

exacerbates neurological symptoms when used to treat uveitis.¹⁶ Tumour necrosis factor antagonists such as infliximab and etanercept are reserved for very aggressive forms of the disease, relapses after immunosuppressant treatment, or poor response to steroids.¹⁷ Venous sinus thrombosis is treated with antithrombotic drugs. Choice of drug and length of treatment are controversial topics.¹⁸

Regarding prognosis, patients tend to respond well to steroids, and only a third of them suffer relapses or progressive disease. Up to 20% of these patients have died at 7 years after diagnosis, which testifies to the severity of the disease.^{11,12}

In conclusion, BD should be included in the differential diagnosis of BME cases. BD is even more likely if BME is associated with mucocutaneous lesions (especially urogenital ulcers) or if the red nucleus is unaffected.

References

- Behçet H. Über residivierende, aphtöse durch ein Virus verursachtes Geschwüre am Mund, am Auge und an der Genitalien. *Derm Wschr.* 1937;105:1152–7.
- Yazici H. Behçet syndrome: an update. *Curr Rheumatol Rep.* 2003;5:195–219.
- Akman-Demir G, Serdaroglu P, Tasçi B. Clinical patterns of neurological involvement in Behçet's disease: evaluation of 200 patients. The Neuro-Behçet Study Group. *Brain.* 1999;122:2171–82.
- International Study Group for Behçet's Disease. Criteria for diagnosis of Behçet's disease. *Lancet.* 1990;335:1078–80.
- Mondéjar Marín B, García Montero R, López Ariztegui N, García Aparicio A. Meningoencefalitis recurrente como inicio de enfermedad de Behçet. *Neurología.* 2007;22:324–8.
- Yazıcı H, Fresko I, Yurdakul S. Behçet's syndrome: disease manifestations, management, and advances in treatment. *Nat Clin Pract Rheumatol.* 2007;3:151–5.
- Miranda G, Orellana P, Dellien H, Switt M. Rombencefalitis por *Listeria monocytogenes*. Patología emergente en relación al brote epidémico. Presentación de 3 casos clínicos. *Rev Med Chile.* 2009;137:1602–6.
- Moragas M, Martínez-Yélamos S, Majos C, Fernández-Viladrich P, Rubio F, Arbizu T. Rhombencephalitis. A series of 97 patients. *Medicine.* 2011;90:256–61.
- Kural-Seyahi E, Fresko I, Seyahi N, Ozyazgan Y, Mat C, Hamuryudan V, et al. The long-term mortality and morbidity of Behçet syndrome: a 2-decade outcome survey of 387 patients followed at a dedicated center. *Medicine.* 2003;82:60–76.
- Al-Araji A, Kidd DP. Neuro-Behçet's disease: epidemiology, clinical characteristics, and management. *Lancet Neurol.* 2009;8:192–204.
- Kidd D, Steuer A, Denman AM, Rudge P. Neurological complications of Behçet's syndrome. *Brain.* 1999;122:2183–94.
- Siva A, Kantarci OH, Saip S, Altintas A, Hamuryudan V, Islak C, et al. Behçet's disease: diagnostic and prognostic aspects of neurological involvement. *J Neurol.* 2001;248:95–103.
- Lueck CJ, Pires M, McCartney AC, Graham EM. Ocular and neurological Behçet's disease without orogenital ulceration? *J Neurol Neurosurg Psychiatry.* 1993;56:505–8.
- Park KS, Ko HJ, Yoon CH, Park SH, Cho CS, Kim HY, et al. Magnetic resonance imaging and proton magnetic resonance spectroscopy in neuro-Behçet disease. *Clin Exp Rheumatol.* 2004;22:561–7.
- Borhani Haghighi A, Pourmand R, Nikseresht AR. Neuro-Behçet disease: a review. *Neurologist.* 2005;11:80–9.
- Kotake S, Higashi K, Yoshikawa K, Sasamoto Y, Okamoto T, Matsuda H. Central nervous system symptoms in patients with Behçet disease receiving cyclosporine therapy. *Ophthalmology.* 1999;106:586–9.
- Abalos-Medina GM, Sánchez-Cano D, Ruiz-Villaverde G, Ruiz-Villaverde R, Quirosa Flores S, Raya Alvarez E. Successful use of infliximab in a patient with neuro-Behçet's disease. *Int J Rheum Dis.* 2009;12:264–6.
- Aguiar de Sousa D, Mestre T, Ferro JM. Cerebral venous thrombosis in Behçet's disease: a systematic review. *J Neurol.* 2011;258:719–27.

