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A Case of Anti-NDMA Receptor Encephalitis with Bilateral Ovarian Teratomas

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Abstract

Anti-NMDA receptor encephalitis is an autoimmune encephalitis precipitated by antibodies generated against NMDA receptors, resulting in abnormalities in behavior, cognition, memory, and movement. Anti-NMDA receptor encephalitis most commonly occurs as a paraneoplastic disorder associated with an ovarian teratoma. While the symptomatic presentation of NMDA receptor encephalitis can be severe, removal of an associated ovarian teratoma generally causes significant improvement and the resolution of symptoms. Thus, accurate detection and early treatment are crucial for superior outcomes in these rare cases.

We present a case of a 19-year-old African American female with a strong family history of epilepsy who was brought to the ED after having two weeks of headaches, multiple seizures over the course of the previous two days, and behavioral changes. An MRI of the brain was normal, but she had an elevated opening pressure. The constellation of symptoms and elevated lumbar pressure raised the suspicion of a paraneoplastic syndrome, and a subsequent CT of the abdomen and pelvis revealed a 5 x 4.2 cm ovarian cyst. Further examination of the features of the cyst with ultrasound revealed areas of calcification. IVIG treatment was started for treatment of a suspected paraneoplastic related encephalitis. After a discussion with neuroradiology about the differential, repeat MRI brain and an MRI abdomen revealed minimal hyperintensities within the temporal lobes bilaterally in the former and evidence of previously unidentified bilateral dermoid cysts in the latter. The patient tested positive for NMDA CSF and underwent left salpingoophorectomy and right ovarian cystectomy. After treatment, she returned to her baseline but still experiences spontaneous jerking movements.

This case report discusses the early use of IVIG, communication with radiology, and the importance of maintaining a broad differential. After a thorough review of the literature, there are very few cases in which NMDA receptor encephalitis has presented with bilateral ovarian teratomas.

Introduction

Anti-NMDA receptor encephalitis is an autoimmune encephalitis precipitated by antibodies generated against NMDA receptors, resulting in abnormalities in behavior, cognition, memory, and movement [1]. Anti-NMDA receptor encephalitis is believed to affect one out of 1.5 million people per year and most commonly occurs as a paraneoplastic disorder associated with an ovarian teratoma [2]. The main method of treatment is the surgical removal of the associated ovarian teratoma, which typically causes significant improvement and the resolution of symptoms [3]. Accurate detection and early treatment of encephalitis are crucial for improving outcomes. Thus, we present a case of a 19-year-old female with anti-NMDA receptor encephalitis with bilateral ovarian teratomas.

Case presentation

This case describes a 19-year-old African American female with a strong family history of epilepsy who presented to the ED following two weeks of headaches, multiple generalized, tonic-clonic seizures, and significant behavioral changes. Her family witnessed her having a generalized seizure lasting about eight minutes, which prompted them to bring her to the hospital. This episode included tongue biting, urinary incontinence, and a post-ictal state lasting 20 minutes. The patient had no previous history of seizures, head trauma, or substance use. After being brought to the ED, she had another tonic-clonic seizure involving all four extremities, which lasted 90 seconds and was terminated with Ativan. Her symptoms progressed from post-ictal psychosis with hallucinations and somnolence to a full loss of consciousness. She also had dysautonomia, ileus,

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intermittent urinary retention, and papilledema.

Initial blood work was unremarkable, and a MRI of the brain was interpreted as normal. EEG recording was abnormal and suggested a right frontal potential for seizures and a nonspecific, generalized disturbance of cerebral function. A lumbar puncture revealed an elevated opening pressure of 55 cmH20. A meningitis panel was negative, and a CSF culture revealed no organisms. A subsequent CT of the abdomen and pelvis revealed a 5 x 4.2 cm ovarian cyst. Further examination of the cyst with ultrasound revealed areas of calcification. The initial differential diagnosis included a paraneoplastic syndrome and HSV encephalitis, and she was subsequently started on acyclovir and IVIG.

