Anti-GFAP Astrocytopathy: A Rare Inflammatory CNS Disorder Presenting as Meningoencephalitis and Polyradiculoneuritis

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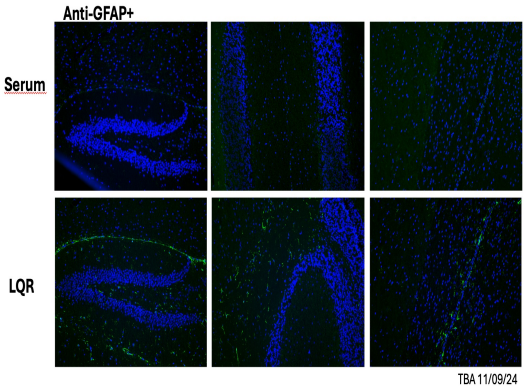
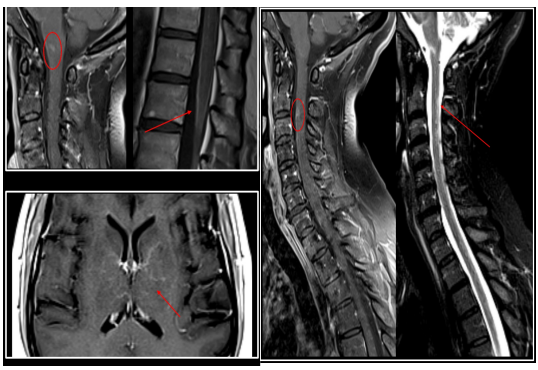
Background and Objectives

Anti-glial fibrillary acidic protein (GFAP) astrocytopathy is a rare inflammatory disorder of the central nervous system (CNS), characterized by the presence of GFAP IgG. Its clinical presentation is often heterogeneous, frequently mimicking viral or tuberculous meningitis. In rare cases, it can mimic neuroinfectious conditions (e.g., tuberculous meningitis) or central and peripheral neuroimmune disorders.



Acute phase MRI



Case report

A 49-year-old Indian woman presented to the emergency department with a one-month history of persistent headache, high-grade fever, nausea, vomiting, photophobia. Brain MRI showed isolated leptomeningeal enhancement. Empirical antibiotic and antiviral treatments were ineffective. Approximately ten days later, the patient developed progressive paraparesis. Electromyography (EMG) findings were consistent with acute motor axonal neuropathy (AMAN). Intravenous immunoglobulin (IVIG) therapy was initiated, resulting in initial clinical improvement.

About one month later, she developed a positive Lhermitte's sign. Cervical spine MRI revealed enhancement of both anterior and posterior spinal cord columns. A repeat lumbar puncture showed positivity for anti-GFAP IgG antibodies. A diagnosis of autoimmune GFAP astrocytopathy was made.

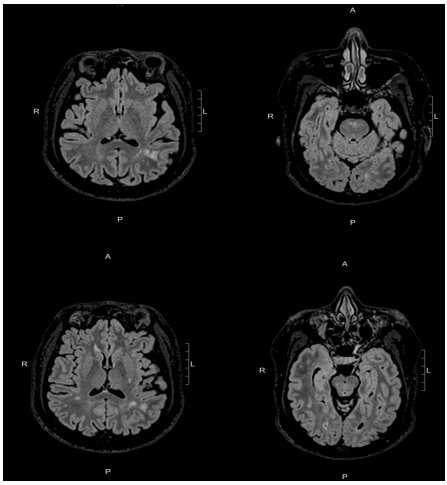
High-dose corticosteroid therapy was initiated immediately, resulting in a modest initial clinical improvement.

The patient was subsequently started on second-line therapy with rituximab, which led to further clinical improvement.

At 9-month follow-up, the patient was able to walk independently, with only residual weakness of the proximal lower limb muscles.

After corticosteroid discontinuation, she experienced a recurrence of severe headache. Due to the persistent GFAP-IgG positivity in cerebrospinal fluid and a cytokine profile suggestive of ongoing immune activation, the patient was considered for an additional cycle of immunomodulatory therapy.

Follow-up MRI



Discussion

Our case highlights that certain clinical presentations can be challenging, as they may mimic a wide range of neurological conditions, potentially leading to delayed diagnosis. The progression to paraparesis with AMAN-like features on EMG is an unusual manifestation and highlights the potential for overlapping central and peripheral nervous system involvement in GFAP astrocytopathy. The clinical response to immunotherapy supports an autoimmune etiology. The identification of anti-GFAP IgG in cerebrospinal fluid was crucial for diagnosis. Importantly, the disease course was relapsing, with symptom recurrence upon corticosteroid withdrawal. This underlines the need for long-term immunosuppressive therapy in selected patients

Conclusion

This case reinforces the importance of considering GFAP astrocytopathy in the differential diagnosis of atypical meningoencephalitis, particularly when initial infectious workup is negative and radiologic findings are non-specific. Early recognition and initiation of immunotherapy are essential to prevent neurological sequelae. Further studies are needed to define optimal treatment duration and to understand the mechanisms underlying relapses and long-term outcomes.

Reference

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