

Anti-N-methyl-D-aspartate receptor encephalitis in a 17-year-old female patient with 3 years of follow-up

Jie-Ping Lu, Xiao-Kai Song, Huai-Yu Li, Guo-Ping Wang

Department of Neurology, The First Affiliated Hospital of USTC, Division of Life Sciences and Medicine, University of Science and Technology of China, Hefei, Anhui 230001, China.

To the Editor: Anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis is a potentially lethal autoimmune disease characterized by prominent psychiatric symptoms and seizures. It usually occurs in young female patients with ovarian teratomas.^[1] Here, we describe a severe case of anti-NMDAR encephalitis. The patient was originally misdiagnosed as psychosis and did not improve within the first 4 weeks of first-line immunotherapy. Although she did not continue to receive second-line immunotherapy, the patient still recovered well and had no recurrence or tumor observed during 3 years of follow-up.

A 17-year-old female patient was transferred from a mental health center to our hospital with psychiatric symptoms, behavioral changes, and seizures. Eleven days earlier, paranoia and behavioral changes of the patient obviously developed, followed by confusion, low-grade fever, and generalized seizures. She had no history of epilepsy or psychosis. On physical examination, the patient remained nonverbal and unresponsive to external stimulus. She demonstrated bilateral Babinski signs and no meningeal signs. Blood tests showed erythrocyte sedimentation rate 58 mm/h. Chest computed tomography suggested mild lung infection. Abdominal and pelvic color ultrasound and cranial magnetic resonance imaging were normal. Cerebrospinal fluid (CSF) analysis showed 15 nucleated cells/ μ L, protein of 0.40 g/L, glucose of 3.81 mmol/L, and chloride of 117.5 mmol/L. A 2-h video electroencephalogram showed nonspecific generalized slow activity without epileptic discharges.

The patient was initially suspected as viral encephalitis and received intravenous (IV) acyclovir and antiepileptic drugs. She manifested oro-lingual-facial dyskinesias and hypersalivation within 1 week of admission. The latter symptom was similar to “bubbles of a crab” covering her nose and mouth. NMDAR antibodies were detected in CSF and serum 4 days after admission. We treated the patient twice

with IV immunoglobulin (IVIG) 0.4 g/kg per day for 5 days at a 2-week interval and IV methylprednisolone 500 mg per day for 5 days. The seizures were under control, but the psychiatric symptoms were not improved. She had a modified Rankin Scale (mRS) score of 5 at 4 weeks after treatment. Then the patient was transferred to the rehabilitation hospital. The mRS score was 2 after 6 months. So far, the patient has been followed up for 3 years with no recurrence or tumor observed.

Our patient was misdiagnosed as a psychiatric disorder at the onset of the disease. Around 80% of patients with anti-NMDAR encephalitis present initially with psychiatric symptoms.^[2] Therefore, it is necessary to consider anti-NMDAR encephalitis in the differential diagnosis of patients with acute onset psychiatric symptoms.^[2] Oro-lingual-facial dyskinesias and hypersalivation were impressive in this case. The former may be easily misdiagnosed as seizures and is often a clue to the diagnosis of anti-NMDAR encephalitis.^[3] In particular, the latter symptom with “bubbles of a crab” covering her nose and mouth was rarely reported before.^[4] Around 60% of patients with anti-NMDAR encephalitis are detected with tumors.^[3] In this case, the tumor was not found during a 3-year follow-up period. However, she should continue to be followed up as ovarian teratoma may occur in patients with anti-NMDAR encephalitis through the years.^[5] The patients who fail first-line immunotherapy (steroids, IVIG, plasmapheresis) within the first 4 weeks continue to have a poor prognosis and second-line immunotherapy (rituximab, cyclophosphamide) can improve the outcome of those patients.^[1] Considering the potential side effects, the second-line immunotherapy was not subsequently performed in this case.

