; **Published:** December 31, 2021

Abstract

This article presents the case of a young woman with a case of Wernicke's encephalopathy and polyneuropathy associated with Beri-Beri, vitamin B deficiency and we focus on the electroneurographic and electromyographic description of the case and its differences with the AIDP. Guillen Barre syndrome. In this patient, the presence of an axonal involvement, not demyelinating, of sensory and motor nerves of predominance in MMII of asymmetric distribution and acute course was clearly verified. If polyneuropathy is associated with encephalopathy, the picture known as Wernicke's syndrome and polyneuropathy due to thiamine deficiency occurs [2,3,5,6]. This patient was seen by us in January and then in October 8 months later ostensibly improving sensory and motor neurography.

His article presents the case of a young woman with a case of Wernicke encephalopathy and polyneuropathy associated with beriberi, vitamin B deficiency, and we focus on the electroneurographic and electromyographic description of the case and its differences with the AIDP. Guillén Barre syndrome. In this patient, the presence of non-demyelinating axonal involvement of sensory and motor nerves, predominantly in MMII of asymmetric distribution and of acute course, was clearly verified. If the polyneuropathy is associated with encephalopathy, the condition known as Wernicke syndrome and thiamine deficiency polyneuropathy occurs. This patient was seen 8 months later with marked improvement in sensory and motor neurography.

***Keywords**: Polyneuropathy; Beri-Beri; Thiamine Deficiency; Axonal Lesion; EMG; TIQA MINA; PYRIDOXINE*

Figure a

Introduction

Some vitamin deficiencies in the countries of the first world are almost eradicated or are very difficult to see or are subject to diseases of the digestive tract, however Wernicke's syndrome and pernicious anemia are more frequent, in this review we will see the case of a patient seen with the symptomatology in a high degree (tetraparesis with arreflexia and sd confusional) and as after 8 months the symptomatology improves markedly with an adequate treatment. Thiamine is the precursor of the most active form thiamine pyrophosphate, if it is associated as in this case an encephalopathy to the presence of a polyneuropathy produces the clinical picture that is called Wernicke's encephalopathy due to thiamine deficiency [2,3,5,6].

Presentation of the clinical case

A 28-year-old woman from Guinea Bisau, admitted from December 4 to 12, 2020 for hyperemesis gravidarum, of more than one month of evolution, then was admitted again from December 16 to 30, 2020, for persistence of vomiting with metabolic acidosis with elevated GAP anion (lactate, ketonemia) and severe hypokalemia. Gastroscopy was performed and targeted (peptic esophagitis, B and petechial hemorrhagic gastritis in fundus, in relation to repeated vomiting).

New admission in January 2021 for confusional syndrome and weakness. With hyperemesis gravidarum with fetal death at week 20+3 with curettage for fetal remains on 24/12/2020 being discharged on 30/12/2020. The husband reports that one afternoon he begins to be more drowsy and finally the next day he fails to wake her up or mobilize. She is taken to the emergency room with pulmonary CT angiography, simple CT and normal lumbar puncture. It presents confusional syndrome, weakness, a tetraparesis of predominance in MMII with arreflexia. Acute polyneuropathy secondary to acute thiamine deficiency vs GBS is suspected. The patient is re-entered on 06.01.21 due to confusional symptoms and areflectic tetraparesis suggestive of Wernicke's syndrome, with improvement after thiamine of the level of consciousness, associated with acute polyneuropathy (secondary to thiamine deficiency vs GBS). Possible hemilitical anemias of unaided cause. SB: Independent for daily activities, cognitive preserved.

Personal records

No known drug allergies. No HTN, NO DM, no dyslipidemia. Study of negative autoimmunity and negative serology.

Admitted from December 4-14, 2020 for hyperemesis gravidarum of one month of evolution with frequent daily vomits, nausea, very frequent vomits with metabolic acidosis, with elevated GAP anion. Re-entry on 06.012021 by cuonfusinal frame with tetraparesis and arreflexia. it is then when we perform the electromyogram in our laboratory.

Analitica

January 2021: the notable increase in GAMMA GT in 164 (normal value 6-42 u/l.) increase in GPT, and increase in GOT, presented anemia, with low levels of red blood cells (3.6), decrease in hemoglobin, decrease in hematocrit, normal white series, increase in ESR: 69, normal values of 0-20.

