



HYPOTHALAMIC LESIONS & NMO IGG POSITIVITY RESTRICTED TO CSF



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OBJECTIVE

We present a unique and diagnostically-challenging case of Neuromyelitis optica spectrum disorders (NMOSD) in a patient who presented with somnolence and autonomic dysfunction consistent with disturbance of the hypothalamic-pituitary axis with MRI lesions restricted to the hypothalamic pathways. Serum NMO-IgG was negative but cerebrospinal (CSF) NMO-IgG was positive.

BACKGROUND

Neuromyelitis optica (NMO) is a severe demyelinating disorder of the central nervous system that predominantly affects the optic nerves and the spinal cord. NMOSD include other clinical syndromes associated with white matter lesions restricted to areas of high aquaporin-4 (AQP4) expression and can present with unique clinical features including encephalitic symptoms, intractable hiccups or nausea and vomiting, the syndrome of inappropriate antidiuretic hormone secretion and isolated optic neuritis (ON) or transverse myelitis. NMOSD are unified by the presence of serum and/or CSF NMO autoantibodies (Anti-AQP4 antibodies). Anti-AQP4 antibodies are 73% sensitive and 91% specific for the diagnosis of NMO. A review of the literature identifies only 3 out of 26 NMO patients with NMO-IgG positivity restricted to CSF.

CASE REPORT

22-year-old, previously healthy woman presented with a three-week history of progressive somnolence, hypothermia, bradycardia, hypotension and hyponatremia. Neurological exam was only significant for inability to maintain wakefulness. Patient did not have clinical signs of optic neuritis (ON) or myelitis.

MRI of the brain revealed T-2 hyper intensities involving the hypothalamus, thalami and anterior head of caudate with patchy contrast enhancement in the hypothalamus. MRI of the spine and orbits were normal. Antinuclear antibody was weakly positive at ANA 1:40, serum NMO-IgG was negative, however CSF NMO-IgG was positive. Other CSF studies revealed 14 WBC, no RBC, normal glucose and protein, mildly elevated IgG index at 0.8, 3 oligoclonal bands (OCB) and myelin basic protein (MBP) of 28.1.

Patient's wakefulness, hypothermia and hypernatremia improved after a five-day course of intravenous methylprednisolone 1-gram daily. Hypotension and bradycardia persisted for several weeks. She was able to return to work and school, however required modafinil to maintain wakefulness as outpatient. She did not have exacerbation on 3-month follow-up, however 4-month follow-up revealed new onset of transverse myelitis.

