Anti-NMDA Receptor Encephalitis: A Masquerade Ball of Neuropsychiatric Symptoms

Sir,

Anti-NMDA (N-methyl-d-aspartate) receptor encephalitis, categorized officially in 2007, is a rare disorder which is potentially fatal at one end of the spectrum but may demonstrate astounding recovery, especially if the disease is mild and detected early.[1] Being a relatively modern disease, it poses great challenges in the diagnosis and subsequent management.

A 39-year lady presented with two episodes of typical generalized tonic clonic seizures. She had a preceding two weeks history of low-grade fever and headache. At presentation, she was vitally stable. Apart from slight agitation, general and systemic examinations were unremarkable. Plain CT scan of the brain was normal. Cerebrospinal fluid (CSF) examination showed pleocytosis with 93% lymphocytes. CSF was negative for bacteriologic and fungal cultures, acid-fast bacilli, mycobacterial DNA, and viral Polymerase chain reaction (PCR). She was started on ceftriaxone, vancomycin and acyclovir at presentation for the suspicion of partially treated bacterial meningitis or viral encephalitis. She was not showing any significant improvement. Repeat CSF examination after two days showed rising proteins. She was started on antituberculous medications with steroids but she continued to deteriorate. Later, patient started to develop behavioral and cognitive changes. She then developed muscular rigidity and autonomic instability. Thus, suspicion of autoimmune encephalitis was raised and her CSF was sent for anti-NMDA receptor antibodies which came out to be positive. She was diagnosed to have anti-NMDA receptor encephalitis. Full body imaging did not show any tumor including ovarian teratomas. Patient was started on high-dose methylprednisolone and IVIGs for 5 days, which did not bring any improvement. Because of the non-responsiveness, she was then placed on rituximab and cyclophosphamide, which dramatically improved the patient condition. She continued to show improvement in her clinical and cognitive condition and was shifted under care of the rehabilitation team. On follow-up one year after the presentation, she had good performance status without any neurological or intellectual disability.

Anti-NMDA receptor encephalitis manifests in a variety of overlapping clinical patterns. Some reviews describe a simplified sequence of prodromal phase, psychotic and/or seizure phase, unresponsive phase and finally hyperkinetic phase.[2] Testing for the specific anti-NMDA receptor antibodies in CSF should be performed for confirmation.[3,4] As it is frequently associated with tumors in the body, especially ovarian teratoma, full body imaging is suggested.[3] Specific treatment involves glucocorticoids, IVIGs, plasma exchange, rituximab and cyclophosphamide, either in sequence or in combination.[3]

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