4. Shy ME, Siskind C, Swan ER, Krajewski KM, Doherty T, Fuerst DR, et al. CMT1X phenotypes represent loss of GJB1 gene function. Neurology. 2007;68:849—55.
5. Kleopa KA, Scherer SS. Molecular genetics of X-linked Charcot-Marie-Tooth disease. Neuromolecular Med. 2006;8:107—22.

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https://doi.org/10.1016/j.nrleng.2017.01.007
2173-5808/

Coeliac disease and neuromyelitis optica: a rare but possible association☆

Enfermedad celíaca y neuromielitis óptica: una rara pero posible relación

Dear Editor:

Coeliac disease (CD) is an enteropathy of autoimmune origin, triggered by the intake of foods containing gluten. The condition not only affects the digestive system, but also presents a broad clinical spectrum, including various neurological manifestations.[1] Neuromyelitis optica (NMO) is a chronic, inflammatory, autoimmune, demyelinating disease of the central nervous system, characterised by severe spinal cord and optic nerve involvement; it can be monophasic or display relapses and remissions, and is a cause of disability in young and adult patients.[2] Given the limited published information on the relationship between these 2 entities, we decided to report the present case.

Our patient is a 9-year-old boy with a family history of CD (mother and a maternal aunt) and personal history of foetal distress, meningitis during lactation, severe sensorineural hearing loss diagnosed at 4 years and treated with cochlear implants, mild intellectual disability, and CD diagnosed at the age of 5. Diagnosis was based on high levels of anti-transglutaminase IgA antibodies and anti-gliadin IgG and IgA antibodies, HLA-DQ2 expression, and compatible duodenal biopsy findings. He was admitted to hospital due to a 4-week history of subacute symptoms of lower limb pain and gait disorder with falls. During the examination the patient was alert and oriented, and displayed developmental delay with poverty of speech and dyslalias, visual deficit, and sensorineural hearing loss. He presented predominantly distal weakness of all 4 limbs, with generalised hyporeflexia, bilateral extensor plantar response, and no clear sensory alterations, although he did have difficulties completing the assessment. The patient displayed positive Romberg sign and difficulty toe and heel walking. The brain MRI scan was of little value due to the cochlear implants, but showed no significant alterations; a spinal MRI scan revealed increased signal intensity from C1 to T2, with patchy distribution and no contrast uptake (Fig. 1). We also requested analyses of lactate, ammonia, amino acids, and organic acids in the blood and urine; very long-chain fatty acids; phytanic, guanidinoacetic, and pristanic acids; alpha-fetoprotein; neuron-specific enolase; human chorionic gonadotropin; creatine; vitamins B_{12} and B_6; folic acid; and immunoglobulins. An autoimmunity test and viral serologic testing were also performed. All results were normal. CSF study results were as follows: 8 cells, proteins 68 mg/dL, glucose 60 mg/dL, and weakly positive oligoclonal bands. An electromyography study showed signs of demyelinating sensorimotor polyneuropathy. Visual evoked potentials revealed bilateral retrobulbar optic neuropathy. The study was expanded with tests for anti-NMO and anti-MOG antibodies, which yielded negative results. The patient was initially treated with immunoglobulins dosed at 0.4 g/kg/day, with no response; he subsequently received methylprednisolone at 1 g/day for 5 days, which achieved favourable outcomes. When interviewed, the mother admitted that the patient was not strictly following a gluten-free diet; in the absence of any other possible aetiology, NMO symptoms were attributed to CD. The patient was discharged and prescribed a tapered dose of the corticosteroid and a strict gluten-free diet. On 2 occasions over the following months, symptoms worsened in association with fever, and the patient was treated with corticosteroids, although radiology studies identified no new lesions; outcomes were excellent after resolution of the infectious symptoms.

