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Case Report

Neuromyelitis optica, psychiatric symptoms and primary polydipsia: a case report

Josh Woolley, M.D., Ph.D.[a,*], Vanja C. Douglas, M.D.[b], Bruce A.C. Cree, M.D., Ph.D., M.C.R.[c]

[a]*Department of Psychiatry, University of California San Francisco, San Francisco, CA, USA*
[b]*Department of Neurology, University of California San Francisco, San Francisco, CA, USA*
[c]*Multiple Sclerosis Center, Department of Neurology, University of California San Francisco, San Francisco, CA, USA*

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Abstract

Neuromyelitis optica (NMO) is an aggressive demyelinating disease that typically affects the optic nerves and spinal cord. While it is increasingly recognized that cerebral lesions are common in NMO, there have been no reported cases of NMO presenting with psychiatric symptoms and polydipsia. We describe a patient with classic signs and symptoms of NMO who also demonstrated prominent psychiatric symptoms and polydipsia that were tied to his flares and resolved with treatment of his NMO. This case expands our understanding of possible presentations of NMO.

Keywords: Neuromyelitis optica; Psychiatric presentation; Psychogenic polydipsia

1. Introduction

Neuromyelitis optica (NMO), also known as Devic's syndrome, is an aggressive demyelinating disease characterized by selectivity for the optic nerves, longitudinally extensive cord lesions and absence of "clinical disease outside the optic nerve or spinal cord" [1]. However, it is increasingly recognized that both silent and symptomatic cerebral lesions are commonly found in NMO [2,3]. Antibodies to aquaporin-4 are often found in and are highly specific for NMO [2,4,5]. There have been no reported cases of NMO presenting with psychiatric symptoms. For the first time, we describe a patient with classic symptoms of NMO and positive aquaporin-4 antibodies who presented with prominent psychiatric symptoms and polydipsia that resolved with NMO treatment.

2. Case report

A previously healthy 40-year-old Asian man with no previous psychiatric history presented with new onset obsessionality, paranoia and severe insomnia. He had finger and facial tingling and a Lhermitte's sign, which were attributed to his psychiatric illness. After 6 months of treatment with risperidone and paroxetine his symptoms resolved, but 3 years later he developed quadriparesis. Magnetic resonance imaging (MRI) showed increased T2 signal with enhancement from T2 to T6 and increased T2 signal in the left midbrain and internal capsule. The patient was diagnosed with multiple sclerosis (MS) and treated with glucocorticoids with complete symptom resolution.

Over the following year, he had two more episodes of transverse myelitis that were treated with glucocorticoids. During the second episode, he developed right-sided optic neuritis (visual acuity was 20/200 on the right and 20/30 on the left with a right relative afferent pupillary defect). There was a right abducens palsy, facial sensation was decreased to light touch on the left and the tongue deviated to the right. Strength was decreased in a pyramidal distribution along

* Corresponding author.
E-mail address: josh.woolley@ucsf.edu (J. Woolley).

