LETTER TO THE EDITOR

Anti-N-metil-D-aspartate receptor encephalitis: a challenge for intensivists

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Dear Editor,

Anti-N-metil-D-aspartate receptor (NMDAR) encephalitis is an autoimmune encephalitis which typically occurs in young women (80%) and it is often a paraneoplastic disorder.[1] The exact incidence is unknown but it seems to be more frequent than reported in literature and NMDAR antibodies have been identified in 1% of the patients admitted to ICU for encephalitis.[1] Among autoimmune encephalitis, anti-NMDAR encephalitis seems to be the most common.[2] Patients may require treatment in Intensive Care Unit (ICU) for complications (coma, seizures, abnormal movements, hypoventilation and dysautonomia).[3, 4] We report a case of critically ill patient with anti-NMDA encephalitis admitted to our ICU.

After a viral-like syndrome characterized by fever, headache and cervical pain, a 25-year-old woman developed hallucinations, agitation, catatonia, abnormal movements and oro-facial dyskinesias; therefore she was admitted to Medicine Department. She was evaluated by psychiatrists and she started a treatment with delorazepam and haloperidol. However, her neurological condition worsened. Brain computed tomography (CT) and magnetic resonance imaging (RMI) were negative. Electroencephalogram showed diffused abnormally slow rhythms, cerebrospinal fluid (CSF) showed lymphocytes (360/mm^3 white cells, 98% lymphocytes), normal glucose and protein, sterile cultures and polymerase chain reaction (PCR) for virus were negative. On the suspicion of viral encephalitis, she received acyclovir therapy. After four days she developed coma, seizures, autonomic dysfunction (hyperthermia with 39 °C and tachycardia) and she was transferred to our ICU. She required mechanical ventilation and then percutaneous tracheostomy for prolonged coma with central hypoventilation and apneas. New CSF showed lymphocytes (90/mm^3 white cells, 90% lymphocytes) and intrathecal synthesis of oligoclonal bands (OCB). After two weeks, as the patient had no improvement, limbic encephalitis was suspected. NMDAR autoantibodies were identified in serum and CSF. The patient received intravenous immunoglobulins (0.4 mg/kg) for five days and methylprednisolone. Chest, abdomen, pelvis CT and transvaginal pelvic ultrasound were negative for detection of tumors. Serological tumor markers were negative. After she had a neurological improvement. After 43 days of hospitalization in the ICU she was transferred in a Medicine Department without neurological sequelae.

When patients are admitted in ICU for encephalitis, after excluding infectious etiology, anti-NMDAR encephalitis should be considered, especially if it occurs in young women with psychiatric symptoms and movement disorders. Our purpose is to focus intensivists' attention on this type of encephalitis because the complications often require admission to ICU. Coma and central hypoventilation require prolonged mechanical ventilation and tracheostomy.[3, 4] Autonomic dysfunction can lead to hypertension, hypotension, tachycardia, bradycardia and asystole.[4] Hyperthermia is often of difficult management.[4] Diagnosis is often late but it is crucial because the immunomodulatory therapies (corticosteroids, immunoglobulins or plasma exchange) determine the resolution of neurological conditions for the most of the patients. Brain CT and RMI are often negative, while CSF shows lymphocytic pleocytosis with OCB. In order to confirm the diagnosis, NMDAR auto-antibodies should be identified in serum and in CSF. The detection and removal of any underlying tumor (often ovarian teratoma) have always to be taken into consideration for the improvement of the prognosis.

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