Neurological complications are estimated to be present in 10% to 22% of cases of CD.[3,4] Reported manifestations include ataxia, epilepsy, myopathy, migraine, cognitive impairment, and, as observed in our patient, axonal or demyelinating peripheral neuropathy, sensorineural hearing loss, and demyelinating diseases.[1,5—7]

The association between NMO and CD has very rarely been reported in the literature.[8—13] Given that CD is relatively common, it would be difficult to demonstrate the cause of this relatively rare neurological syndrome. However, the excellent response to methylprednisolone and the introduction of a strict gluten-free diet suggest an inflammatory process in which gluten plays a fundamental role. Indeed, some authors have proposed that the pathophysiological mechanism may be an autoimmune process in which circulating IgG anti-neuronal

☆ Please cite this article as: Díaz Díaz A, Hervás García M, Muñoz García A, Romero Santana F, Pinar Sedeño G, García Rodríguez JR. Enfermedad celíaca y neuromielitis óptica: una rara pero posible relación. Neurología. 2019;34:547—549.

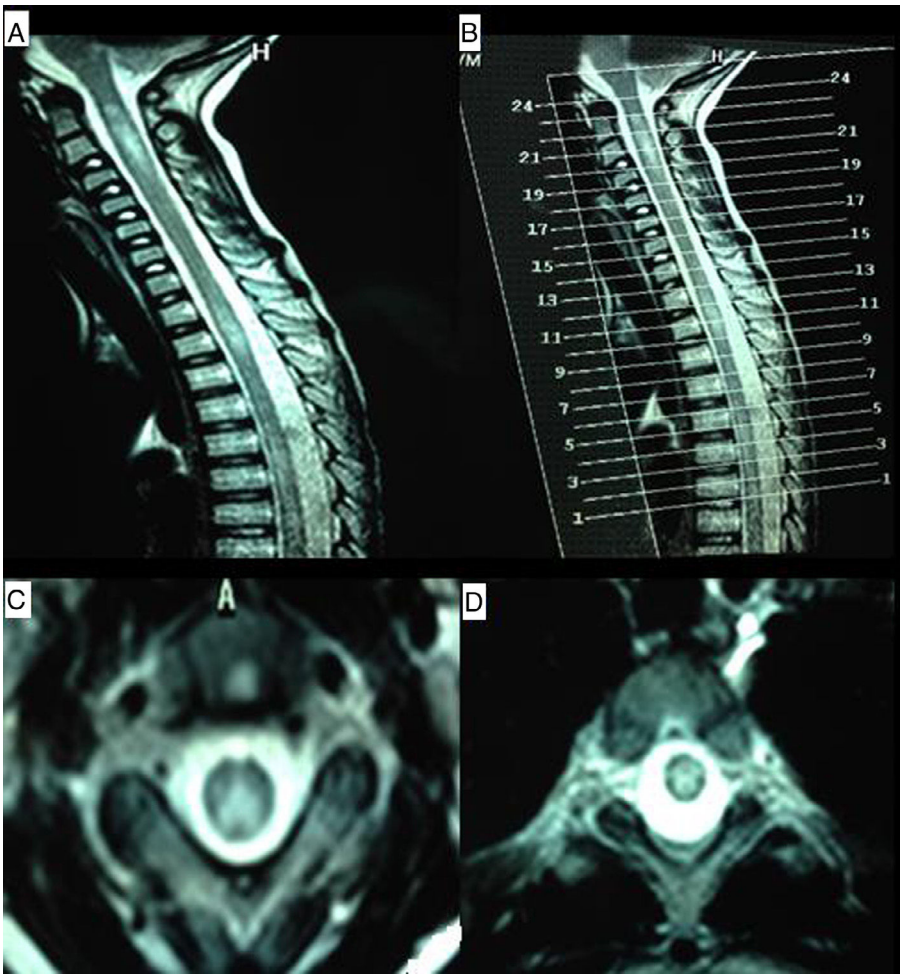

Figure 1 MR images of the cervical and thoracic spine. (A) Sagittal T2-weighted sequence showing hyperintense lesions affecting the C1-C4 and T2-T4 regions. (B) Reference image for axial slices. (C) Axial T2-weighted sequence (slice 21) showing a hyperintense lesion predominantly affecting posterolateral regions of the spinal cord. (D) Axial T2-weighted sequence (slice 11) revealing a centrospinal hyperintense lesion.

antibodies target antigens in the central and enteric nervous systems.[3,14]

Our case highlights the importance of knowing the possible association between CD and NMO, as a strict gluten-free diet may positively affect both diseases.

