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Original Research Paper

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BRAIN ON FIRE- A CASE REPORT ON ANTI NMDA RECEPTOR ENCEPHALITIS IN A TERTIARY CARE HOSPITAL

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ABSTRACT To emphasize the importance of suspecting anti-N-methyl-D-Aspartate-Receptor(NMDAR) encephalitis in young females presenting with new onset psychiatric symptoms and association with ovarian teratoma. Early diagnosis and management can decrease morbidity and disability.

KEYWORDS : Anti NMDA encephalitis, ovarian teratoma, psychiatric symptoms.

INTRODUCTION:

Anti-NMDA encephalitis is a type of autoimmune encephalitis. Autoimmune encephalitis syndromes are due to antibodies to neuronal cell surface, synaptic proteins, intracellular antigens or even as a part of other autoimmune diseases. Prominent feature of anti-NMDA encephalitis is psychiatric symptoms. Sleep disorders, seizures, frequent dyskinesia, autonomic instability, and language dysfunction are the other common clinical features. The NMDA receptor (NMDAR)is an ion channel receptor found at most excitatory synapses, where it responds to the neurotransmitter glutamate, and therefore belongs to the family of glutamate receptors. Anti NMDAR autoimmune pathology are frequently seen in ovarian tumors usually a teratoma as well as in some cases of herpes simplex encephalitis. The disease is prevalent in young females with male to female ratio of 2:8. It is considered that the antibodies to the NR1-NR2 subunits of the NMDA subtype of glutamate receptors develop in response to this abnormal tissue. Differentiating from a primary psychiatric illness is challenging especially at the onset. We present a case report of anti-NMDA encephalitis in a 16-year-old female developing psychiatric symptoms who became symptomatically better on excision of ovarian teratoma.

CASE:

16-year-old female was brought to emergency room with history of altered behaviour of one week duration. She had one episode of abnormal posturing of limbs with facial deviation at the onset of illness. There was no history of fever, relatives also declined any associated headache, vertigo, nausea, or blurry vision. Neurological examination revealed an irritable patient with impaired short term memory, echolalia, echopraxia, rigidity and oro-facial dyskinesias without any motor power deficits. Routine blood evaluation showed raised inflammatory markers including C-reactive protein (CRP). Urine sample showed elevated WBCs. Magnetic resonance imaging (MRI) brain showed no obvious abnormal signal changes. Eelectroencephalography(EEG) had normal background activity without any abnormal discharges. Uultrasound (USG) abdomen showed multiple internal echoes in urinary bladder, ovarian cyst with hyperintense rim was also detected (figure 1.). After initial evaluation she was started on antiepileptic medications and treatment for urinary tract infection with antibiotics. Patient became progressively confused with aggressive behaviour without any fever spikes, meningeal signs, motor or sensory deficits. Cerebrospinal fluid (CSF) study showed 12 cells with lymphocyte predominance, protein and sugar was in normal range. She was started on empirical antiviral, meningitic dose of antibiotics along with antipsychotics for symptom relief. Autoimmune panel was strongly positive for anti- NMDA antibodies. Patient was started on intravenous Immunoglobulin (0.4g/kg per day) administered for 5 days along with pulse corticosteroids. Considering high association of NMDAR encephalitis with ovarian pathology, MRI of pelvis was done which was conclusive with ovarian teratoma of size 2.7*2.3*3.6cm (figure 2a, figure 2b). Surgical removal of teratoma was done three weeks later. Patient had symptomatic improvement and was discharged subsequently. EEG monitoring was performed 2 months after discharge, which revealed a normal pattern. Her cognitive function became normal and her psychiatric symptoms, involuntary movements including oral-facial dyskinesias had a complete recovery.

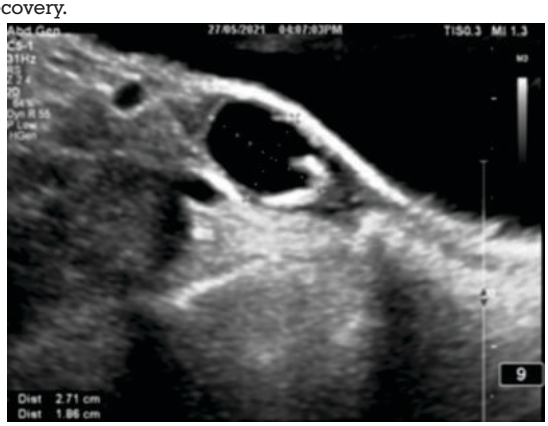

Figure 1, Ultrasonogram Of Pelvis Showing Cystic Ovarian Lesion With Hyperintense Rim

