A 32 years female presented with sequential weakness of left half, followed by right half of body during last two and half months without any remission. She had muscle spasms, urinary retention and constipation for same duration. She had periorbital pain but no visual or hearing problem. She had no preceding respiratory infection or vaccination. She had two abortions. She could not talk properly but followed instructions during examination. Her BP was 160/100 mmHg. She had pseudo-bulbar affect (with alternating euphoric and depressed moods), left sided upper motor facial weakness and torticollis (Fig. 1). She had spastic quadriparesis. Deep tendon jerks including jaw jerk were exaggerated. Planters were bilateral extensor. Ophthalmoscopy was normal. Her Hb was 9 gm%. ESR was 2 mm/1st hour. ANF, RF, VDRL and HIV tests were negative. CSF study showed cell count 4/cumm (all lymphocytes) and protein 42 mg%. CSF oligoclonal band was absent. MRI T2-weighted images showed multiple white-matter hyperintense lesions including periventricular (Fig. 2), centrum semiovale (Fig. 3), right cerebellar peduncle (Fig. 4), brainstem (Fig. 5) and corpus callosal (Fig. 6) lesions. In corpus callosum perpendicular extensions in radiating fashion towards cortex (Dawson’s fingers) were characteristic (Fig. 6). Plaques appeared hypointense on T1W images [hypointense lesions (black holes) are associated with evidence of axonal loss in addition to demyelination]. Axial T1W gadolinium enhanced MR images depicted several characteristic C-shaped or arc-like enhancements (Fig. 7-10). Corpus callosal and pericallosal hypointense lesions were characteristic (Fig. 10). FLAIR of T2W images showed periventricular high-signal-intensity lesions around the lateral ventricles (Fig. 11) with mild mass effect and a cavitary lesion with fluid attenuation at right subcortical white matter (Fig. 12). Marburg’s variant of MS was diagnosed.

Acute MS (Marburg’s variant) is a rare fulminant rapidly progressive demyelinating process with multifocal lesions involving cerebral hemispheres, brainstem or optic nerves. Otto Marburg first described it in 1906. Antibody mediated process appears to be responsible for most cases. It does not seem to follow infection or vaccination. It is associated with developmentally immature myelin basic protein. Usually there are no remissions. A brain tumour may be suspected in solitary, usually cavitary lesion. CSF oligoclonal bands are typically absent. Acute fulminant presentation, rapidly progressive course and MRI findings are diagnostic, despite the absence of CSF oligoclonal bands; but high degree of suspicion is needed for its diagnosis. High dose glucocorticoids, plasma exchange and cyclophosphamide may be beneficial.

Table 1: Demyelinating diseases


Reference