Brain MRI 12/01/2021

Hypersignal FLAIR T2 (fluid -attenuated inversion recovery) of pulvinar and bilateral postero-medial thalamus. No other significant signal alterations in other structures. At least three hypersignal foci are identified in DWI, without clear correlation in ADC or in other sequences, of right precentral and paracentral frontal cortical location, which impress of artifact. Ventricular system, cerebral grooves and cisternal spaces of normal characteristics. Normal Turkish chair. The study with contrast is of very limited assessment by movement, not optimal for diagnosis. The FLAIR T2 sequence with coronal contrast but also artefacted shows no evident meningeal enhancement.

Conclusion: Brain study with findings compatible with Wernicke medial, pulvinar and central thalamus lesions.

Brain MRI control 21/1/21

With respect to the previous study, lower FLAIR-T2 signal of the affected structures, in relation to possible Wernicke, corresponding to radiological improvement and according to the clinical improvement referred. No new lesions are seen

Analytical 12.03.21: Vitamins

- 25-hydroxy vitamin D (D3+D2) * 17.3 ug/l
- Sufficiency: 30-100 ug/L
- Mild insufficiency: 20-30 ug/L
- Moderate insufficiency: 10-20 ug/L
- Deficiencia: <10 ug/L
- Toxicidad: >100 ug/L.

Negative tumor markers.

Anemia control 12.07.21

- Hierro 60 ug/dL 33 - 193
- Folato * 2.6 ng/ml 3.8 - 16.0
- Severe deficit: < 2.7 ng/mL

Result interfered with by hamolysis

- Vitamin B12 542 pg/ml 200 - 770

Clinic

Last admission from December 16 to 30, 2020 for persistence of vomiting with metabolic acidosis with elevated anion gap (lactate, ketonemia) and severe hypokalemia. Medical treatment was performed for gestational loss of second trimester and subsequent evacuation curettage for retention of placental remains on 12/25/20 that occurs without incident. The postoperative period was satisfactorily completed.

Gastroscopy (peptic esophagitis B and hemorrhagic petechial gastritis in fundus, in relation to repeated vomiting), PPIs are indicated.

Abdominal ultrasound

Gallbladder filled with bile mud, with no suggestive signs of acute cholecystitis. Hydronephrosis grade II in right kidney and left grade I, without observing obstructive lithiasis (probably in the context of gestation).

Exploration NRL 11/01/2021

Slight drowsiness. Obey simple commands. It is intoxicated by successive orders. He says his first and last name in a hypophonic voice, without being able to determine if there is dysarthria. MOE: Bilateral pair of Ophltamoparesis VI with endotropia of the left eye, horizontal nystagmus of both eyes. Medium and reactive MOI (cataract carrier), bilateral facial of right predominance, bucinator weakness, with preserved masseter, preserved cervical extension. MMSS motor balance: slightly diminished tone, maintains against gravity for a few seconds, with predominance of proximal weakness 3/-4 over 5 and distal 5/5 MMII: mobilizes in plane with maintained tone, proximal 2/5 distal 3/5 pectoral ROT + bicipital + tricipital + absent styradial, absent patellar, absent aquileo, sensitivity: normal tactile, normal postural, maintained vibratory, algesic maintained.

Diagnoses

- Confusional syndrome and arreflectic tetraparesis in relation to Wernicke's encephalopathy and polyneuropathy
- Moderate-severe axonal motor sensory secondary to thiamine deficiency, improving after replacement therapy and immunoglobulins
- Febrile syndrome without clear focus, probably secondary to systemic symptoms
- Coombs anemia due to consumption secondary to thiamine deficiency
- Reactive depressive picture with psychotic symptoms in improvement
- Hypertrasaminemia, probably pharmacological
- SB: Independent for daily activities, cognitive preserved

Current treatment does not take aripiprazole, mirtazapine, neoxaporin, or pantoprazole.

Thiamine (vit B1) 300 mg daily, pyrodyxine (vit B6) 1 tablet in esayuno, acfol 5 mg, bisoprololo 2.5 mg 1/24 hours.

Lumbar puncture normal.

EMG 20/1/21

Pathological we describe it below.

RNM: Pathological

Conclusion: Brain study with findings compatible with Wernicke [1,19].

