Progressive multifocal leucoencephalopathy with discrete involvement of pyramidal tract

A 35 year old gentleman manifested with subacute onset progressive left hemiparesis, left facial weakness, and dysarthria of 1 month duration. While in hospital, he developed dysphagia, and oesophageal candidiasis was detected. His haemogram, serum biochemistry, and urine analysis were normal. He was seropositive for human immunodeficiency virus (HIV) by enzyme linked immunosorbent assay (ELISA) with CD4 count of 240 cells/cc and CD8 of 945 cells/cc. The magnetic resonance imaging (MRI) of brain revealed a non-enhancing lesion (hypointense on T1W and hyperintense on T2W and fluid attenuated inversion recovery (FLAIR) images) essentially confined to white matter, extending from internal capsule on the right side along the pyramidal tract and crossing over to the left side at pontine level to involve the middle cerebellar peduncle (fig 1A and B).

MRI demonstrated an increased choline peak, reflecting accumulation of myelin breakdown products and rapid cell membrane synthesis. Cerebral spinal fluid (CSF) analysis showed 7 cells/cmm (five lymphocytes, two polymorphs), 34 mg % protein, and 64 mg % glucose. India ink stain did not reveal cryptococci. There was no immunological evidence of cryptococci, mycobacteria, toxoplasma, or syphilis. Stereotactic biopsy of the lesion from the internal capsule (close to the thalamus) showed features of demyelination of the white matter tracts with prominent foamy macrophages and oligodendrocytes. Occasional oligodendrocytic and astrocytic cell nuclei had basophilic large inclusions, while other astrocytes were large and atypical. Immunohistochemcnal localisation with polyclonal antibody to JC viral antigen (VP1) highlighted dark oligodendroglial inclusions and granular neuropil, suggesting a diagnosis of progressive multifocal leucoencephalopathy (PML) (fig 2A and B).

PML commonly involves the parietal subcortical white matter of brain in immuno-compromised individuals. MRI changes in brainstem often occur in association with supratentorial lesions and isolated brainstem involvement is rather rare. This patient had unusual MRI features of discrete involvement of the pyramidal tract. The role of the stage of the disease and host–agent interaction in its pathogenesis needs to be explored.1–5

Figure 2 Stereotactic brain biopsy of the lesion showing basophilic intranuclear oligodendroglial inclusions (arrows), and foamy histiocytes representing PML lesions in the demyelinated zones. HE × 600. (A) Immunohistochemical localisation of JC viral antigen in oligodendroglia (arrow) and in neuropil in granular pattern. Immunoperoxidase × 200. (B) An autopsied case of PML stained with JC viral antigen (VP1) highlighting dark oligodendroglial inclusion (arrow) and granular neuropil labelling. Immunoperoxidase × 300.

References