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Wernicke's encephalopathy secondary to hyperthyroidism and ingestion of thiaminase-rich products[☆]

Encefalopatía de Wernicke secundaria a hipertiroidismo e ingesta de productos ricos en tiaminasas

Wernicke's encephalopathy (WE) is a neurological disease caused by thiamine deficit. In some autopsy series, its

incidence rates range from 0.4% to 2.8%,¹ and prevalence is especially high among susceptible populations, including patients with alcoholism. It is estimated that 70% of all cases are not diagnosed until severe complications have appeared; the mortality rate approaches 17%.

Although alcoholism is its most frequent cause, WE has also been described in patients with malnutrition, hyperemesis, a history of bariatric surgery, cancer, and parenteral nutrition. It may not be detected in these situations, which can lead to serious complications.² This letter presents an exceptional case of WE secondary to hyperthyroidism and consumption of foods rich in thiaminase.

We present the clinical case of a 54-year-old woman referred to the Emergency Department with a 3-week history of asthenia and vomiting. During that time she had been ingesting little more than water and tea. The patient

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Figure 1 FLAIR MRI sequence with increased uptake in the corpus callosum.

reported episodic palpitations, heat intolerance, dyspnoea, and double vision that had appeared 3 days previously, in addition to gait instability. Personal history was anodyne; she lived alone and neither the patient nor family members reported alcohol abuse.

Upon her arrival at the Emergency Department, BP was 154/106 and heart rate was 150 bpm. The patient presented convergent strabismus with bilateral miosis, bilateral sixth cranial nerve palsy, and horizontal nystagmus, plus ataxia and abolished patellar reflexes. In the first hours following admission, as the patient was being treated with glucose solution, her level of consciousness decreased and the ataxia and ophthalmoparesis worsened. Complementary testing revealed the following values: K 2.8 mmol/L, Ca 11.72 mg/dL, GOT 193 U/L, GPT 199 U/L, PTH 9, TSH <0.05, T4L 2.01; antithyroid peroxidase AB >600; tumour markers and HIV, HVA, HVB, and HVC serology tests all negative. ECG showed supraventricular tachycardia with a rate of 140 bpm. Ultrasound revealed a thyroid of borderline size with no nodules. Echocardiography, abdominal ultrasound, gastroscopy, and brain and thoracic-abdominal CTs showed no significant findings. The MRI scan (Fig. 1) revealed deep periventricular alterations, demyelinating lesions in the white matter in predominantly frontal bilateral locations, and well-circumscribed plaques of demyelination in the corpus callosum and pons. All of these signs are compatible with WE.

The patient's clinical condition improved with parenteral thiamine plus metazolam and beta-blockers to correct thyroid alterations. Nystagmus and diplopia resolved and ataxia improved progressively; the patient achieved full recovery. Water–electrolyte imbalances, transaminase levels, and thyroid hormone levels all reverted to normal and the lesions visible in imaging studies resolved almost completely. Symptoms were compatible with WE due to hyperthyroid crisis and consumption of thiaminase-rich beverages.

