01. Title: Super Refractory Status Epilepticus in Paraneoplastic Autoimmune Encephalitis Associated With Ovarian Teratoma: A Fatal Case Report

Yashasvi Sanghvi[1], Srujana Rao[1]

[1] B.J. Medical College, Ahmedabad, India

Background: Autoimmune Encephalitis(AIE) is a group of inflammatory central nervous system(CNS) disorders occurring due to breakdown of immune tolerance against specific CNS antigens. AIE can be idiopathic or can precipitate post-viral infection or as a paraneoplastic complication. Ovarian teratoma is consistently linked with production of immunoglobulin G(IgG) against N-Methyl-D-Aspartate receptors(NMDAR) found on neuronal membranes in the hippocampus and forebrain. The incidence of anti-NMDAR encephalitis is 1.5/million/year. The symptoms may initially mimic a viral prodrome or psychiatric illness. Frank neurological symptoms like seizures, movement disorders, catatonia, memory loss, and impaired cognition occur later. Diagnostic modalities include electroencephalogram(EEG), neuroimaging, and serology and/or cerebrospinal fluid(CSF) analysis for autoantibody detection, followed by computed tomography(CT) of the chest, abdomen, and pelvis for detection of lung, breast, ovarian, testicular, or other abdominal tumors. The condition responds well to tumor removal, plasmapheresis, steroids, intravenous immunoglobulin(IVIG), and other immunotherapeutic agents such as Cyclophosphamide. Anti-NMDAR encephalitis can be complexed by super refractory status epilepticus(SRSE) which is a state of neurological urgency and is accompanied by profound mortality. SRSE refers to seizures that persist for 24 hours or more after starting anaesthetics, including the seizures that recur after stopping the anaesthetics.

The Case: A 39-year-old, previously healthy female, developed three episodes of seizures on a single day, involving involuntary movements of all four limbs with tongue bite and mild fever. She was stable for the next 11 days after which she developed irritability, behavioural abnormalities, and reduced responsiveness. She was brought to a local hospital in an unconscious state. Glasgow coma scale score was 9(G2V2M5). Brain magnetic resonance imaging(MRI) revealed left hippocampal swelling. EEG revealed no significant abnormality. CSF analysis revealed elevated protein and cell count, normal glucose, and negative microscopy for organisms. The patient had a seizure during hospitalisation at the local hospital, for which phenytoin was given. The patient and family decided to continue further management at our hospital. Virological analysis was normal. CSF autoimmune encephalitis panel revealed the presence of anti-NMDA antibodies. A contrast-enhanced CT of the abdomen and pelvis was done which revealed an approximately 37 x 37 x 35 mm-sized right ovarian dermoid cyst(Figure 1). The patient was started on Methylprednisolone, IVIG, and Cyclophosphamide. Antiepileptics Levetiracetam, Valproate, and Phenytoin were ineffective in achieving seizure control. Midazolam, Ketamine, and Propofol were used to sedate the patient. The patient was intubated and provided with oxygen therapy. The patient had persistent seizures for 24 hours despite the anesthetic therapy, leading to the diagnosis of SRSE. Unfortunately, despite aggressive management, the patient died due to cardiorespiratory arrest, before tumor resection surgery could be performed.

Conclusion: The case underlines the importance of maintaining high clinical suspicion for paraneoplastic autoimmune encephalitis in female patients with psychiatric presentations followed by SRSE. Early diagnosis is imperative because immunotherapy and surgical tumor removal dramatically improve clinical outcomes. As seen in this case, delayed tumor management contributes to rapid disease progression and profound mortality rates.

Key Words: Anti-N-Methyl-D-Aspartate Receptor Encephalitis, Teratoma, Ovarian, Status Epilepticus, Case Report

Figure:

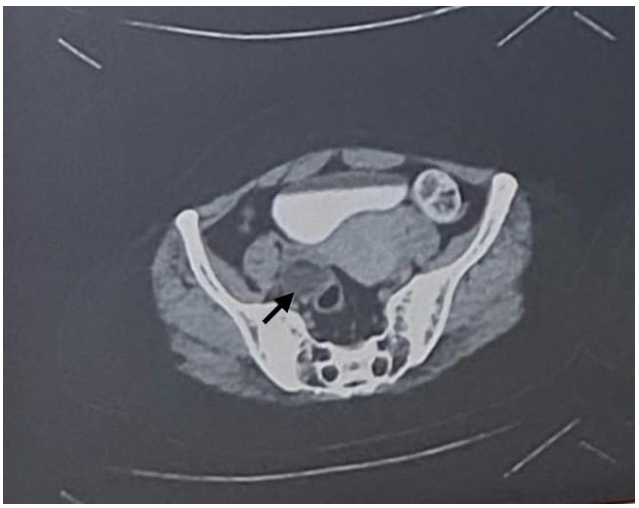

Figure 1: Contrast enhanced computed tomography(CECT) of abdomen and pelvis showing approximately 37 x 37 x 35 mm sized, peripherally enhancing, well-defined, rounded, predominantly cystic lesion with fat component, solid component and curvilinear calcification within, arising from right ovary, hence suggesting the presence of a right ovarian dermoid cyst (white arrow).
IJMS

ISSN 2076-6327

This journal is published by Pitt Open Library Publishing