EEG 12/01/2021

Within Normal limits for the patient's age. Background EEG tracing devoid of specific anomalies assessable for epilepsy. No electrical data of epileptic status. The patient was stuporous, not responding to simple commands. Mild encephalopatia with a preserved alpha rhythm pattern.

EMG 14/1/21

Chronic sensory polyneuropathy, of axonal predominance, without data of active denervation.

EMG control 20/1/21

Sensory polyneuropathy - axonal motor of distal gradient of thick fiber, of moderate-severe degree, of predominance in lower limbs, in acute stage in which signs of acute axonal degeneration begin to be registered in the form of fibrillations and positive waves (+/++++) at the level of proximal and distal musculature of MMII.

Main diagnosis

- Confusional syndrome and arreflectic tetraparesis in relation to Wernicke's encephalopathy and polyneuropathy.
- Moderate-severe axonal motor-sensory secondary to thiamine deficiency, improving after replacement treatment and immunoglobulins
- Febrile syndrome without clear focus, probably secondary to systemic symptoms
- Coombs anemia due to consumption secondary to thiamine deficiency
- Reactive depressive picture with psychotic symptoms in improvement
- Hypertrasaminemia, probably pharmacological.

Electromyography: Description of the EMG.

The EMG is performed on 20.01.21 with the following results:

MMSS DCHO:

- Radial sensitive DCHO. (SNAP) normal.
- Ulnar sensitive DCHO. (SNAP) normal.
- Medium sensitive DCHO. (SNAP) slight delay of latencies and associated with a mild CTS.
- Medium engine DCHO. (CMAP) normal. F of the normal median.

MMII DCHO and left

Sensitive superficial peroneum (SNAP) right and left. marked drop in amplitude, bilateral and symmetrical, with preservation of latencies and VCS.

N. sural dcho. (SNAP) marked drop in amplitude, bilateral and symmetrical, with preservation of latencies and VCS.

- N. ciatico popliteo externa DCHO: CPE DCHO motor. (CMAP) at the pedio level: No motor potential is obtained.
- N. ciatico popliteo interno DCHO: CPI motor DCHO. (CMAP): marked drop in amplitude, Not blocking of motor conduction. Preserved latencies, and conservation of VAW. Normal CPI F wave. Normal H soleo reflex.

- N. ciatico popliteo externa IZQ: CPE motor (CMAP) at the level of order: fall of amplitude, and preservation of latencies and VCM. Normal f wave.
- N. ciatico popliteo interno IZQ: CPI motor (CMAP): Normal. No blocking of motor driving, preserved latencies, and conservation of Vaw. Normal CPI F wave. Normal H soleo reflex.