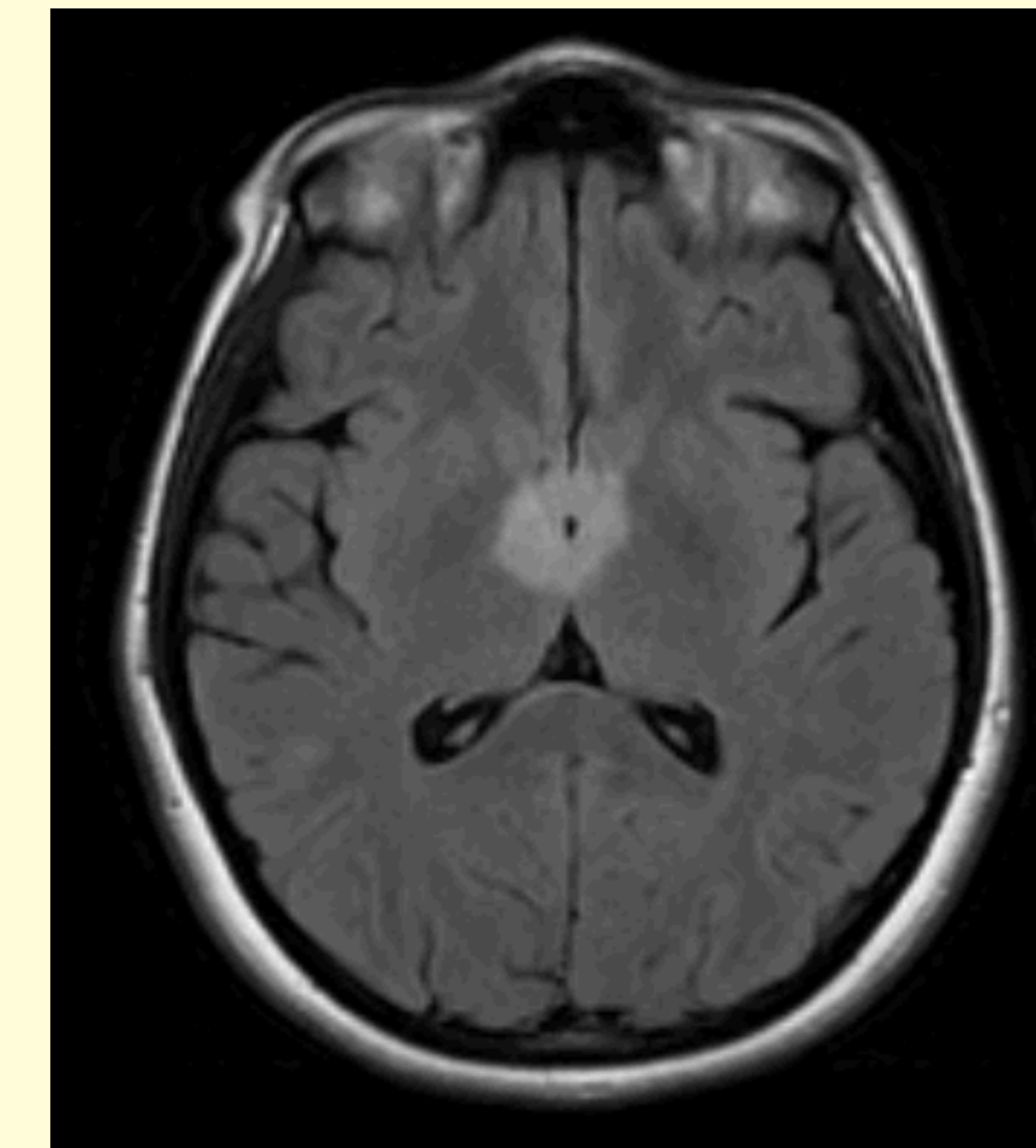


Fig 1: FLAIR changes in the thalamus, hypothalamus and anterior head of the caudate

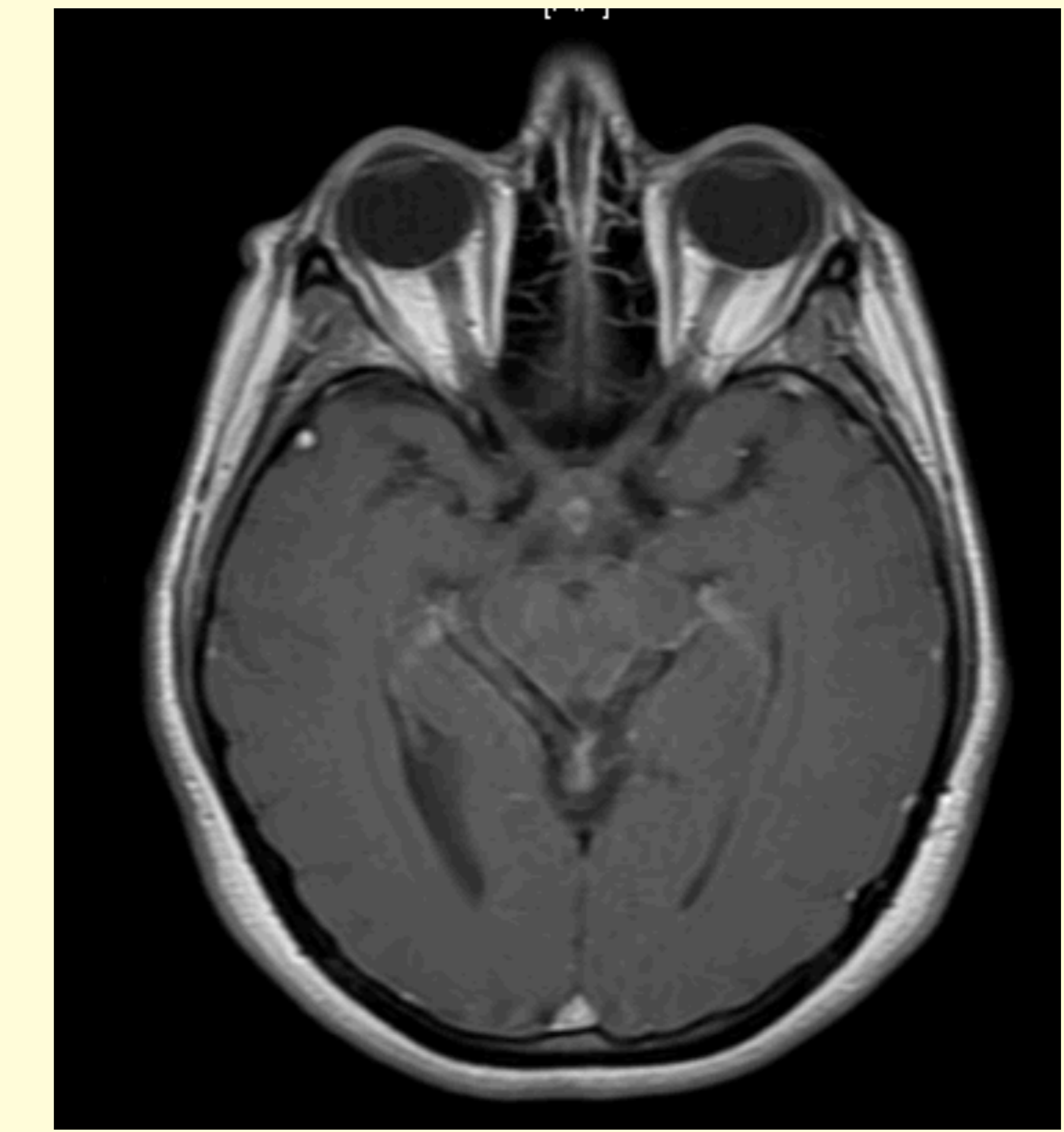


Fig 2: Patchy contrast enhancement in hypothalamus on T-1 with contrast

CONCLUSION

Hypothalamic-pituitary axis dysfunctions can be the presenting features of NMO in the absence of signs of ON or myelitis. While helpful when positive, serum NMO-IgG sensitivity is insufficient to exclude the diagnosis of NMO. If clinical suspicion persists, CSF NMO-IgG should be pursued.

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