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RESEARCH ARTICLE

YOUNG FEMALE WITH AUTOIMMUNE ENCEPHALITIS – A CASE REPORT

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ABSTRACT

Autoimmune encephalitis (AE) is a type of brain inflammation where the body's immune system mistakenly attacks healthy cells and tissues in the brain or spinal cord. It is a rare, complex disease with incidence of 1 in 100000 population that can cause rapid changes in both physical and mental health. It has multiple etiologies.AE was recognized as a disease relatively recently. Because of this, some doctors may not be familiar with this condition, and it may be misdiagnosed as a psychiatric or neurological disorder. Early diagnosis and treatment are critical to minimizing both short- and long-term complications of this disease. We hereby present to you an interesting case of a 30 year old female with symptoms similar to a wide spectrum of Neurological diseases. Nevertheless to our surprise it came out to be a one in one lakh cases.

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INTRODUCTION

Encephalitis is an inflammatory condition of the brain with multiple etiologies. The autoimmune encephalitis syndrome have a wide clinical spectrum that ranges from typical limbic encephalitis to syndromes with complex neuropsychiatric symptoms such as deficits of memory, cognition, psychosis, seizures, abnormal movements, or coma (1-6). Anti-NMDA receptor encephalitis is associated with a predictable set of symptoms that combine to make up a characteristic syndrome (10-11). Patients present with a prodromal headache, fever, or a viral-like process, followed in a few days by a multistage progression of symptoms that include: Prominent psychiatric manifestations (anxiety, agitation, bizarre behavior, hallucinations, delusions, disorganized thinking, psychosis)(8-9). Rarely, the disease can be monosymptomatic (eg, manifesting as isolated psychosis), or psychiatric symptoms. Sleep disorders, including sleep reduction at disease onset and hypersomnia during recovery, Memory deficits, Seizures, Decreased level of consciousness, stupor with catatonic features.Frequent dyskinesias: orofacial, choreoathetoid movements, dystonia, rigidity, opisthotonic postures. Autonomic instability: hyperthermia, fluctuations of blood pressure, tachycardia, bradycardia, cardiac pauses, and sometimes hypoventilation requiring mechanical ventilation.

Language dysfunction: diminished language output, mutism, echolalia.An approach to the diagnosis should include neuroimaging, EEG, lumbar puncture, and antibody testing on serum and CSF(8-9)

CASE REPORT

CASE: A 30 years old lady with no medical co-morbidities presented to Emergency Room (ER) with complaints of headache- Holocranial, mild to moderate intensity, double vision – side by side / horizontal, generalised weakness, confused state, reduced oral intake since 2-3 days. MRI brain showed changes which has been discussed below in detail. On arrival to ER patient was conscious, but confused. Vitally stable. No history of nausea/ vomiting.

On Clinical Examination:

Primary Survey: Airway-Patent Was Speaking In Full Sentence Breathing- Saturation: 99%, Rr:16/Min. Circulation- Pulse : 86 B/M, B.P: 100/60 mmhg, Peripheral Pulses: Palpable, Rbs: 110 Mg/Dl Disability- Gcs: E4v4m6, Conscious But Confused

History (Mn: SAMPLE): S(Symptoms)-Headache, Double Vision, Generalised Weakness With Confused State. A(Allergy)-No Known Drug Allergy. M(Medications)-Not On Any Chronic Medications. P(Past History)-No Significant Past Medical History. L(Last Meal)-3 Hours Ago. E(Events) -There Is No Significant Event Prior To Arrival

SECONDARY SURVEY

HE-ENT: No Pallor/ Cyanosis/ Icterus

Chest: B/L AE Equal.

Cvs: S1S2 Heard.

Abdomen: Soft, Non-Tender, Bowel Sounds Heard.

