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Case Report

Ovarian teratoma induced encephalitis-an ignited brain in the pelvis

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ABSTRACT

Ovarian teratomas, are common benign ovarian germ cell tumours that can yield diverse manifestations. Rarely they are associated with anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis, a serious, frequently overlooked pathology affecting young women. It is characterized by neuropsychiatric symptoms, seizures, involuntary movements, rapidly progressing to unresponsiveness and coma. Discerning the gynaecological origin of this neurological condition, especially in young women, followed by timely treatment, can significantly improve patient outcomes. We present a case of anti-NMDR encephalitis associated with ovarian teratoma in a 19-year-old female. The patient was successfully managed via ovarian cystectomy and immunomodulatory therapy. A multidisciplinary strategy remains pivotal in managing such cases.

Keywords: Ovarian teratomas, NMDAR, Pelvis

INTRODUCTION

Anti-N-methyl-D-aspartate receptor (anti-NMDAR) encephalitis, a rare paraneoplastic syndrome, is often associated with various tumours, mostly ovarian teratoma and in some cases extraovarian teratomas in mediastinum, lung or thyroid. leading to pronounced neuropsychiatric manifestations. The condition involves antibodies targeting the NR2 subunit of NMDAR receptors in the hippocampus, causing synaptic function decline.

Ovarian teratoma associated anti-NMDAR encephalitis has diverse presentation encompassing psychiatric symptoms, cognitive dysfunction, memory deficits, seizures, movement disorders, and autonomic dysfunction.[1] EEG often reveals diffuse delta slowing waves. MRI is typically normal, occasionally showing high signals in the cerebral cortex, cerebellum, or medial temporal lobe.

Diagnosis requires thorough clinical evaluation, cerebrospinal fluid (CSF) analysis for NMDAR antibodies, brain imaging studies, and detection of ovarian teratomas via ultrasound or MRI. Treatment involves tumour resection and immunotherapy, including high-dose corticosteroids, intravenous immunoglobulin (IVIG), plasma exchange, and immunosuppressive drugs such as rituximab or cyclophosphamide.[2] Early detection and tumour resection may lead to improved outcomes. If left untreated severe disability or even death can occur with mortality reported between 5 to 14% in various case series.[2]

We present a case of 19 years old woman with anti-NMDAR encephalitis and ovarian teratoma, who experienced symptom resolution following multidisciplinary treatment.

CASE REPORT

A 19-year-old female with no prior medical history presented to the emergency department with a three-week history of fever, headache, followed by incoherent speech, facial twitching, irritability, seizures, and altered consciousness. On May 11, 2023, when she developed fever for two days followed by a buzzing sensation in her

left ear; an otolaryngology evaluation was unremarkable. She subsequently developed neuropsychiatric symptoms, including emotional outbursts, self-muttering, paranoia, and suicidal ideation, and was diagnosed with acute transient psychotic disorder (ATPD) at a private hospital, where she was started on antipsychotic medications. However, her condition worsened with the development of abnormal movements and impaired consciousness. She was referred to our institution on May 29, 2023.

On examination, she was stuporous with a Glasgow coma scale (GCS) score E2M1V4 indicating eye-opening on painful stimuli but no motor response. Neurological examination revealed bilateral 3 mm pupils reacting to light, absence of bilateral plantar reflexes, and neck rigidity. Intermittent jerky movements of both upper limbs were also observed. Vital signs demonstrated tachycardia, blood pressure and oxygen saturation remained within normal limits. Other systemic examinations were unremarkable.

Suspecting encephalitis, she was admitted to the neurology intensive care unit. She received empirical broad-spectrum antibiotics, anticonvulsants, and antipsychotics. Routine blood investigations revealed Hb 12.5 gm/dl, TLC 8900/mm^3, platelet 244×10 mm^3 creatinine 1.1 mg/dl, electrolytes normal, TSH 2.19, LFT normal, HIV, hepatitis B and C non-reactive. Blood and urine culture came sterile. Anti-nuclear antibody profile, vasculitis profile, serum angiotensin-converting enzyme were all normal. Electroencephalography displayed generalized beta activity slowing. Contrast-enhanced MRI brain revealed subtle hyperintensities in basi-frontal and medial temporal regions. The CSF analysis showed lymphocytic pleocytosis and a CSF autoimmune encephalitis panel was positive for NMDA antibodies.

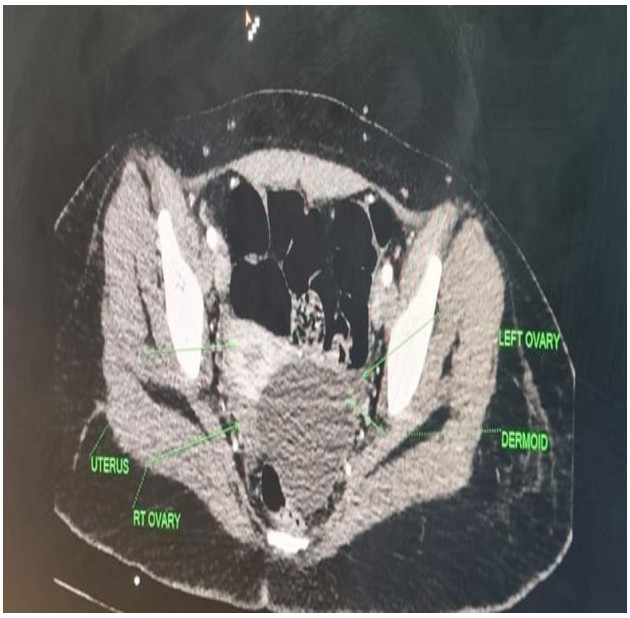

Figure 1: CECT abdomen showing cystic lesion of size 55×77×55 mm in left adnexa with fat fluid level.