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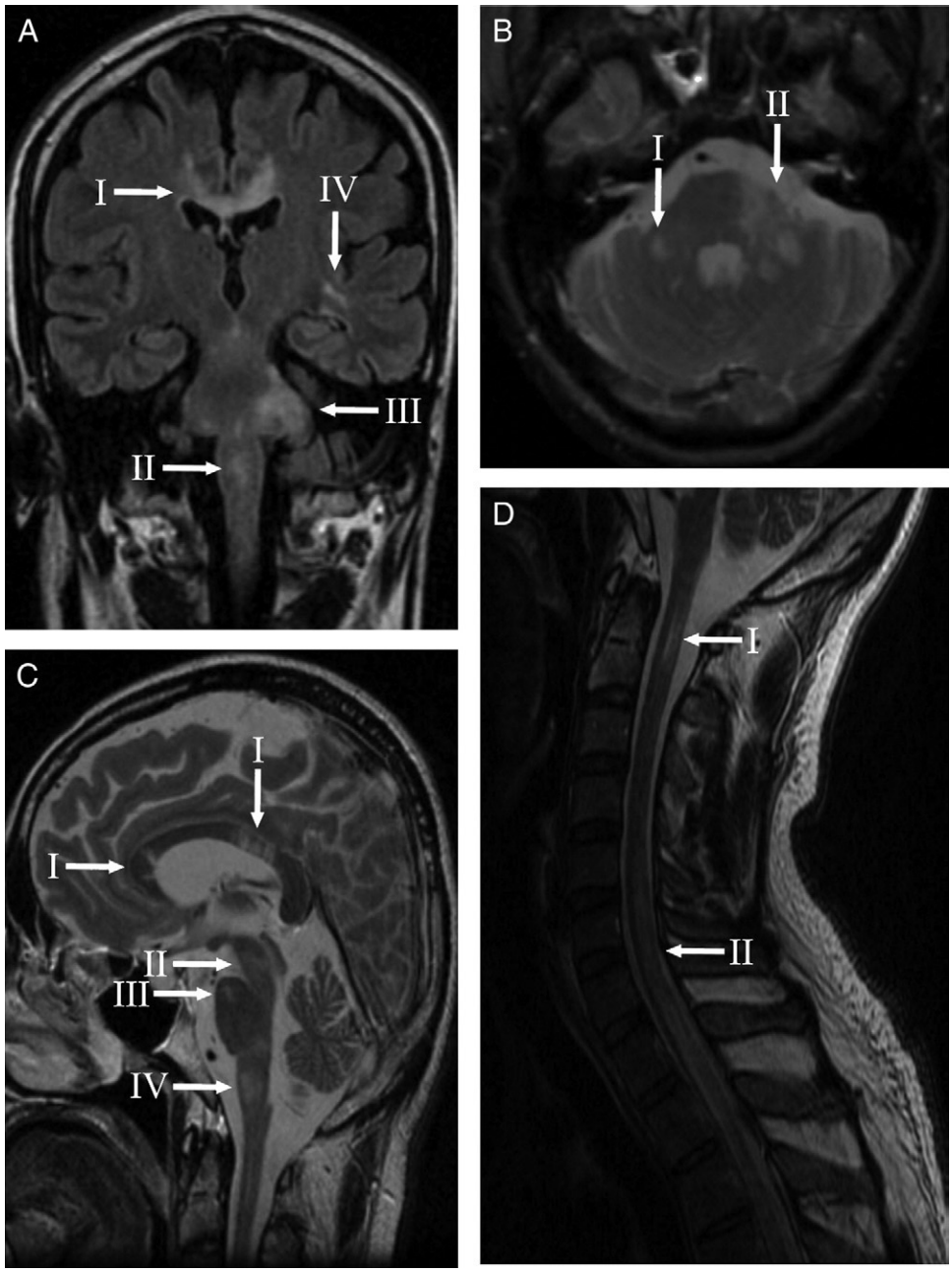

Fig. 1. Structural MRI obtained from the patient. White arrowheads indicate areas of abnormality. The coronal flair (A) demonstrates T2 prolongation in the corpus callosum (I), medulla (II), left pons (III) and left temporal lobe (IV). The axial T2 image (B) demonstrates T2 prolongation in the right brachium pontis (I) and left pons/brachium pontis extending into the cerebellar white matter (II). The sagittal T2 image (C) demonstrates T2 prolongation in the corpus callosum (I), midbrain (II), pons (III) and medulla (IV). The sagittal spinal T2 image (D) demonstrates T2 prolongation at the C2 level (I) as well as a longitudinal lesion from the C4/5 level extending into thoracic cord to at least T4 with associated volume loss (II).

with spasticity in all extremities. He had absent vibration sense and reduced cold sensation in all extremities with a T10 sensory level. Reflexes were trace in the upper extremities, 3+ at the knees and 4+ at the ankles with up-going toes bilaterally. He also developed urinary retention and steroid-induced hyperglycemia.

