ABSTRACT

When to Think Zebras, Not Horses: A Case of Delayed Care in anti-NMDA Receptor Encephalitis.

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Winner at ACP MO Chapter 2019 (not peer-reviewed or edited by this Journal)

Am J Hosp Med 2020 Jan;4(1):2020.011 https://doi.org/10.24150/ajhm/2020.011

INTRODUCTION

Anti-NMDA receptor encephalitis (NMDAr) presents a diagnostic challenge for clinicians due to its varying symptom presentation such as psychosis, abnormal behavior, and seizures. Given the presentation, and it only newly recognized as a diagnosis, anti-NMDAr is difficult to distinguish from other neuropsychiatric disorders, and delaying treatment leads to increased morbidity and mortality.

CASE

A 22-year-old female presented to the emergency department with 1-month history of increasing agitation, hypersexuality, and self-asphyxiation. She had been recently hospitalized in a psychiatric facility for 3 weeks for acute psychosis. Her abnormal behavioral reportedly started after trying a synthetic stimulant known as Flakka and given her history of chronic marijuana use was thought to be drug-induced psychosis, and an extended toxicology screen was collected. She underwent an extensive workup that included head CT, brain MRI, EEG, and paraneoplastic panel which were negative. Repeated lumbar puncture results showed elevated oligoclonal bands but was otherwise normal. CSF anti-NMDA receptor antibodies were positive at a 1:20 titer. She

was started on a course of intravenous immune globulin and solumedrol without improvement in mental status. Evaluation for possible second-line treatment with rituximab was started. Transabdominal ultrasound was performed to evaluate for possible ovarian teratoma because it is an associated trigger for anti-NMDAr encephalitis, which showed a 2.4 cm right ovarian cyst thought to represent a corpus luteal cyst. Pelvic MRI to further evaluate the ovarian cyst, showed a stable 2.5 cm ovarian cyst, but ovarian teratoma could not be ruled out. A right oophorectomy was recommended. A multi-disciplinary meeting was held prior to possible surgery to elucidate medical plan. Due to worsening mental status and inability to safely manage patient, she was transferred to intensive care unit prior to surgery for Precedex sedation and central venous catheter placement for plasmapheresis exchanges. Patient underwent laparoscopic right oophorectomy; pathology demonstrated a mature solid teratoma. She received her first plasmapheresis treatment prior to surgery, with significant improvement in mental status within two days of surgery. She received a total of five plasmapheresis treatments while hospitalized and was discharged home, with weekly plasmapheresis and solumedrol for one month. She had complete return to baseline

without recurrence of symptoms one year out from treatment completion.

DISCUSSION

The rarity and presentation in otherwise healthy, young women of anti-NMDAr encephalitis presents the challenge of delineating it from common neuropsychiatric conditions such as schizophrenia or substance-induced psychosis. Such challenges lead to bias, and result in delays in diagnosis, which in the case of anti-NMDAr encephalitis can lead to fatal outcomes. The next time a young female presents with neuropsychiatric symptoms consider autoimmune encephalitis--a zebra, not horse.