INTRODUCTION

N-methyl-D-aspartate (NMDA) receptor encephalitis is an uncommon disorder originally described in women with ovarian teratomas. Usual symptoms include paroxysmal sympathetic hyperactivity, progressive neurological impairment and declining respiratory status. If associated with a tumor, resection along with aggressive immunotherapy can facilitate recovery. In the pediatric population, NMDA receptor encephalitis is less likely to be associated with tumors, having a negative impact on overall outcome. This disease process presents a unique set of challenges to the anesthesiologist.

PATIENT HISTORY

An 11-year-old female with an unremarkable medical history presented at a outside hospital with new onset seizures. MRI and CT scans were unremarkable and she was started on carbamazepine and discharged. Four days later she awoke with a severe headache, followed by three seizures, vomiting and fever. Carbamazepine was increased, and she was transferred to the University of Iowa Children’s Hospital.

Upon admission to our institution her mental status was stable; she subsequently developed continuous abnormal movements including left hand twitching and lip smacking. Considering the abnormal movements to be seizure activity, levetiracetam and fosphenytoin were started. Despite a decreased mental status, GCS remained 14-15. An EEG was consistent with encephalitis without seizure activity and LP showed elevated WBCs. Empiric antibiotics were started. Further CSF testing was negative.

She developed a progressive decline in her mental status and continued to have abnormal movements. An urgent MRI was considered for encephalitis. While awake, she began having hallucinations as well as fast, incomprehensible speech. Her mental status continued to decline and she was intubated and an ICP monitor was placed. ICP’s fluctuated consistent with encephalitis without seizure activity and LP showed elevated WBCs. Empiric antibiotics were started. Further CSF testing was negative.

With continued decline, she was taken to the operating room for placement of a gastrostomy tube and tracheostomy. Anesthetic management included continuation of propofol infusion with the addition of isoflurane, fentanyl and rocuronium. The medications used in this case were selected after a careful review of the literature. Phenylephrine infusion was required to maintain adequate perfusion.

ANESTHETIC MANAGEMENT

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DISCUSSION

It is believed that anti-NMDA receptor encephalitis may be responsible for up to 4% of cases of encephalitis and nearly 400 cases have been reported; however, very few cases have been reported in the pediatric population. The disease usually progresses from symptoms of psychosis and memory impairment to seizures, decreased responsiveness and, later, autonomic dysfunction requiring respiratory support. The majority of patients are young women and most have a co-existing tumor. Positive outcome is associated with resection and aggressive immunotherapy.

Several drugs used in anesthesia work through the NMDA receptor, namely ketamine and nitrous oxide. NMDA and GABA receptors are the two receptors most commonly associated with the actions of anesthetics. Halogenated anesthetics may act on the NMDA receptor, but their effects on the GABA receptor appear to be more dominant. Nitrous oxide and ketamine were avoided because of their known action at NMDA receptors and their unpredictable behavior in the setting of anti-NMDA receptor antibodies. Those medications with primary action at the GABA receptor, such as propofol, were used with caution because studies have shown knock out mice to have resistance to some of these medications, suggesting that the NMDA receptor may play some indirect role in the action of these drugs. Even though propofol is less likely to work through the NMDA receptor to produce its anesthetic effect, one report described a case in which a patient had an exaggerated response to propofol on induction resulting in extreme hypotension that was refractory to fluid resuscitation. Our patient did not have this sensitivity to propofol and her anesthetic was uneventful. Unfortunately, her clinical status did not improve and she was transferred for long term hospitalization.