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Rare Case of Seronegative Neuromyelitis Optica Presenting with Seizure

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Abstract: ***Neuromyelitis optica spectrum disorder (NMOSD) is a rare autoimmune condition primarily characterized by optic neuritis and longitudinally extensive transverse myelitis (LETM). Seropositivity for aquaporin-4 antibodies (AQP4-IgG) has been considered a hallmark of NMOSD. However, this case presentation focuses on an exceptional case of NMOSD in a 28 year old female presenting with 1st episode of seizure who didn't exhibit evidence of AQP4-IgG seropositivity. Keywords • Seronegative Neuromyelitis Optica • Generalized Tonic-Clonic Seizures.***

Keywords: Seronegative Neuromyelitis Optica, Generalized Tonic-Clonic Seizures, Atypical NMO Presentation, AQP4-IgG Negative, Autoimmune Neurological Disorder

1. Introduction

Neuromyelitis Optica is a disorder characterized chiefly by optic neuritis and acute myelitis with MRI changes that extend over at least 3 segments of spinal cord. Patients rarely present with atypical presentations like features of endocrinopathy, intractable hiccoughs or manifestation of features of encephalopathy like seizures. Previously, this disease was known as Devic's Disease and was thought to be a variant of Multiple Sclerosis. This disease is strongly associated with presence of specific antibody marker (NMO IgG) targeting water channel aquaporin-4 in 80% of cases and Anti-MOG IgG in about 30% of NMO-IgG seronegative patients. Management of these patients is by suppressing acute flares with corticosteroids and long term immunosuppression for maintaining remission as required. Here we discuss such a case of NMO IgG seronegative female presenting with generalized tonic clonic seizures and past history of temporary visual loss and paraplegia.

2. Case

A 28 year old Muslim female, who is a house wife in a lower middle class family residing in Gondal with known case of hypothyroidism on thyroxine (100) since 2 years presented to Civil Hospital Rajkot with sudden onset of involuntary movements of all 4 limbs with up-rolling of eyeballs and frothing of mouth the night before as witnessed by her husband. Patient had 1 such episode and patient regained consciousness after 10 to 15 minutes. Patient had not been fully oriented in form of occasionally not following commands and even developed aphasia. Patient was also not able to swallow any solids or liquids after the episode (gag lost). Patient had no history of any such episode in past. Patient was afebrile during the episode. There was no association with headache, nausea, vomiting. There was no complain of weakness, tingling sensations or numbness in any of the limbs. There was no history of dizziness, or fall. Patient's relative gave a remote history of transient loss of vision bilaterally 1 month back which recovered spontaneously and a history of paraplegia 3 years ago which was relieved after 1 month. Patient had no addiction history. Patient had fairly regular sleeping, bowel and bladder habits and was on mixed diet before the episode. Patient had 2.5 month full term male child vaginally delivered after a normal uneventful gestation period. Patient was examined in supine position under natural light. On examination, patient was conscious not fully oriented. Patient was well nourished and well-built as per age. Patient was vitally stable. There were no abnormal findings on general examination. Pupils of the patient were bilaterally equal, reactive to light for direct and consensual light reflex. Fundus exam didn't show any changes of papilledema but revealed changes of left sided optic neuritis. There were no other cranial nerves abnormalities detected. Patient had equal movements with no reduction of tone and 4 power in all 4 limbs. There was no evidence of ataxia and nystagmus was absent. Abnormal involuntary movements were absent. All reflexes were intact and bilateral plantar reflexes were flexor. All peripheral and cortical sensations were intact and no abnormalities in gait or posture were seen. Her Cardiovascular system, respiratory system and abdominal examination were unremarkable.

Differential Diagnosis

- Meningitis and Meningoencephalitis
- Structural Brain Lesion
- Multiple Sclerosis
- NMOSD
- ADEM

Investigations

Her routine investigations including CBC, Electrolytes, RFT, LFT and CRP were normal. ESR, thyroid and coagulation profile were normal and serology was negative. Chest X- ray didn't reveal any abnormalities. MRI Brain with contrast revealed " Multiple cortical subcortical hyperintensities involving bilateral frontal parietal and bilateral basal ganglia region without any post contrast enhancement suggesting underlying changes of ACUTE DISSEMINATED ENCEPHALOMYELITIS (ADEM)". MRI Study of spine revealed "Evidence of few multifocal long segment

intramedullary hyperintense lesions involving upper cervical cord at C2, C3 and C4 level with hyperintense signal in anterior aspect of medulla suggesting underlying demyelinating etiology like MS / NMOSD with possibility of infective etiology less likely. " CSF studies revealed mild lymphocytic pleocytosis with protein and sugar within normal limits. CSF didn't detect oligoclonal bands. Serum didn't show presence of Anti Aquaporin- 4 antibodies but shows strong positivity for Anti MOG antibodies.

Treatment

Patient was started with antiepileptic Levetiracetam. Patient was given Methylprednisolone 1000 mg for five days after which patient recovered from the episode and patient is currently on tapering dose of steroids and will be considered for long term immunosuppresives after completely weaning of from steroids after assessing requirement and side effect profiles.

3. Discussion

This atypical presentation of NMOSD underscores the diagnostic complexities faced by clinicians when confronted with patients who lack the typical biomarkers. The case discusses the patient's clinical course, neuroimaging findings, cerebrospinal fluid analysis, and antibody testing, ultimately leading to the diagnosis of seronegative NMOSD. It highlights the importance of considering alternative diagnostic avenues, such as the presence of anti-myelin oligodendrocyte glycoprotein antibodies (MOG-IgG), and the significance of ruling out other neurological and autoimmune conditions with similar presentations. This case presentation emphasizes the need for ongoing research and clinical vigilance to better understand the heterogeneous nature of NMOSD and to improve diagnostic accuracy in cases that defy conventional expectations.

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