Leptomeningeal Gadolinium Enhancement in Autoimmune GFAP-Astrocytopathy

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A 46-years-old woman presented with urinary retention, headache, tremor, ataxia, mental slowness, and tetra-hyperreflexia following urinary infection. MRI showed signs of encephalomyelitis, with diffuse post-contrast-3D-FLAIR leptomeningeal enhancement, a common finding in several neuroinflammatory conditions\(^1\) (Figures 1 and 2). An extensive diagnostic work-up (serum and CSF neurotropic viruses, borrelia burgdorferi, treponema pallidum, serum anti-MOG and anti-AQP4 antibodies, ANA, ENA, ANCA, angiotensin-converting enzyme and alpha-chitotriosidase levels, quantiferon, total-body CT scan) was unrevealing. CSF analysis disclosed lymphocytic pleocytosis (204 leukocytes/MCL), high protein levels (123 mg/dl), CSF-restricted oligoclonal bands, and anti-GFAP antibodies (cell-based assay), leading to the diagnosis of autoimmune GFAP-astrocytopathy\(^2\). After receiving IV-methylprednisolone, the patient significantly improved.
FIGURE 1. 3D-FLAIR: occipital sulcal hyperintensities (A); linear perivascular radial hyperintensities in basal ganglia (B); linear and punctate leptomeningeal (arrow) and ependymal (empty arrow) brainstem hyperintensities (C). These findings could be related to raised CSF proteins and cells, leading to unsuppressed FLAIR CSF signal. STIR: C4-C5 and D1 lesions (D).
FIGURE 2. Sagittal and axial post-contrast MPRAGE 3D-T1: perivascular radial gadolinium enhancement in periventricular regions (A) and basal ganglia (B). Sagittal post-contrast SE-T1: linear leptomeningeal enhancement in whole spine (cervical tract detail, C), more evident in conus medullaris (D).

BIBLIOGRAPHY


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