Ext: No Pedal Edema

Neurological Findings

GCS – E4V4M6
Conscious But Confused/Irritable And Dull
Pupils B/L 3mm Equal And Reactive, Eom Full, Dilopia Present
Neck Rigidity Present. No Nystagmus No Facial AssymetryNo DysarhriaNo Pronator DriftNo Past Pointing. Motor Power 5/5 All Four Limbs , Sensations intact

Based On Our Findings, Some Of The Possible Causes Are Listed As

- Tuberculosis
- Toxic Metabolic Causes
- Wernickes Encephalopathy
- Neoplastic Disorders
- Encephalitis- Bacterial/ Viral/ Fungal/ Autoimmune
- Demyelinting Disorders – Ms/Nmo/Mog
- Vasculitis- Cns Lupus/ PcnsOrSeconday Vasculitis/ Neuro Sarcoiddois

TREATEMENT IN ER: In view of the finding, point of care (POC) Test were sent which includes, electrocardiogram which was suggestive of Normal Sinus Rhythm with. Patient's Blood Gas Analysis was normal pH 7.36, Pco2 35.8, HCO3 24, NA+ 136, K+ 3.7, LAC 1.0. Patient was canulated with 18G canula over the left anti-cubital vein and administered below medications.

- INJ.ACYCLOVIR 500MG IV STAT
- INJ.CEFTRIAXONE 2 GM IV STAT
- INJ.LEVETIRACETAM 1000MG IV STAT
- INJ.PAN 40MG IV STAT
- IV NS @80ML/HR

She was then subjected to a plethora of investigations to find out the cause of her symptoms.

LAB FINDINGS

- Routine Blood Investigations-Cbc, Lft, Kft: Wnl
- Paired Culture: Wnl
- Viral Markers: Wnl
- Ana/ Cta: Positive
- Autoimmune Encephalitis Panel: Negative
- Para Neoplastic Panel: Negative
- Anti Thyroid Peroxidase Antibody: Negative
- Pet: Negative For Any Mass Lesion
- Compliment C3, C4: Negative
- Ana Immunofluorescence Test
- Anti Cardiolipin Antibody: Negative

LUMBAR PUNCTURE-CSF ANALYSIS

TEST NAME	RESULT
APPEARANCE	Clear
CSF GLUCOSE	65 mg/dl (15-40 mg/dl)
PROTEIN	123.8 mg/dl (15-40mg/Dl)
TLC	35 (0-8)
RBC	Nil
GRAM STAIN	No Organism Seen
ADA	1.2 U/L (0-10)
GENE XPERT	Negative
ME PANEL	Virus, Bacteria And Fungi Not Detected
INDIA INK	No Cryptococcus Like Organism Seen

Radiological Investigations

MRI BRAIN

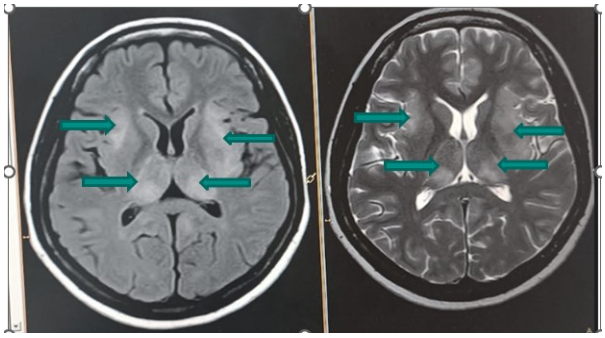


Image 1. B/L symmetrical, T2/Flair, Hyperintensities are seen in B/L Thalami AND B/L external capsule

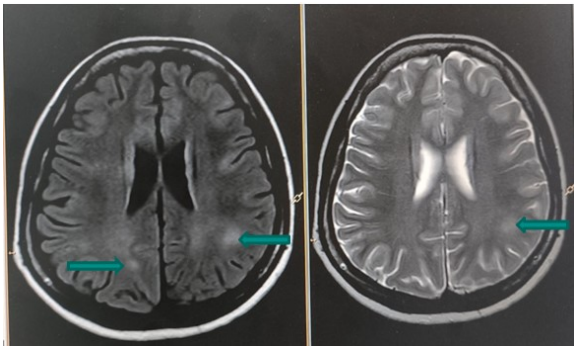


Image 2. B/L symmetrical, T2/Flair, Hyperintensities are seen in occipital and left parietal lobe

