



Anti-NMDA Receptor Encephalitis Presenting with Psychiatric Symptoms and Seizures in a Young Female with Ovarian Teratoma: A Case Report

Shraddha Khandelwal^{*}, Shahad Mahmoud and Meeshti Shethia

*Corresponding author: Shraddha Khandelwal, MBBS, MS, Department of Obstetrics and Gynaecology, Sheikh Shakhbout Medical City (SSMC), Abu Dhabi, United Arab Emirates. Tel: 506934287

Abstract

Background: Anti-NMDAR encephalitis is a rare autoimmune encephalitiswith 50 % cases associated with ovarian teratoma. It often presents with psychiatric symptoms, seizures, and autonomic instability with rapid deterioration. Early diagnosis is critical for favourable outcomes.

Case report: 21 years old Emirati female, previously healthy, was brought to the emergency department with altered mental status following episodes of generalized tonic-clonic seizures. She had erratic behaviour and persistent sleeplessness for past few days. Her Glasgow Coma Scale (GCS) on arrival was 8/15. CT brain was normal. Admission electroencephalogram revealed an abnormal slow rhythm. Cerebrospinal fluid analysis (CSF) was normal except mild leucocytosis. Infectious screen returned negative results. Two EEG diffuse days later, showed severe epileptogenic encephalopathy and foci in frontotemporal regions. At this stage, autoimmune encephalitis was strongly suspected. She was treated with methylprednisolone, therapeutic plasma exchange (TPE) and Rituximab. However, clinical course was complicated with frequent seizures requiring intubation. Pelvic imaging showed a 3 x 3 cm right ovarian teratoma for which Laparoscopic right ovarian cystectomy was performed and pathology later confirmed mature teratoma. Serum and CSF analysis confirmed a diagnosis of NMDA receptor encephalitis. Postoperatively, the patient showed gradual clinical improvement. On discharge after a month, she was able to sit, whisper, and move her limbs, although

with residual weakness. Two weeks later patient went abroad for rehabilitation and showed significant improvement upon her return after 5 months. She now follows up routinely and continues to show steady progress in her neurological function over 15 months from diagnosis.

Discussion: Early initiation of therapy, multidisciplinary approach, and early diagnosis and removal of an ovarian teratoma results in successful outcome in patients with anti NMDAR encephalitis.

Keywords: Anti-NMDA receptor encephalitis; Autoimmune encephalitis; Acute psychosis; Seizures; Case report; Ovarian Teratoma

Introduction

Encephalitis is defined as inflammation of the brain parenchyma leading to neurological symptoms resulting from the inflammatory response within the brain tissue [1]. Anti-NMDA receptor encephalitis is an autoimmune encephalitis that is identified by the presence of Immunoglobulin G (IgG) antibodies that act against the NR1 subunit of the NMDA receptors in the central nervous system [2]. It often affects young adults and children, with females being more commonly affected. The clinical presentation may mimic primary psychiatric illness, posing a diagnostic challenge. Ovarian teratomas, also known as dermoid cysts, are germ cell tumours that contain different kinds of tissue, such as skin, hair, and even muscle, or bone. Since these tissues originate from various layers of an embryo (ectoderm, endoderm, and mesoderm), they are usually quite diverse in composition [3]. One of the first associations between these two disorders was described by Vitaliani et. al. [4] and Dalmau et al. [5]. They characterized this associated condition as a anti-N-methyl-D-aspartate "paraneoplastic

(NMDA) receptor encephalitis associated with ovarian teratoma." Findings from the two referenced studies indicated that affected women presented with seizures, memory impairment, psychiatric conditions, and a reduced level of consciousness that required ventilatory support in most cases. Dalmau et al. [5] further identified that the target autoantigens of this paraneoplastic syndrome were NR1 and NR2 subunits of the NMDA receptor. This syndrome is now referred to as anti-NMDA receptor encephalitis. It can be present in males and in children [6] but almost 50% of cases of anti-NMDA receptor encephalitis is associated with ovarian teratomas [7].