Cognitively, he was alert and fully oriented but scored 27/30 on a mini-mental status exam missing points for repetition, recall and serial seven's. He appeared extremely depressed, displaying a passive wish to die, anhedonia, hopelessness, and decreased energy and concentration, progressing to severe psychomotor retardation. When asked about his illness, he admitted that his doctors thought he had MS but that he thought his symptoms were due to "medications." He consistently complained of thirst and reported that despite drinking copious water it "wouldn't

stay" in his body. Serum sodium was 135 on admission. With unrestricted water intake, his urine output was 10 L/day with a urine osmolality of 107 mmol/kg and a urine sodium of 16 mmol/L in the face of a serum osmolality of 298 and serum sodium of 135. After restricting water intake, his urine osmolality rose to 394 and then to 773 mmol/kg within 24 h, urine sodium rose to 45 and then to 129 mmol/L and urine output rapidly fell into the normal range. Serum sodium was 136 mmol/L and serum osmolality was 289 mmol/kg. Therefore, he was diagnosed with primary polydipsia.

Lumbar puncture demonstrated a lymphocytic pleocytosis (23 WBCs, normal protein and IgG index and no oligoclonal bands). MRI showed multiple areas of T2 prolongation within brain white matter (Fig. 1A–C), at C2 and from C4/5 to the thoracic cord with volume loss (Fig. 1D) without reduced diffusion or enhancement. Laboratory studies for connective tissue, thyroid and vasculitic diseases were unremarkable. Antibodies to aquaporin-4 were positive at 1:1950, so his diagnosis was revised from MS to NMO. After treatment with glucocorticoids, interferon beta-1a, sertraline and aripiprazole, his symptoms improved. Five months later, he again developed transverse myelitis with cord enhancement that was treated with glucocorticoids and plasma exchange. He was started on rituximab [6] and over the next 3 years was tapered off all psychiatric medications and has been relapse free.

3. Discussion

This patient's initial symptoms were psychiatric and most likely derived from his inflammatory disease: his symptoms developed before treatment with glucocorticoids, returned during a later flare, resolved completely with time and did not require ongoing psychiatric treatment. Furthermore, schizophrenia onset is rare at age 40. While a bipolar illness may explain his first presentation, the co-occurrence of neurological symptoms with this flare and a re-occurrence of his psychotic symptoms in the setting of a later documented NMO flare make this explanation less likely. Psychiatric symptoms are well described in MS; depression has a prevalence of 30% compared to <10% in patients without MS [7]. Rarely, patients with MS can present with psychosis [8,9], catatonia [10], persecutory delusions and Capgras syndrome [11], and may be misdiagnosed with primary psychiatric conditions [8]. While psychiatric presentations are well described in MS, there have been no previous reports of patients with NMO presenting with psychiatric symptoms or polydipsia.

Hyponatremia in psychiatric patients is often due to the syndrome of inappropriate ADH secretion (SIADH) associated with SSRIs. However, patients with SIADH produce a normal volume of concentrated urine with high sodium; the fact that our patient produced a high volume of dilute urine with low sodium rules out SIADH and raises the specter of psychogenic polydipsia or diabetes insipidus (DI). DI is characterized by polyuria and polydipsia caused by inadequate urinary concentration due to three main defects: insufficient antidiuretic hormone (ADH) production in the hypothalamus (neurogenic DI), resistance of ADH action on the kidney (nephrogenic DI), or excessive water intake (primary polydipsia) due to damage to neural circuits regulating thirst (dipsogenic DI) or psychiatric derangement (psychogenic polydipsia) [12]. The fact that our patient's urine osmolality rose rapidly with water restriction indicates that ADH is present and the kidney responded normally, ruling out nephrogenic or neurogenic DI. Diagnosis of dipsogenic DI requires documented polyuria, normal urinary concentrating capacity and no psychiatric illness that could produce dysregulated water intake [13,14]. Given his prominent psychiatric symptoms, it is difficult to determine whether his excessive drinking was due to damage to neural circuits regulating thirst, his psychiatric illness or both. While hypothalamic lesions are relatively common in NMO [15–18] and he did demonstrate impaired thirst mechanisms, severe insomnia and supratentorial brain involvement, there was no evidence of hypothalamic involvement on MRI although this may have been missed due to slice thickness. Regardless, the fact that his polydipsia and quadriparesis resolved with immunosuppression suggests that his polydipsia was related to the NMO flare.

In summary, we report a patient with NMO and prominent psychiatric symptoms, including exacerbation associated primary polydipsia, which were likely due to his underlying inflammatory disease. This case demonstrates that patients with NMO may present with psychiatric symptoms that resolve with NMO treatment.

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