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Letter to the Editor

Therapeutic Plasma Exchange in Anti-N-Methyl-D-Aspartate-Receptor (Anti-NMDA-R) Encephalitis associated with Benign Ovarian Teratoma

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To the Editor:

Two recent articles [1,2] discussed the role of therapeutic plasma exchange (TPE) in the treatment of patients with anti-N-methyl-D-aspartate-receptor (anti-NMDA-R) encephalitis. Mirza et al. [1] discuss the case of a 14-year-old girl with anti-NMDA-R encephalitis with no response to TPE but with no underlying ovarian teratoma. A series of nine patients described by Pham et al. [2] showed good therapeutic response in their patients with and without ovarian teratomas. We share our experience of one such patient who presented to our hospital and had a favorable response to TPE.

A 34-year-old woman, with no significant past medical history, experienced auditory hallucinations. She also had visual hallucinations, agitation, generalized fatigue, anorexia, and paranoia. Her physical exam was normal. Complete blood count with platelet and differential, electrolytes and coagulation studies were within normal limits. Cerebrospinal fluid (CSF) examination was normal. CSF polymerase chain reaction for Herpes Simplex Virus (HSV1), HSV2, Cytomegalovirus, Human herpes virus (HHV-6), Varicella zoster, and Epstein Barr Virus were negative. Brain Magnetic Resonance Imaging (MRI) showed mild edema of left medial temporal lobe/hippocampus consistent with limbic encephalitis. Subsequent CT of the abdomen and pelvis showed a complex, cystic mass in the left ovary. The serum titer of anti-NMDA-R antibodies was high. She underwent unilateral salpingo-oophorectomy. Histopathology confirmed a benign, mature, cystic teratoma. She was subsequently treated with intravenous immunoglobulin (IVIG), steroids, and acyclovir without improvement. Hence, TPE was considered as an adjunctive treatment. She underwent a total of 5 TPE's (1.2 plasma volume, alternate day) with 2.5L of 5% albumin solution used as replacement fluid. Although there was minimal response in the first 2 weeks, she became progressively more coherent, alert, and ambulatory a month after treatment in conjunction with negative serum antibodies.

Paraneoplastic anti-NMDA-R encephalitis results in a syndrome with prominent psychiatric manifestations. Symptoms include memory deficits followed by decline of consciousness, seizures, involuntary movements, and dysautonomia [3,4]. In most cases, neuropsychiatric symptoms precede the diagnosis of teratoma [4]. Prompt resection of the teratoma expedites recovery [3,4]. Immunotherapy, including steroids and IVIG, is often effective. Delay in treatment can lead to mortality in these patients [4]. The benefit of TPE in these disorders is uncertain and the American Society for Apheresis assigns it as a category III (Grade 2C) recommendation [5]. Although we cannot definitively state that TPE was the main cause for improvement in our patient, we believe that it had a significant positive impact. Thus, our case corroborates evidence of a beneficial effect on anti-NMDA-R encephalitis reported by Pham et al. [2] and is in contrast to the report by Mirza et al [1]. The value of TPE in the latter report may have been negatively impacted by the delay (3 months from onset) in its use. TPE in our patient was started 15 days after admission. This result is consistent with greater improvement observed in patients with a shorter median time between admission and start of TPE [2].

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