After a discussion with neuroradiology about the differential diagnosis, a repeat MRI brain and a MRI of the abdomen, to establish better detail of the cyst, were done. The MRI brain revealed minimal hyperintensities within the temporal lobes bilaterally (**Figure 1**), and the MRI abdomen and pelvis showed evidence of previously unidentified bilateral dermoid cysts (**Figure 2**). Testing for anti-NMDA receptor in the CSF came back positive, and the patient underwent left salpingoophorectomy and right ovarian cystectomy. Microscopy of the excised lesions revealed both to be mature cystic teratomas. In addition to IVIG, she was also treated with Solu-Medrol, PLEX, and rituximab. She also had to have a ventriculoperitoneal shunt placed for grade II papilledema and recurrent elevated lumbar pressures. Testing for HSV eventually came back negative and acyclovir was discontinued.

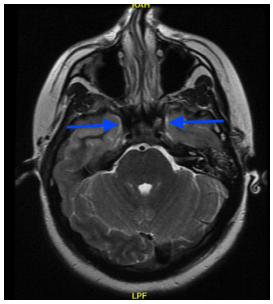

Figure 1: *Dilated cecum and small-bowel.*

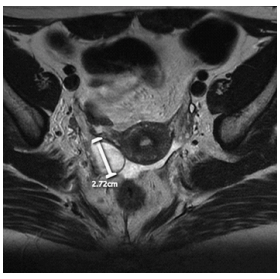

Figure 2: *One of the two ovarian teratomas identified in the MRI abdomen.*

Despite removal of the bilateral teratomas, she showed minimal improvement over the next two months, with continued poverty of speech, impaired cognition, and a fluctuating level of consciousness. She continued to have hallucinations along with violent and psychotic behavior. After a four-month hospital course, her condition gradually improved enough so that she could be discharged. Currently, the patient's mental status has returned to and is maintained at her baseline. However, she continues to have intermittent, spontaneous movements once every 30 minutes. An EEG will be done in the near future to further characterize these movements.

Discussion

This case describes a unique form of anti-NMDA receptor encephalitis that presented with bilateral ovarian teratomas. There are very few cases of bilateral ovarian teratomas associated with anti-NMDA receptor encephalitis found in the literature. This disease process is life-threatening and clinically significant, making early assessment and diagnosis of anti-NMDA receptor encephalitis critical. In a review of 119 documented cases of anti-NMDA receptor encephalitis, 12 (7%) of the cases resulted in death [4]. Typically, after timely diagnosis and removal of an ovarian teratoma, patient's rapidly improve and have a favorable outcome [3]. In this case, despite early identification and intervention, her presentation was more severe and her functional disability persisted. We hypothesize that this may be attributed to her increased burden of ovarian teratomas, but the pathophysiology underlying this association is poorly described and further investigation is warranted.

Despite the complicated recovery, one of the key therapeutic steps taken in this case was the early use of IVIG and acyclovir. Given our high clinical suspicion for encephalitis, either caused by an antibody or herpes virus, it was imperative to start early treatment, while testing was pending, to prevent further clinical deterioration and long-term complications. While it can be difficult to determine the diagnosis, it may be important in cases of suspected encephalitis to rely on the clinical presentation and treat early until further testing results return.

Initial imaging of the patient was insignificant and didn't show any signs suggestive of a paraneoplastic syndrome. Despite the negative imaging, the diagnosis of a paraneoplastic syndrome remained at the top of the differential. Her clinical presentation was discussed with radiology, and upon examination of her repeated imaging, very subtle findings, indicative of a paraneoplastic syndrome, were discovered. Context was important in this case when assessing the radiologic images, and it stresses the importance of open and clear communication with radiology in complicated presentations. Furthermore, in anti-NMDA receptor encephalitis, a tumor may not be present on initial imaging, so continual imaging to monitor for the development of an ovarian teratoma is indicated [5].

Conclusions

In conclusion, we present the case of a 19 year old female who developed anti-NMDA receptor encephalitis with bilateral mature ovarian teratomas. This case report describes the early use of IVIG in suspected encephalitis, communication with radiology, and the importance of maintaining a broad diagnostic differential. After a thorough review of the literature, there are very few documented cases in which anti-NMDA receptor encephalitis has presented with bilateral ovarian teratomas. Lastly, further investigation into the pathophysiology and impact of the burden of ovarian teratomas on the severity of

anti-NMDA receptor encephalitis is warranted.

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