Answer to Medical Quiz - 2

Answers:

A. Right cerebellar hemisphere, in the left pons & superior colliculus and adjacent to the lateral ventricles and cervical cord lesion with marked swelling .

B. Neuromyelitis optica spectrum disorder (NMSOD)

C. Multiple sclerosis (MS)/Acute disseminated encephalomyelitis (ADEM)/ Sarcoidosis,/CNS vasculitis/ Leber hereditary optic neuropathy,

D. Serum Anti-AQP4 antibody

E. Intravenous methylprednisolone/ Plasmapheresis

Review:

Neuromyelitis optica spectrum disorder (NMOSD) is a rare and chronic autoimmune disorder of the central nervous system (CNS) that typically presents with inflammation to the optic nerves (optic neuritis) and the spinal cord (acute transverse myelitis) caused by an autoantibody to the aquaporin-4 water channel. The classic presentation of NMO is with the triad of optic neuritis, longitudinally extensive myelitis, and positive anti-AQP4 antibody, although a far wider range of manifestations are now recognized as part of NMOSD[1].

Neuromyelitis optica is typically found in patients somewhat older than those with multiple sclerosis (MS), with an average age of presentation of 41 years, and there is an even stronger female predilection (F:M 6.5:1) [2,3] . It is found more frequently in patients of Asian, Indian, and African descent [4]. NMO is characterized by bilateral optic neuritis and myelitis resulting in blindness and paraplegia. Although the two usually present concurrently, it is not uncommon for one to precede the other by up to several weeks [2]. Additionally, it is now recognized that some patients present with unilateral optic nerve involvement.

Although NMO was initially thought of as a monophasic illness, it is now evident that, as with MS, it is usually a relapsing-remitting disease with symptomatic events separated by many years [5].

Furthermore, NMOSD also encompasses non-neurological manifestations in anti-AQP4 antibody seropositive patients including systemic lupus erythematosus (SLE) and Sjogren's [6].In approximately 70% (sensitivity of 70-90%; specificity of 90%) of patients with established NMO, a specific immunoglobulin can be isolated (AQP4-IgG) which targets a transmembrane water channel (aquaporin 4) present on astrocyte foot processes abutting the limiting membrane [6].

The most common diagnostic test used for diagnosing NMOSD is MRI of the brain, orbits, and spinal cord. Imaging of the CNS is typically performed with gadolinium, and follow-up examinations are obligatory.[7]

A curative treatment for NMOSD does not currently exist. Intravenous methylprednisolone is the first-line therapy for treatment of acute NMOSD. Plasma exchange is used as a second-line therapy. Treatment in the acute or early stages aims to improve relapse symptoms and restore neurological function .[8]

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