In summary, the probable anti-NMDAR encephalitis should be seriously considered when the patients originally present acute psychiatric symptoms. Although some

Access this article online

Quick Response Code:



Website:

www.cmj.org

DOI:

10.1097/CM9.0000000000000190

Correspondence to: Dr. Jie-Ping Lu, Department of Neurology, The First Affiliated Hospital of USTC, Division of Life Sciences and Medicine, University of Science and Technology of China, Hefei, Anhui 230001, China
E-Mail: Lujp1984@163.com

Copyright © 2019 The Chinese Medical Association, produced by Wolters Kluwer, Inc. under the CC-BY-NC-ND license. This is an open access article distributed under the terms of the Creative Commons Attribution-Non Commercial-No Derivatives License 4.0 (CCBY-NC-ND), where it is permissible to download and share the work provided it is properly cited. The work cannot be changed in any way or used commercially without permission from the journal.

Chinese Medical Journal 2019;132(8)

Received: 28-12-2018 Edited by: Li-Shao Guo

patients with anti-NMDAR encephalitis who initially fail first-line immunotherapy are not further treated with second-line immunotherapy, they may still recover well.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initial will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Conflicts of interest

None.

References

1. Titulaer MJ, McCracken L, Gabilondo I, Armangué T, Glaser C, Iizuka T, *et al.* Treatment and prognostic factors for long-term outcome in patients with anti-NMDA receptor encephalitis: an observational cohort study. *Lancet Neurol* 2013;12:157–165. doi: 10.1016/S1474-4422(12)70310-1.
2. Sarkis RA, Coffey MJ, Cooper JJ, Hassan I, Lennox B. Anti-N-methyl-D-aspartate receptor encephalitis: a review of psychiatric phenotypes and management considerations: a report of the American Neuropsychiatric Association Committee on Research. *J Neuropsychiatry Clin Neurosci* 2018; Epub ahead of print. doi: 10.1176/appi.neuropsych.18010005.
3. Barry H, Byrne S, Barrett E, Murphy KC, Cotter DR. Anti-N-methyl-D-aspartate receptor encephalitis: review of clinical presentation, diagnosis and treatment. *BJPsych Bull* 2015;39:19–23. doi: 10.1192/pb.bp.113.045518.
4. Taguchi Y, Takashima S, Nukui T, Tanaka K. Hypersalivation in a patient with anti-NMDAR encephalitis with ovarian teratoma. *Intern Med* 2010;49:803–804. doi: 10.2169/internalmedicine.49.3221.
5. Iizuka T, Sakai F, Ide T, Monzen T, Yoshii S, Iigaya M, *et al.* Anti-NMDA receptor encephalitis in Japan: long-term outcome without tumor removal. *Neurology* 2008;70:504–511. doi: 10.1212/01.wnl.0000278388.90370.c3.

How to cite this article: Lu JP, Song XK, Li HY, Wang GP. Anti-N-methyl-D-aspartate receptor encephalitis in a 17-year-old female patient with 3 years of follow-up. *Chin Med J* 2019;132:996–997. doi: 10.1097/CM9.0000000000000190

Corrigendum

Corrigendum: Association of pre-pregnancy body mass index and gestational weight gain with labor stage

In the article titled “Association of pre-pregnancy body mass index and gestational weight gain with labor stage,” published on pages 483–487, Issue 4, Volume 132 of *Chinese Medical Journal*,^[1] the affiliation of authors is written incorrectly as “Department of Perinatal Medicine, Beijing Obstetric and Gynecology Hospital, Capital Medical University. Beijing 100026, China” instead of “Department of Perinatal Medicine, Beijing Obstetrics and Gynecology Hospital, Capital Medical University. Beijing 100026, China,” and the name of funding is also written incorrectly as “the Capital Characteristics Project (Beijing Municipal Science and Technology Commission No.151100004015182)” instead of “Beijing Municipal Science and Technology Commission (No.Z151100004015182).”

Reference

1. Zhou L, Yang HX, Zhao RF, Zhang WY. Association of pre-pregnancy body mass index and gestational weight gain with labor stage. *Chin Med J* 2019;132:483–487. doi:10.1097/CM9.0000000000000093.

DOI:10.1097/CM9.0000000000000207