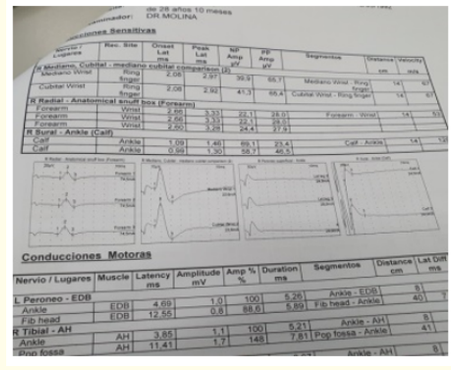

Figure 1

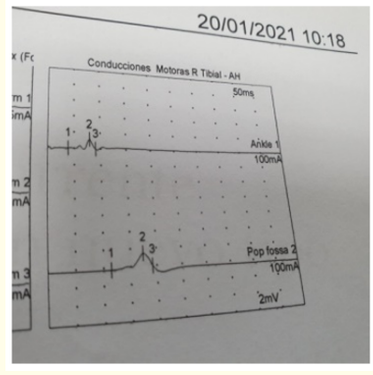

Figure 2

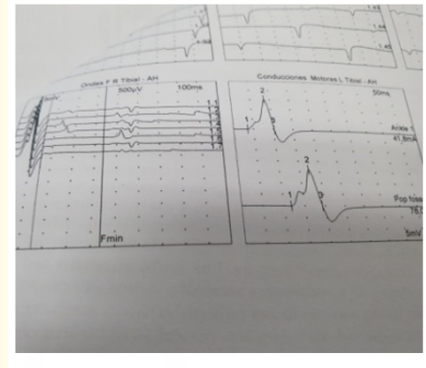

Figure 3

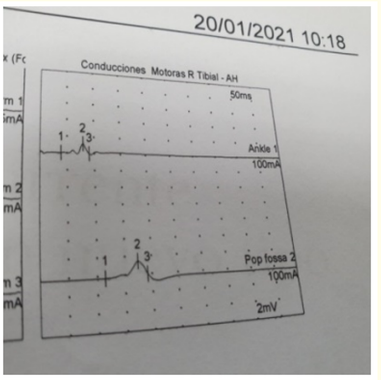

Figure 4

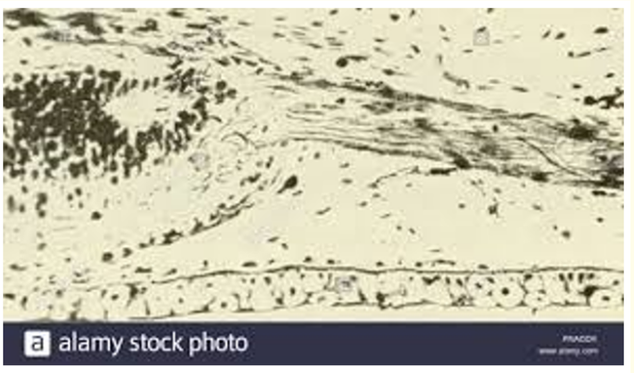

Figure 5

The known deterioration of nerve growth in thiamine deficiency (beri-beri) is observed only if the cell bodies are bathed by the deficient medium (*in vitro*), although the rest of the nerve, including the growth tips, is in normal medium (Burt, '43a).

EMG

Resting denervative activity is detected in the form of fibrillations and positive waves (++/++++) in proximal and distal musculature of both MMII (ILIAC PSOAS, anterior rectum, vastus lateral, anteroior tibials and both geme. LOS) denervative activity is also detected, in a smaller amount (+/++++) in MMSS at the level of dorsal intersoeo I dcho. And biceps dcho.

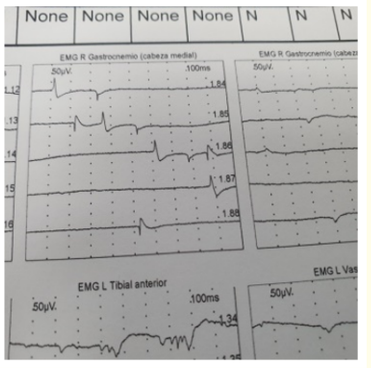

Figure 6

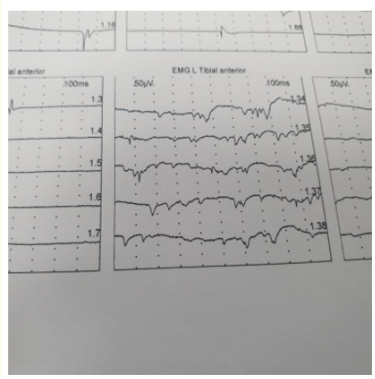

Figure 7

Conclusion

JANUARY 2021: Sensory-motor axonal polyneuropathy of distal gradient, of coarse fiber, of moderate-severe degree most affected the MMII, in acute-subacute stage (10-30 days) at the level of distal and proximal musculature of both MMII and to a lesser extent in MMSS. A picture that given tetraparesis and arreflexia can clinically resemble a Guillan syndrome sweeps-AIDP, the polyneuropathy found is axonal, acute, sensory-motor within the context of a dry Beri-Beri. Indicative of axonal fiber degeneration secondary to sustained Thiamine deficiency (vit B1) AidP in addition to being acute is inflammatory demyelinating unlike what was found in this patient.

Peripheral polyneuropathy is common in vitamin B1 thiamine deficiency, with a similar rate between motor and sensory neuropathy, usually with greater involvement of both MMII [1,19,28] as occurred with our patient.

The electroneurogrqafria associated with electromyography (EMG) is the test of choice for the diagnosis of polyneuropathy due to nutritional deficiency [4,12,13,15,19]. finding a diffuse axonal neuropathy with sensory and motor involvement, acute, subacute, which more often produces greater involvement of lower limbs than of upper limbs, as we could see in our study and in reviewed studies [1,4,11-15,19].