References

1. Hernández-Lahoz C, Mauri-Capdevila G, Vega-Villar J, Rodrigo L. Neurogluten: patología neurológica por intolerancia al gluten. Rev Neurol. 2011;53:287—300.
2. Carnero-Contentti E, Leguizamón F, Colla-Machado PE, Alonso R. Neuromielitis óptica: actualización clínica y terapéutica. Neurol Arg. 2013;5:259—69.
3. Caio G, de Giorgio R, Venturi A, Giancola F, Latorre R, Boschettin E, et al. Clinical and immunological relevance of anti-neuronal antibodies in celiac disease with neurological manifestations. Gastroenterol Hepatol Bed Bench. 2015;8:146—52.
4. Briani C, Zara G, Alaedini A, Grassivaro F, Ruggero S, Toffanin E, et al. Neurological complications of celiac disease and autoinmun mechanisms: a prospective study. J Neuroimmunol. 2008;195:171—5.
5. Zelnik N, Pacht A, Obeid R, Lerner A. Range of neurologic disorders in patients with celiac disease. Pediatrics. 2004;113:1672—6.
6. Solmaz F, Unal F, Apuhan T. Celiac disease and sensorineural hearing loss in children. Acta Otolaryngol. 2012;132:146—51.
7. Thawani SP, Brannagan TH 3rd, Lebwohl B, Green PH, Ludvigsson JF. Risk of neuropathy among 28 232 patients with biopsy-verified celiac disease. JAMA Neurol. 2015;72:806—11.
8. Jacob S, Zarei M, Kenton A, Allroggen H. Gluten sensitivity and neuromyelitis optica: two case reports. J Neurol Neurosurg Psychiatry. 2005;76:1028—30.
9. Jarius S, Jacob S, Waters P, Jacob A, Littleton E, Vincent A. Neuromyelitis optica in patients with gluten sensitivity associated with antibodies to aquaporin-4. J Neurol Neurosurg Psychiatry. 2008;79:1084.
10. Bergamaschi R, Jarius S, Robotti M, Pichiecchio B, Wildemann B, Meola G. Two cases of benign neuromyelitis optica in patients with celiac disease. J Neurol. 2009;256:2097—9.
11. Meyts I, Jansen K, Renard M, Bossuyt X, Roelens F, Regal L, et al. Neuromyelitis optica-IgG (+) optic neuritis associated with celiac disease and dysgammaglobulinemia: a role for tacrolimus? Eur J Paediatr Neurol. 2011;15:265—7.
12. McNamara PH, Costelloe L, Langan Y, Redmond J. Aquaporin-4 seropositivity in a patient with coeliac disease but normal

neurological examination and imaging. J Neurol. 2011;258: 702—3.
13. Matijaca M, Pavelin S, Kaliterna DM, Bojic L, Matijaca A. Pathogenic role of acuaporin antibodies in the development of neuromyelitis optica in a woman with celiac disease. Isr Med Assoc J. 2011;13:182—4.
14. Volta U, de Giorgio R, Petrolini N, Stanghellini V, Barbara G, Granito A, et al. Clinical findings and anti-neuronal antibodies in coeliac disease with neurological disorders. Scand J Gastroenterol. 2002;37:1276—81.