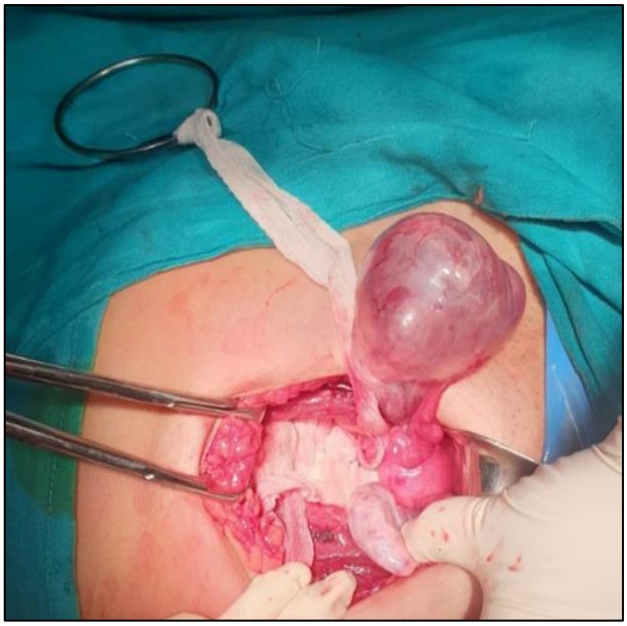

Figure 2: Laparotomy with tumour removal preserving the ovary.

Despite administering high-dose intravenous methylprednisolone and immunoglobulin, the patient's neurological condition did not improve. A contrast-enhanced CT scan of the abdomen revealed a large thin walled peripherally enhancing cystic lesion of size 55×77×55 mm in left adnexa showing fat fluid level within it. Tumour markers AFP-2.68, CA-125 18, β-Hcg<0.5, CA-19.9-5, CEA-0.79 was all negative. After discussion with parents and attendants the patient underwent laparotomy with left ovarian cystectomy on June 8, 2023. Histopathology confirmed a mature teratoma containing mature glial and neural tissue, choroid plexus-like structures, and cerebellar tissue.

Postoperatively, the patient demonstrated significant cognitive and behavioural improvement within 48 hours. Following supportive treatment and intensive rehabilitation, her recovery was excellent and she was discharged on June 20, 2023.

DISCUSSION

Anti-NMDAR encephalitis secondary to ovarian teratoma is a rare but fatal paraneoplastic syndrome. The case detailed herein emphasizes the diagnostic complexities related to this disorder.

Dalmau et al first delineated the causal relationship between anti-NMDA encephalitis and ovarian teratoma in 2007.[3] As patients frequently present to neurology and psychiatry services, gynaecologists may be less familiar with this clinical entity. Our patient had an initial prodromal phase of fever and headache followed by psychiatric symptoms, aligns with existing literature. This underscores the necessity to consider this condition in cases of unexplained neuropsychiatric presentations.

The detection of anti-NMDAR antibodies in CSF or serum, in conjunction with supportive clinical features, is pivotal in confirming the diagnosis.[4] Although electroencephalography (EEG) often shows nonspecific slow and disorganized epileptic activity, it remains a useful diagnostic tool. Magnetic resonance imaging (MRI) findings are typically nonspecific. The differential diagnoses may include acute primary psychiatric disorder, neuroleptic malignant syndrome, malignant catatonia, drug intoxications, viral encephalitis, and lethargic encephalitis. However, due to complex neuro-psychiatric presentations achieving a definitive diagnosis is challenging.

The management of ovarian teratoma-induced anti-NMDAR encephalitis necessitates a multidisciplinary strategy, including both tumour resection and immunotherapy.[5] Successful outcomes, as demonstrated in our patient, can be achieved through this combination of surgical and immune interventions, which in this case comprised steroids, intravenous immunoglobulins (IVIG), and rituximab. Long-term care in intensive settings over several weeks to months, as well as comprehensive rehabilitation, may be warranted. Close monitoring and follow-ups are crucial to evaluate long-term outcomes and avert relapse.

It must be noted that a subset of patients may endure a severe clinical trajectory, resulting in lasting disability or mortality. Therefore, prompt recognition, diagnosis, and initiation of appropriate treatment strategies are essential to enhance positive outcomes.

CONCLUSION

In conclusion, this report contributes to the existing knowledge regarding ovarian teratoma-induced anti-NMDAR encephalitis, detailing a prototypical clinical presentation, diagnostic approach, and treatment response. It emphasizes the consideration of this condition in patients manifesting neuropsychiatric symptoms, particularly when associated with ovarian teratomas. Ongoing research and case report contributions are critical to deepen our understanding of this rare disease in order to optimize patient management strategies.

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