Within 48 hours after starting thiamine there was progressive recovery of consciousness with spontaneous eye opening but with persistence of flaccid tetraparesis and arreflexia, 72 hours after starting treatment the patient began to perform spontaneous movements, with facial response to painful stimuli and progressive mobilization of limbs and the patient evolved slowly but progressively. The findings found in the EMG coincide with other similar studies and articles such as the one described by Carvajal and collaborators in their article, Wernicke's encephalopathy and polyneuropathy associated with B-complex deficiency after bariatric surgery [1,6,13].

Control EMG October 2021

View of the patient 8 months later with a correct treatment of vitamin deficiency and rehabilitation it is verified: Current Datos from the neurographic point of view of a notable improvement of the acute sensory-motor polyneuropathy suffered by this patient giving way to the current normalization of the sensory nerves and motroes of MMSS and both MMII. If a chronic neurogenic pattern remains residual (more than 6 months) at the level of proximal muscles and is far from MMII (l3, l4, l5, s1, s2 bilateral) with absence of acute and subacute denervation.

Discussion of the case

Deficiencies of vitamin B1 (thiamine) and vitamin B12 (cyanocobalamin) are the most common causes of polyneuropathies due to malnutrition, gastric atrophy, gastritis, bariatric surgery, those caused by thiamine deficiency are called dry BERI-BERI. Thiamine or vitamin B1 found in unrefined cereals, wheat germ, yeast, soybean meal, and pork, thiamine, vitamin B1, acts as a coenzyme in the metabolism of carbohydrates, lipids, and branched-chain amino acids, also involved in the synthesis of myelin, also has an unidentified role in the initiation of nerve impulse propagation, possibly in cholinergic and serotonergic transmission through effect on the sodium channel [7,13]. Also polyneuropathy can be seen in patients with niacin (vit B3) and pyridoxine deficiency (vit B6 that give clinically similar pictures to Guillain barre syndrome, myasthenia gravis, and polyneuropathy associated with acute chronic hepatitis. Thiamine deficiency causes tissue damage, inhibiting metabolism in vulnerable brain regions, which are those with the highest metabolic activity and highest thiamine reserve [12,13,15,16], after 3 weeks of thiamine deficiency, serum levels drop and lead enzymes that require thiamine pyrophosphate to decrease their activity, producing accumulation of toxic intermediates such as lactic acid – acidosis, which causes tissue damage. The decrease of the enzyme transcetolase in astrocytes begins a week after deficiency so it increases the production of oxygen free radicals that causes osmotic tissue damage and edema. The increase in lactic acid leads to a situation of persistent metabolic acidosis that perpetuates toxic damage to tissues [1]. The permeability of the cerebral capillaries is altered producing hemorrhagic areas and cellular edema. Macrophage infiltration and DNA fragmentation are observed in the thalamus and mammillary bodies, (in this patient the presence of acute axonal degeneration through EMG was confirmed in January 2021, confirming the presence of fibrillations and positive waves (++/++++) at the level of proximal and distal musculature of both MMII. And also lesions in medial, pulvinar and central thalamus. There is also cellular apoptosis two weeks after thiamine deficiency, the clinical picture can be reversible up to this point, if the repositioning of thiamine is managed as a priority, in order to reduce lactate, acidosis and tissue injury triggered with decrease in the production of amino acids, a prolonged delay in time can lead to permanent neurological sequelae [1,12-15,17,18].

The demand for thiamine pyrophosphate depends on glucose consumption; This venous route could lead to severe pathology in patients with latent thiamine deficiency (necessary for glucose oxidation).

The most frequent cause is alcoholism, because in these patients inadequate intake, reduced gastrointestinal absorption, reduced conversion to active metabolites, increases in demand for carbohydrate metabolism and reduced liver storage are joined. Other causes are basic malnutrition due to the origin of an underdeveloped country, repeated vomiting, this patient vomited on the order of 10-20 times a day during her admission, can also be seen redeemed by serious systemic diseases, bariatric surgery, antacid drugs, prolonged parenteral feeding, pregnancy 8 in this case this woman was pregnant, although she lost her child), critical diseases, paraneoplastic syndromes, and treatments for quimioterapia.la Wernicke's encephalopathy is one of the neurological alterations that can produce thiamine deficiency (b1) that although known as a disease attributed only to Alcoholics, are not only produced by this cause [9,10,12,15].