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https://doi.org/10.1016/j.nrleng.2018.12.007
2173-5808/

Posterior reversible leukoencephalopathy syndrome associated with psychosis: An unusual presentation☆

Síndrome de leucoencefalopatía posterior reversible asociado a psicosis: una presentación inusual

Dear Editor:

Posterior reversible leukoencephalopathy syndrome (PRES) is a clinical condition whose actual incidence is still unknown. It was first described in 1996 and may be observed in acute patients, a majority of whom present such signs and symptoms as headache, altered level of consciousness, seizures, and/or bilateral loss of vision (cortical blindness). Neuroimaging studies usually reveal posterior leukoencephalopathy.[1] This condition has been reported in the literature in association with various clinical entities, including hypertension, eclampsia, systemic neoplasms, kidney disease, sepsis, transplants, and immunosuppressive therapy.[2—4]

We present the case of a patient with postpartum eclampsia who developed acute psychotic symptoms and attempted suicide in the hospital. Imaging findings were characteristic of PRES, which disappeared after the clinical improvement which followed effective treatment for postpartum eclampsia.

Our patient was an 18-year-old pregnant woman (39.3 weeks) from Cali, Colombia, with no relevant history and no alterations detected in the prenatal checkups. She consulted the emergency department due to premature rupture of membranes of 2 hours' progression. An obstetric examination revealed that the foetus was in the breech position; 12 hours after admission the patient was taken to the operating theatre for caesarian delivery, with no complications for the mother or baby.

Fourteen hours after delivery, the patient reported intense holocranial headache with phosphenes; there were no signs of increased blood pressure. She subsequently presented a generalised tonic—clonic seizure lasting 1 minute, associated with sphincter relaxation; this resolved with endovenous administration of benzodiazepines. An emergency brain computed tomography (CT) scan revealed hypodensities in the white matter of both hemispheres, involving the temporal-occipital region bilaterally and mainly the left-sided basal ganglion region (Fig. 1). These signs are highly indicative of symmetrical bilateral posterior leukoencephalopathy.

Complementary tests detected proteinuria and an increased level of lactate dehydrogenase (LDH) (Table 1), suggesting possible eclampsia with no initial presence of

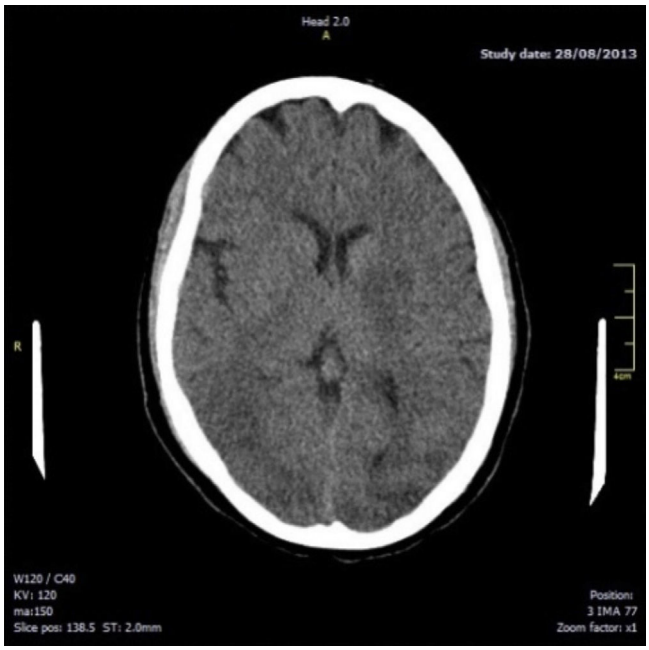

Figure 1 Simple brain CT showing changes suggestive of reversible posterior leukoencephalopathy syndrome.

☆ Please cite this article as: Díaz-Ramírez GS, Salgado-Cifuentes CA, Zúñiga-Escobar G, Morales-Plaza CD. Síndrome de leucoencefalopatía posterior reversible asociado a psicosis: una presentación inusual. Neurología. 2019;34:549—551.