Neurol Sci (2015) 36:1257–1258
DOI 10.1007/s10072-014-1966-3

LETTER TO THE EDITOR

A case of NMDAR encephalitis misdiagnosed as postpartum psychosis and neuroleptic malignant syndrome

Ayhan Koksal · Sevim Baybas · Belgin Mutluay · Yavuz Altunkaynak · Aslı Keskek

Received: 11 May 2014 / Accepted: 23 September 2014 / Published online: 30 September 2014

Keywords Neuroleptic malignant syndrome · NMDAR antibody · NMDAR encephalitis · Ovarian teratoma · Postpartum psychosis

Dear Editor,

Anti-*N*-methyl-D-aspartic acid receptor (NMDAR) encephalitis is a rare and recently described disease that is characterized by a neuropsychiatric syndrome and associated with ovarian teratomas [1–3].

We report a 25-year-old female who presented with insomnia, agitation, irritability and fear of death which began 3 months after a normal delivery. Shortly after that, she developed delusions and hallucinations and psychomotor excitement. Her past medical history as well as that of her family's was unremarkable. She complained of seeing demons in her dream and believed that they were inside her body and would kill her. She was suspected of having postpartum psychosis and admitted to the psychiatry clinic. She was given risperidone 2 mg/day and benzodiazepine 10 mg/day treatment, but there was no improvement. One week later, she developed confusion, cog-wheel rigidity, fever, increase in serum creatine phosphokinase (CPK) levels, and leukocytosis. She was diagnosed as neuroleptic malignant syndrome and transferred to the neurology clinic. Peripheral leukocyte count and serum CPK levels were 18.000 $\times$ 10^3/μL and 8,500 IU/L, respectively. Neuroleptic treatment was stopped and bromocriptine 10 mg/day, biperiden 6 mg/day and benzodiazepine 10 mg/day treatment were given. Two weeks later, extrapyramidal rigidity resolved substantially and CPK values decreased gradually and she was sent back to the psychiatry clinic for further treatment. She was treated with aripiprazole 10 mg/day and two sessions of electroconvulsive therapy. After having a generalized seizure, she was transferred to the neurology clinic for the second time to investigate an organic etiology. Since her delusions and hallucinations continued and she wanted to strangulate herself and tried to jump from windows, she was tied to bed almost 24 h a day. The results of laboratory tests including paraneoplastic antibodies were within the normal range, and there was no marked elevation of antiviral antibody titers. Magnetic resonance imaging (MRI) of the brain with contrast and repeat EEG's was normal. Cerebrospinal fluid (CSF) analysis showed neither pleocytosis nor protein increase. Herpes simplex virus polymerase chain reaction was negative in the CSF. Although NMDAR antibody was negative in serum, it was positive in CSF and she was diagnosed as anti-NMDAR encephalitis. Abdominal ultrasound examination and MRI revealed a right ovarian cystic teratoma (Figs. 1, 2). She was treated with intravenous immunoglobulin (IVIG) at a dose of 0.4 g/kg for 5 days, and then intravenous methylprednisolone was initiated at a dose of 1 g/day for 7 days. One month later having failed with this therapy, she was treated for the second time with a 5 day course of IVIG, but there was no improvement. Within the third month of the disease, 2 weeks after a successful laparoscopic ovarian tumor removal, a dramatic improvement was seen in her symptoms and she was discharged without any maintenance

A. Koksal · S. Baybas · B. Mutluay · Y. Altunkaynak · A. Keskek
Department of Neurology, Bakirkoy Research and Training Hospital for Neurologic and Psychiatric Diseases, Istanbul, Turkey

A. Koksal (✉)
Department of Neurology, Bakirkoy Research and Training Hospital for Psychiatry, Neurology and Neurosurgery, Bakirkoy, Istanbul 34147, Turkey
e-mail: ayhan.koksal@yahoo.com

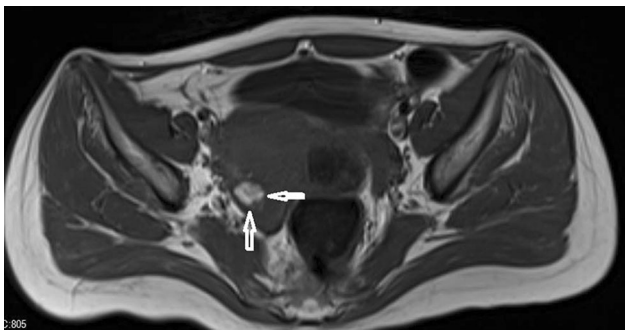

Fig. 1 T1-weighted axial pelvic MRI showing (*arrows*) right ovarian cystic teratoma

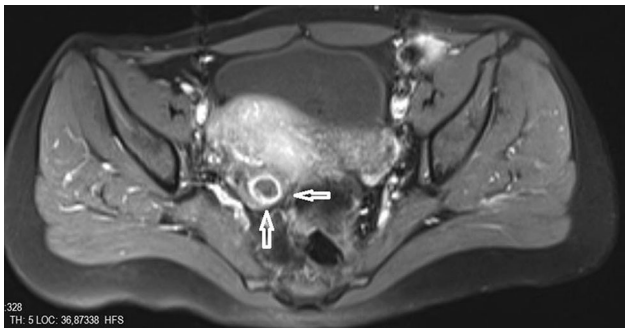

Fig. 2 T1-weighted axial pelvic MRI with fat suppression showing (*arrows*) contrast enhancing right ovarian cystic teratoma

treatment. After more than 6 months follow-up, she had no relapse and her sequelae are limited to minimal behavioral changes.

Discussion

Anti-NMDAR encephalitis is a severe but treatable disorder first described in 2007 by Dalmau [1] and is mostly under recognized. Most cases develop seizures, followed by an unresponsive or catatonic state, prolonged impairment of consciousness, central hypoventilation, bizarre dyskinesias, and autonomic symptoms [2]. The sensitivity of NMDAR antibody testing is higher in CSF than in serum which may be related to intrathecal synthesis of antibodies [3]. We found NMDAR antibody positive in CSF in this case although it was negative in serum. The diagnosis is based on the characteristic clinical picture and supportive findings in MRI, EEG and the CSF [1]. Immunosuppression with steroids and intravenous immunoglobulin is considered as the first-line treatment and, in paraneoplastic cases, complete tumor removal [1–3]. Our case improved substantially after tumor removal although immunosuppression with steroids and intravenous immunoglobulin failed. Although there are a few cases of NMDAR encephalitis during pregnancy [4], there is not much information on the postpartum period. We conclude that patients with anti-NMDAR encephalitis initially may present after pregnancy with predominantly psychiatric symptoms; this is especially important for psychiatrists and neurologists to have a high index of suspicion in the differential diagnosis of postpartum psychosis.

Conflict of interest There is no conflict of interest and this report has not been supported financially or other manner.

References

1. Dalmau J, Tuzun E, Wu HY et al (2007) Paraneoplastic anti-N-methyl-D-aspartate receptor encephalitis associated with ovarian teratoma. Ann Neurol 61:25–36
2. Iizuka T, Sakai F (2008) Anti-NMDA receptor encephalitis—clinical manifestations and pathophysiology. Brain Nerve 60:1047–1060
3. Titulaer MJ, McCracken L, Gabilondo I et al (2013) Treatment and prognostic factors for long-term outcome in patients with anti-NMDA receptor encephalitis: an observational cohort study. Lancet Neurol 12(2):157–165
4. Kumar MA, Jain A, Dechant VE et al (2010) Anti-N-methyl-D-aspartate receptor encephalitis during pregnancy. Arch Neurol 67:884–887