The deficiency of thiamine causes an aumneto of the lactic acid, and deficirt of the capatacion of cellular oxygen, also the matabolismo of the glucose at the cellular level is diminished, the nervous system is the most exposed (alteration of the osmotic gradients, accumulation of glutamic acid, alteration of the permeability of the hematoencephalic membrane. In this case due to dry Beri-Beri there was a sensory neuropathy- axonal motroa [7,10] in Wewrnicke syndrome in just over half of the patients are seen in the rnm in T2 and FLAIR shows hyperintense thalamic periacueductal and roof signals of the IV ventricle, in the MRI of this patient comaptible signs were seen with Wernicke's sd. both the lesions at the level of the CNS and the SNP level improved in this patient ostensibly remaining residual at the level of EMG, a chronic neurogenic pattern (more than 6 months) at the level of proximal musculature and far from MMII (l3, l4, l5, s1, s2 bilateral) with absence of acute and subacute denervation and a slightly ataxic gait.

Figure 8

Bibliography

1. Carvajal., *et al*. "Wernicke's encephalopathy and polyneuropathy associated with B-complex deficiency after bariatric surgery". 1 (2012).

2. Ba F and Siddiqi ZA. "Neurologic complications of bariatric surgery". *Reviews in Neurological Diseases* 7 (2010): 119-124.

3. Pournaras D and le Roux C. "After bariatric surgery, what vitamins should be measured and what supplements should be given?". *Clinical Endocrinology71* (2009): 322-325.

4. Priya S., *et al*. "Neurologic Manifestations of Nutritional Disorders". 15 (2014): 273-290.

5. Thaisetthawatkul P., *et al*. "Good nutritional control may prevent polyneuropathy after bariatric surgery". *Muscle & Nerve* 42 (2010): 709-714.

6. Ziegler O., *et al*. "Medical follow up after bariatric surgery: Nutritional and drug issues. General recommendations for the prevention and treatment of nutritional deficiencies". *Diabetes & Metabolism* 35 (2009): 544-57.

7. Reynolds EH . "The neurology of folic acid deficiency". *Handbook of Clinical Neurology* 120 (2014): 927.

8. Donnino M., *et al*. "Myths and misconceptions of Wernicke's encephalopathy: What every emergency physician should know". *Annals of Emergency Medicine* 50 (2007): 715-721.

9. Cirignotta F., *et al*. "Wernicke-Korsakoff encephalopathy and polyneuropathy after gastroplasty for morbid obesity: Report of a case". *Archives of neurology* 57 (2000): 1356-1359.

10. Huertas-González N., *et al*. "Wernicke's Encephalopathy, Wet Beriberi, and Polyneuropathy in a Patient with Folate and Thiamine Deficiency Related to Gastric Phytobezoar". *Case Reports in Neurological Medicine* (2015).

11. Singh S and Kumar A. "Wernicke encephalopathy after obesity surgery: A systematic review". *Neurology* 68 (2007): 807-811.

12. Aluka KJ., *et al*. "Guillain-Barré syndrome and postbariatric surgery polyneuropathies". *JSLS* 13 (2009): 250-253.

13. Koike H., *et al*. "Rapidly developing weakness mimicking Guillain-Barré syndrome in beriberi neuropathy: Two case reports". *Nutrition* 24 (2008): 776-780.

14. Teitleman M and Katzka DA. "A case of polyneuropathy after gastric bypass surgery". *Archive of "Medscape General Medicine"* 7 (2005): 21.

15. Wilson RK., *et al*. "Wernicke's encephalopathy: Beyond alcoholism". *Nature Clinical Practice Neurology* 2 (2006): 54-58.

16. Sechi GP and Serra A. "Wernicke's encephalopathy: New clinical settings and recent advances in diagnosis and management". *The Lancet Neurology* 6 (2007): 442-455.

17. Fei GQ., *et al*. "Clinical characteristics and MR imaging features of nonalcoholic Wernicke encephalopathy". *American Journal of Neuroradiology* 29 (2008): 164-169.

18. Kumar V., *et al*. "The central nervous system". En: Robbins and Cotran pathologic basis of disease". (2009): 1328-1329.

19. Nakamura K., *et al*. "Polyneurophaty following gastric by pass surgery". *The American Journal of Medicine* 115 (2003): 679-680.

Volume 14 Issue 1 January 2022