WE consists of the classic triad of ophthalmoparesis with ataxia and confusional syndrome, although only a third of all patients present the 3 signs simultaneously. Altered mental

status presents in 82% of all patients, with manifestations that include disorientation, apathy, lethargy, confusion and agitation, memory impairment, and even stupor and coma. In contrast, ophthalmoparesis presents in 29% of cases and includes horizontal nystagmus, paralysis of the lateral rectus muscles with diplopia and internal strabismus, conjugate gaze palsy, ptosis in rare cases, and miosis in long-standing processes. Ataxia appears in 23% of cases with unstable tandem gait, short strides, and increased base of support. Other signs may include polyneuropathy as a manifestation of nutritional deficiency, and tachycardia with postural hypotension due to autonomous nervous system disorders or cardiac beriberi. Diagnosis is basically clinical, and the disease is reversible with administration of thiamine. MR imaging studies provide additional information with a low sensitivity of about 53% and a higher specificity of 93%. Studies show increased T2-weighted signal intensity in the periventricular area, mamillary bodies, and periaqueductal grey matter, as well as demyelinating lesions in the cortex and corpus callosum.³

Treatment consists of administering intravenous thiamine. Ophthalmoparesis was the first manifestation to improve (resolving over a few hours except for nystagmus), followed by ataxia within a few days (40% recovery) and state of consciousness (improving in 2–3 weeks). Our review of the literature yielded few cases of Wernicke's encephalopathy associated with or secondary to hyperthyroidism.^{4–9} Most cases were associated with hypercatabolism and increased nutritional needs. Thiaminases are known to be present in raw fish, shellfish, tea, and coffee. These enzymes act as natural thiamine antagonists and may have contributed to the development of WE in our patient, who had been consuming little more than tea in the preceding weeks.¹⁰

In conclusion, the case was diagnosed as WE secondary to hyperthyroidism that was probably exacerbated by the thiaminases present in tea. The case we present is interesting as it demonstrates that symptoms compatible with WE may develop without the classic antecedents of malnutrition or alcoholism.

References

- Galvin R, Brathen G, Ivashynka A, Hillbom M, Tanasescu R, Leone MA. EFNS guidelines for diagnosis, therapy and prevention of Wernicke encephalopathy. *Eur J Neurol*. 2010;17:1408–18.
- Gascón-Bayarri J, Campdelacreu J, García-Carreira MC, Estela J, Martínez S, Palasí A, et al. Encefalopatía de Wernicke en pacientes no alcohólicos: una serie de 8 casos. *Neurología*. 2011;26:540–7.
- Zuccoli G, Siddiqui N, Cravo I, Bailey A, Gallucci M, Harper CG. Neuroimaging findings in alcohol-related encephalopathies. *Am J Roentgenol*. 2010;195:1378–84.
- Wierzbicka-Chmiel J, Wierbicki K, Kajdaniuk D, Sedziak R, Marek B. Wernicke–Korsakoff syndrome in the course of thyrotoxicosis – a case report. *Endokrynol Pol*. 2011;62:178–80.
- Bonucchi J, Hassan I, Policeni B, Kaboli P. Thyrotoxicosis associated Wernicke's encephalopathy. *J Gen Intern Med*. 2008;23:106–9.
- Ohmori N, Tushima T, Sekine Y, Sato K, Shibagaki Y, Ijuchi S, et al. Gestational thyrotoxicosis with acute Wernicke encephalopathy: a case report. *Endocr J*. 1999;46:787–93.

7. Otsuka F, Tada K, Ogura T, Hayakawa N, Mimura Y, Yamauchi T, et al. Gestational thyrotoxicosis manifesting as wernicke encephalopathy: a case report. *Endocr J.* 1997;44:447–52.
 8. Millson CE, Harding K, Hillson RM. Wernike–Korsakoff syndrome due to hyperemesis gravidarum precipitated by thyrotoxicosis. *Postgrad Med J.* 1995;71:249–50.
 9. Enoch BA, Williams DM. An association between Wernicke's encephalopathy and thyrotoxicosis. *Postgrad Med J.* 1968;44: 923–4.
 10. Haas RH. Thiamin and the brain. *Annu Rev Nutr.* 1988;8: 483–515.
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