Case Presentation

The patient is a 21 y/o female of Emirati descent with no past medical conditions was brought to the emergency department with altered mental status and a history of three generalized tonic-clonic seizures that lasted for less than a minute. Her Glasgow Coma Scale (GCS) on arrival was 8/15. The patient's parents stated that, for the past four days before arriving at the emergency department, the patient had been experiencing erratic behaviour and persistent sleeplessness. Because of these symptoms, the patient's parents took her to a different hospital two days later, where she was given intravenous midazolam to aid sleeping. The patient's CT was unremarkable, and her labs showed elevated CK, mild metabolic acidosis, and positive benzodiazepine in the urinalysis. Based on her presentation, she was admitted in the ICU to continue further workup. Upon admission, the patient's electroencephalogram revealed an abnormal pattern, with background rhythm slowing suggestive of encephalopathy. However, her brain MRI showed no significant abnormalities. Analysis of her cerebrospinal fluid indicated mild leucocytosis, with normal protein, glucose, and

lactate levels, while bacterial and viral PCR panels returned negative results. Two days after admission, EEG showed а repeated severe diffuse encephalopathy and epileptogenic foci in the right and left frontotemporal regions. There was, however, no evidence of subclinical seizures. At this stage, autoimmune encephalitis was strongly suspected. A CT scan of the chest, abdomen, and pelvis ruled out any underlying malignancies. Despite this, the patient's condition continued to decline. She was started on Methylprednisolone 1000 mg daily for five days, but her neurological symptoms showed no significant improvement. To further manage her condition, Therapeutic Plasma Exchange (TPE) was initiated every other day,

totalling five sessions. However, following the second TPE procedure, she experienced another seizure episode, accompanied by a drop in oxygen saturation (SpO2). As a result, she required intubation and sedation to maintain respiratory stability. A pelvic ultrasound revealed small heterogeneous lesions in the right adnexa, prompting further investigation with an MRI. The imaging confirmed a 3 x 3×1.8 cm lesion within the right ovary, exhibiting characteristic features consistent with an ovarian teratoma. The gynaecology team then proceeded with а laparoscopic right ovarian cystectomy. The teratoma contained mix of skin and hair (see below pic).



Pathology later confirmed mature teratoma. Serum and CSF analysis confirmed a diagnosis of NMDA receptor encephalitis. Postoperatively, the patient showed gradual clinical improvement. On discharge after a month, she was able to sit, whisper, and move her limbs, although with residual weakness. Two weeks later patient went abroad for rehabilitation and showed significant improvement upon her return after 5 months. She now follows up routinely and continues to show steady progress in her neurological function over 15 months from diagnosis.

Discussion

Anti-N-Methyl-D-Aspartate Receptor (anti-NMDAR) encephalitis is a form of autoimmune encephalitis. It predominantly affects young women and is often associated with underlying particularly ovarian neoplasms, teratomas. However, it can also occur in the absence of a detectable tumour. The disorder is characterized by the production of antibodies against the GluN1 subunit of the NMDA receptor, resulting in dysfunction of glutamatergic neurotransmission. Clinically, it often presents with a multiphasic course, beginning with nonspecific flu-like symptoms, followed by acute psychiatric manifestations (e.g., hallucinations, agitation, or catatonia), seizures. movement disorders, autonomic dysfunction, and reduced consciousness. Our patient initially presented with acute psychosis and generalized seizures, a pattern that may mimic primary psychiatric disorders, thereby delaying diagnosis and treatment. Diagnostic confirmation relies on the detection of anti-NMDAR antibodies, preferably in the cerebrospinal fluid, as CSF testing is more sensitive and specific than serum testing. In our case, the patient had a normal brain MRI and nonspecific EEG findings, which are common. CSF analysis showed mild lymphocytic pleocytosis and elevated protein, aligning with findings in previously published cohorts. Timely initiation of immunotherapy significantly improves outcomes. First-line therapy typically includes corticosteroids, Intravenous Immunoglobulin (IVIG), or plasmapheresis. In patients who do not respond adequately, second-line agents such as rituximab or cyclophosphamide are recommended. Our patient showed gradual improvement after receiving highdose corticosteroids and IVIG, with sustained recovery following rituximab therapy.

Conclusion

This case reinforces the importance of including autoimmune encephalitis in the differential diagnosis of new-onset psychiatric symptoms, particularly in young adults with accompanying neurological features. Early recognition and treatment can substantially improve outcomes and prevent long-term neurological deficits.

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Citation of this Article

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