

Paraneoplastic Anti-N-Methyl-D-Aspartic Acid Receptor Encephalitis

Chi Niu, Wei Guo

Emergency Department, Beijing Tiantan Hospital, Capital Medical University, Beijing 100050, China

To the Editor: Anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis is a severe autoimmune disease. Dalmau identified anti-hippocampal and prefrontal nerve cell membrane NMDAR antibodies in this patient population and first named this syndrome.^[1] Here, we describe a case of one patient with anti-NMDAR encephalitis accompanied by ovarian teratoma.

A 22-year-old woman presented at the Emergency Department, Beijing Tiantan Hospital, Beijing, China with a fever that had been accompanied by headache for 18 days, and an exacerbation accompanied by conscious disturbance for 1 day. Head computed tomography (CT) scan did not reveal any finding. Lumbar puncture indicated a cerebrospinal fluid (CSF) pressure of 245 mmH₂O (1 mmH₂O = 0.0098 kPa). Routine CSF analysis indicated a total cell count of $255 \times 10^6/L$, a white blood cell (WBC) count of $101 \times 10^6/L$. CSF biochemistry indicated a protein level of 8090 mg/dl. The diagnosis of viral encephalitis was made, followed by antiviral therapy. The patient was referred to our hospital for further treatment. On admission at our hospital, physical examination of the subject showed a temperature of 38.1°C, pulse rate of 120 beats/min, respiratory activity of 20 breaths/min. Nervous system physical examination showed the patient was in a moderate coma; both pupils were poorly responsive to light. The patient could not comply with an instruction to stick out her tongue and showed no physical reactions to a painful stimulus applied to the limbs. The muscle tension of limbs was reduced in the presence of bilateral tendon reflexes.

The emergency diagnosis was made as central nervous system infection with a possible viral, tuberculosis (TB), or autoimmune encephalitis. Rounds of antiviral, anti-TB, and dehydration treatments were administered. However, no improvement was noted. The lumbar puncture was repeated which indicated a CSF pressure of 140 mmH₂O. Routine CSF analysis indicated a total cell count of $404 \times 10^6/L$, a WBC count of $104 \times 10^6/L$. CSF biochemistry indicated acid-fast staining (–), TB-SPOT (–), and anti-NMDAR antibodies (+). Pelvic CT indicated the abnormal signal shadow in the right appendage. Rectal ultrasound indicated the masses in the right appendage. Based on these, a final diagnosis of anti-NMDAR encephalitis was made. Anti-TB and antiviral therapies were replaced with hormone and gamma globulin pulse therapy. A laparoscopic excisional surgery for the teratoma was

subsequently performed. The postoperative pathological findings confirmed the presence of a mature teratoma in the right ovary. The patient gradually regained consciousness following the combination therapy, with improved cognitive function and reduction in involuntary limb movements. She recovered after 2 months of treatment.

Anti-NMDAR encephalitis is an antibody-mediated autoimmune rare disease. The activation of NMDARs commences immune responses ultimately resulting in the production of anti-NMDAR antibodies, followed by the occurrence of anti-NMDAR-related clinical manifestations. Some tumors are related to anti-NMDAR encephalitis because of the expression of NMDAR. Even infection and genetic alterations may also contribute to the occurrence of anti-NMDAR encephalitis.^[2] At the nonspecific prodromal period, symptoms include headache, fever, nausea, and diarrhea. The prominent psychiatric period shows symptoms including anxiety, hallucination, restless, crankiness, or stereotyped behaviors. Multiple nervous injuries may occur with rapid progression of the disease. The most characteristic manifestations are the involuntary movements of the mouth, tongue, and face. Auxiliary examinations: (1) CSF analysis shows the WBC count and albumin quantification can be normal or slightly increased. (2) Neuroimaging can show normal results; however, abnormal signal changes in the cerebral cortex, limbic system, basal ganglia, cerebellum, and brain stem may be observed with the progression of disease. (3) The final diagnosis is made based on positive anti-NMDAR antibodies in CSF and blood.^[3] The surgical excision of tumor and immunoregulation therapy should be immediately administered as soon as the diagnosis is established. The early surgical excision of tumor is important for recovery or improvement of symptoms.^[4]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given consent for her

Address for correspondence: Dr. Wei Guo,

Emergency Department, Beijing Tiantan Hospital, Capital Medical University, Beijing 100050, China
E-Mail: guowei1010@126.com

Access this article online

Quick Response Code:



Website:
www.cmj.org

DOI:
10.4103/0366-6999.218020

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

© 2017 Chinese Medical Journal | Produced by Wolters Kluwer - Medknow

Received: 30-06-2017 **Edited by:** Li-Min Chen

How to cite this article: Niu C, Guo W. Paraneoplastic Anti-N-Methyl-D-Aspartic Acid Receptor Encephalitis. Chin Med J 2017;130:2765-6.

clinical information to be reported in the journal. The patient understands that her identity will be made to conceal.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Dalmau J, Tüzün E, Wu HY, Masjuan J, Rossi JE, Voloschin A, *et al.* Paraneoplastic anti-N-methyl-D-aspartate receptor encephalitis associated with ovarian teratoma. *Ann Neurol* 2007;61:25-36.
2. Kayser MS, Dalmau J. Anti-NMDA receptor encephalitis, autoimmunity, and psychosis. *Schizophr Res* 2016;176:36-40. doi: 10.1016/j.schres.2014.10.007.
3. Gresa-Arribas N, Titulaer MJ, Torrents A, Aguilar E, McCracken L, Leypoldt F, *et al.* Diagnosis and significance of antibody titers in anti-NMDA receptor encephalitis, a retrospective study. *Lancet Neurol* 2014;13:167-77. doi: 10.1016/S1474-4422(13)70282-5.
4. Mann AP, Grebenciucova E, Lukas RV. Anti-N-methyl-D-aspartate-receptor encephalitis: Diagnosis, optimal management, and challenges. *Ther Clin Risk Manag* 2014;10:517-25. doi: 10.2147/TCRM.S61967u.