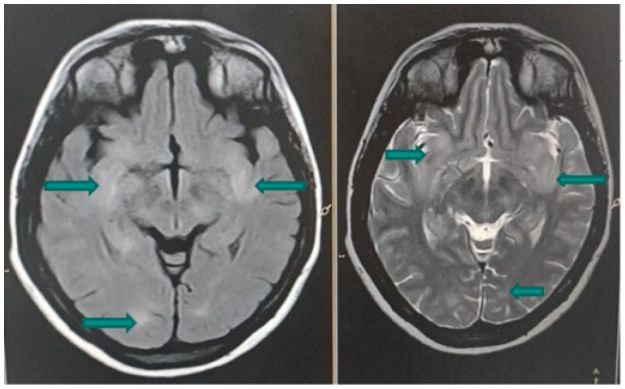


Image 3. B/L symmetrical, T2/Flair, Hyperintensities seen in sub cortical white matter of occipital lobe

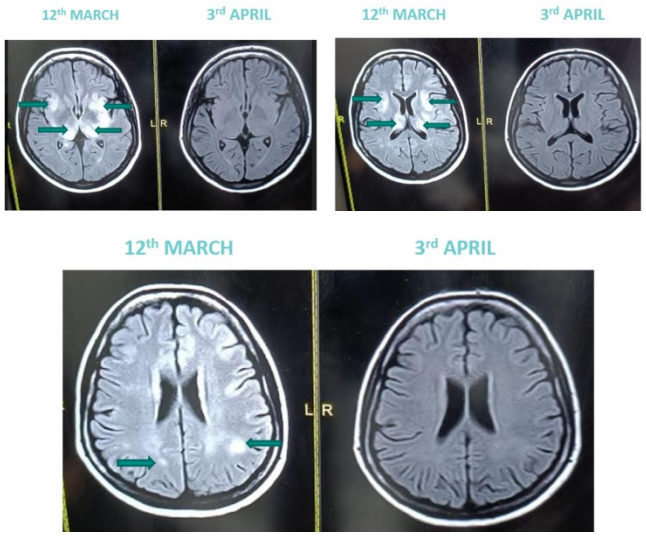
Ancillary Investigations: She was Positive for ANA (PM-Scl). Her Pet-CT showed no evidence of FDG avid metabolically active lesions anywhere in the body.

DISCUSSION

On arrival of the patient to the ER, We were suspicious of a bacterial or viral encephalitis, vasculitis, neoplasm and other above described differentials and line of management was started accordingly. The treatment included empirical anti virals and antibiotics. Other investigations as per our differentials were sentonly ANA (PM-Scl) was positive, which raised our doubt toward autoimmune encephalitis or a possible neoplasm. Lumbar puncture and MRI brain were ordered. Lumbar puncture showed a CSF glucose=65 mg/dl, protein=123.8mg/dl, TLC =35, while ,MRI brain showed hyperinrensities as described above. To rule out neoplasm PET-CT was ordered which was normal. Hence, we could get to the diagnosis of Autoimmune Encephalitis based on the clinical summary mentioned above. Its worthy noting the low incidence of the case ,(incidence of 1 in 100000) that makes it a rare entity in our clinical setting. Empricial treatment received by the patient was then stopped and the patient was shifted to a critical care unit and received the following treatment:

- Iv methylprednisolone 1 gm for 5 days followed by oral steroids
- Planned for -inj.rituximab 2 doses 2 weeks apart

After starting on iv methyprednisolone and 1st dose of rituximab, patient showed significant improvement on symptoms. Double vision and language improved, headache was better. A Rheumatology consult was sought in view of positive ANA and no active intervention was advised.The patient was then discharged with an advise of oral steroids ;Inj Rituximab 2 weeks apart. Second dose of rituximab was delivered on follow up and repeat MRI brain was ordered which showed 90% resolution of hyperintensitiies, as evident by the images below



CONCLUSION

In a nutshell, A rare disease like autoimmune encephalitis can happen even in a young patient with no significant past medical history. Any patient who is presenting with new onset non-specific neurological symptoms like confusion, diplopia, headache should not be neglected or mislead as psychiatric illness and investigated thoroughly to rule out all possible neurological and neoplastic causes.

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