Autoimmune glial fibrillary acidic protein (GFAP) meningoencephalomyelitis

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DESCRIPTION

A woman in her 20s presented with urinary retention, headache, cervicalgia and fever for 1 week. She was disoriented and had bilateral abducens palsy and papilloedema. Hypotonia of the lower limbs, appendicular ataxia, choreoathetotic movements of the trunk and neck rigidity were also noted. The clinical picture was interpreted as a meningoencephalomyelitis of probable infectious nature, given the neurological examination and presentation with fever. The main differential diagnosis included immune-mediated encephalitis, which could be related to a primary autoimmune disorder (neuromyelitis optica (NMO) spectrum disorders), a post-infectious (acute disseminated encephalomyelitis (ADEM)) or a paraneoplastic cause.

MRI showed T2 and fluid attenuated inversion recovery (FLAIR) hyperintensities in the periventricular white matter, basal ganglia and thalami, brainstem and dentate nuclei of cerebellum. There were also FLAIR hyperintense sulci on the posterior temporal and occipital lobes (figure 1A–D). Imaging of the spinal cord revealed a longitudinally extensive transverse myelitis (figure 1E,F). No enhancement was present. Lumbar puncture revealed lymphocytic pleocytosis and hyperproteinorrachia (protein 253.5 mg/dL (range 15–45), 18 cells/mm^3 with lymphocyte predominance, glucose 36 mg/dL (range 40–70)). Multiplex PCR was negative for viruses. A presumptive diagnosis of autoimmune meningoencephalomyelitis was considered. The patient was started on induction therapy with methylprednisolone IV (1 g/day for 5 days) and immunoglobulin IV (0.4 g/kg/day in a total of 20 g/day for 5 days). Cerebrospinal fluid (CSF) antibodies for IgG anti-glial fibrillary acidic protein (GFAP) were positive, and AQP4-IgG were negative. A paraneoplastic cause was investigated, and transvaginal ultrasonography found a right ovarian teratoma with 20 mm. A laparoscopic oophorectomy was performed, and maintenance immunotherapy was started with oral prednisolone (5 mg/day), with subsequent tapering. During hospitalisation, there was a progressive neurological and dysautonomic improvement. The patient started a physical rehabilitation programme, regaining the ability to walk and at discharge maintaining only a slight left abducens palsy without visual impairment. At 6 months'

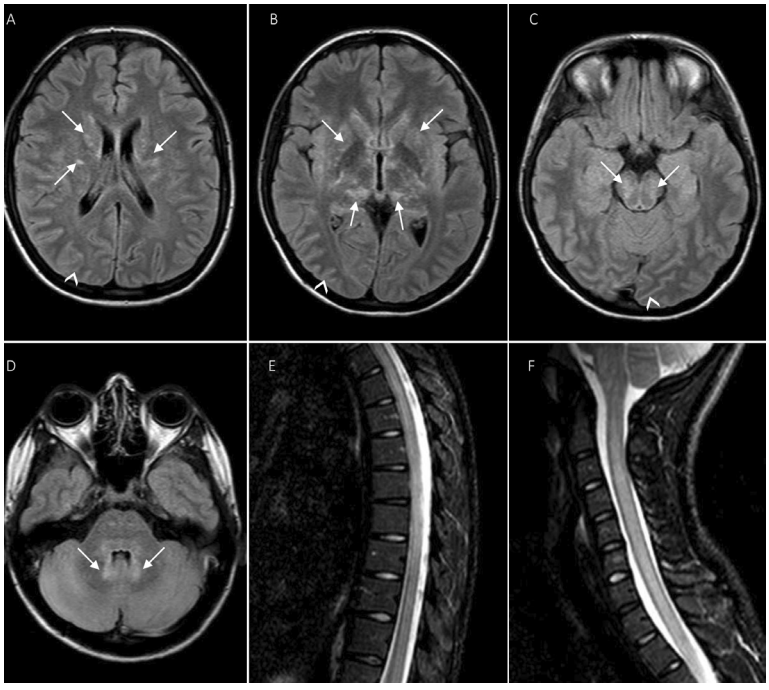

Figure 1 Brain MRI showing bilateral axial FLAIR hyperintensities (arrows) in the periventricular white matter (A), basal ganglia and thalami (B), brainstem (C) and dentate nuclei of cerebellum (D) and hyperintense sulci on the posterior temporal and occipital lobes (arrowheads A–C). Spinal MRI sagittal STIR showed a longitudinally extensive transverse myelitis (E–F), extending from the cervical spinal cord to the conus medullaris. FLAIR: fluid attenuated inversion recovery; STIR: short tau/TI inversion recovery.

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follow-up appointment, the patient had fully recovered and showed no neurological deficits.

Autoimmune GFAP astrocytopathy is an autoimmune inflammatory central nervous system disease for which a GFAP-IgG antibody serves as a biological marker.[1 2] Clinical findings of meningoencephalomyelitis as well as movement disorders and autonomic dysfunction are typical.[1–3] Brain imaging features range from absence of changes to T2 hyperintensities and T1 post-gadolinium enhancement, with a striking perivascular radial enhancement and a vasculitis pattern.[3] Also, a longitudinally extensive myelitis is the most common spinal finding, with the differential diagnosis including NMO spectrum disorders and other autoimmune inflammatory encephalomyelitis. GFAP-IgG detection is highly specific for inflammatory meningoencephalomyelitis when screened for in CSF. A paraneoplastic aetiology is found in some patients, with ovarian teratoma being the most common.[3 4] Tumour resection and immunotherapy are of paramount importance to improving clinical outcomes.[5]

Learning points

- Autoimmune GFAP astrocytopathy is a steroid-responsive central nervous system disorder presenting with symptoms and signs in the spectrum of meningoencephalomyelitis.
- Approximately one in four patients have a paraneoplastic cause, most commonly ovarian teratoma and tumour resection can be important to stop neurological progression.
- Despite the absence of post-gadolinium enhancement on imaging, a longitudinally extensive myelitis in a patient with glial fibrillary acidic protein (GFAP)-IgG detected in cerebrospinal fluid (CSF) with additional paraclinical findings (inflammatory CSF) warrants clinical suspicion.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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