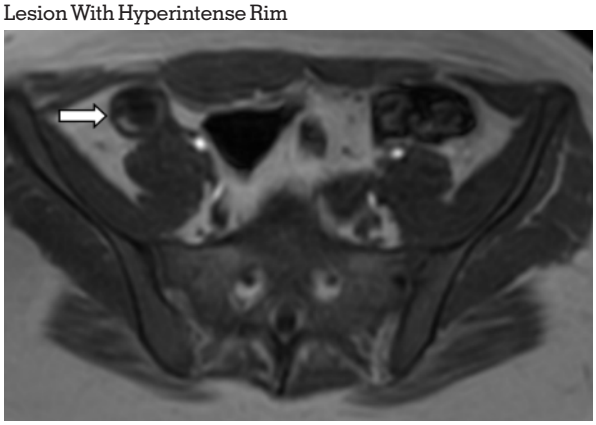

Figure 2a, T1 MRI sequence images of pelvis showing right ovarian teratoma

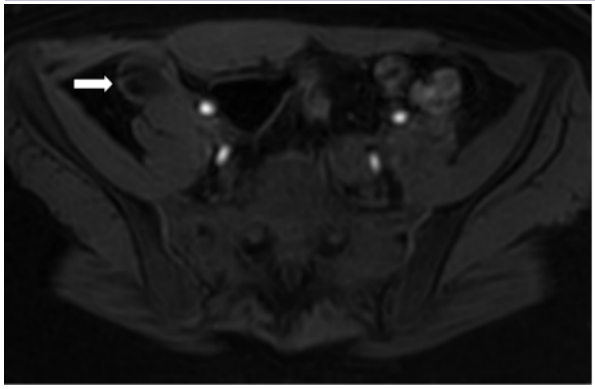

Figure 2b, T1 SAT MRI images of pelvis showing loss T1 hyperintense rim of the ovarian mass suggesting fat stranding

DISCUSSION:

Anti-NMDAR encephalitis is a type of limbic encephalitis that is typically found in young women with teratomas. Common clinical features include prominent psychiatric manifestations[1,2],sleep disorders[3], memory deficits, seizures, decreased level of consciousness ,frequent dyskinesias, autonomic instability and language dysfunction. CSF of this patients shows lymphocytic pleocytosis or oligoclonal bands. Electroencephalography may show infrequent epileptic activity, but frequent slow, disorganized activity may not correlate with most abnormal movements. Brain MRI may be normal or contrast enhancing abnormalities in cortical and subcortical regions are seen. PET reportedly shows a characteristic increase in the fronto-occipital gradient of cerebral glucose metabolism, which correlates with disease severity. The diagnosis of anti-NMDA receptor encephalitis is confirmed by the detection of IgG antibodies to the GluN1(also known as NR1)subunit of the NMDA receptor in CSF[4].The differential diagnosis includes primary psychiatric disorders(acute psychosis or schizophrenia), malignant catatonia, neurolept malignant syndrome[5],viral encephalitis[6],and encephalitis lethargica[7].The detection of an ovarian teratoma is age dependent; approximately 50 percent of female patients older than 18 years have unilateral or bilateral ovarian teratomas, while less than nine percent of girls younger than 14 years have a teratoma[8].This patient was steroid unresponsive, since a high dose intravenous administration of steroids failed to improve her symptoms. After IVIG infusion and tumour resection she recovered to normal status in a short period of time, and we gradually tapered down all her medications. Although most studies indicated recovery was a slow process for anti-NMDAR encephalitis, our experience in patients with teratoma and tumor resection, had good prognosis and fast recovery time.

CONCLUSION:

Early diagnosis and intervention in anti-NMDAR encephalitis patients with ovarian teratoma can decrease morbidity and improve clinical outcome.

REFERENCES:

1 Kayser, Matthew S., Titulaer, Maarten J., Gresa-Arribas, Núria and Dalmau, Josep (2013) 'Frequency and characteristics of isolated psychiatric episodes in anti–N-methyl-d-aspartate receptor encephalitis'. *JAMA neurology*, 70(9), pp. 1133–1139.

2 Guasp, Mar, Giné-Servén, Eloi, Maudes, Estibaliz, Rosa-Justicia, Mireia, et al. (2021) 'Clinical, Neuroimmunologic, and CSF Investigations in First Episode Psychosis'. *Neurology*, 97(1), pp. e61–e75.

3 Ariño, Helena, Muñoz-Lopetegi, Amaia, Martinez-Hernandez, Eugenia, Armangue, Thaís, et al. (2020) 'Sleep disorders in anti-NMDAR encephalitis'. *Neurology*, 95(6), pp. e671–e684.

4 Prüss, H., Dalmau, J., Harms, L., Höltje, M., et al. (2010) 'Retrospective analysis of NMDA receptor antibodies in encephalitis of unknown origin'. *Neurology*, 75(19), pp. 1735–1739.

5 Sansing, Lauren H, Tüzün, Erdem, Ko, Melissa W, Baccon, Jennifer, et al. (2007) 'A patient with encephalitis associated with NMDA receptor antibodies'. *Nature clinical practice. Neurology*, 3(5), pp. 291–296.

6 Gable, M. S., Gavali, S., Radner, A., Tilley, D. H., et al. (2009) 'Anti-NMDA receptor encephalitis: report of ten cases and comparison with viral encephalitis'. *European Journal of Clinical Microbiology & Infectious Diseases: Official Publication of the European Society of Clinical Microbiology*, 28(12), pp. 1421–1429.

7 Dale, Russell C., Irani, Sarosh R., Brilot, Fabienne, Pillai, Sekhar, et al. (2009) 'N-methyl-D-aspartate receptor antibodies in pediatric dyskinetic encephalitis lethargica'. *Annals of Neurology*, 66(5), pp. 704–709.

8 Titulaer, Maarten J., McCracken, Lindsey, Gabilondo, Iñigo, Armangué, Thaís, et al. (2013) 'Treatment and prognostic factors for long-term outcome in patients with anti-NMDA receptor encephalitis: an observational cohort study'. *The Lancet. Neurology*, 12(2), pp. 157–165.