Hypothalamic Demyelination Process Complicated by Functional Neurological Disorder and Regressive Behavior

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Introduction

Key Points:

The hypothalamus plays a crucial role in regulating various homeostatic processes, including mood and stress response.

In Multiple Sclerosis (MS), hypothalamic demyelination can occur, leading to a range of non-localizing symptoms that may be mistaken for non-neurological issues.

This case study presents a patient with FND and acute regressive behavior coinciding with newly diagnosed hypothalamic demyelination.

The authors suggest that hypothalamic dysfunction may contribute to the development of FND symptoms in this case and could be an unusual presentation of MS.

Additional Resources:

National Multiple Sclerosis Society:
https://www.nationalmssociety.org/

Functional Neurological Disorder Society:
https://www.fndsociety.org/

Case Presentation, inpatient

24-year-old female without psychiatric history presented to urgent care with presyncope, chest pain and fatigue. She was treated with IV fluids and discharged after negative cardiac work-up. She presented 1 week later to the ED with similar symptoms and was discharged. Patient was concurrently diagnosed by PCP with major depressive episode after reporting stressors and scoring 13 on PHQ-9. She was started on bupropion and took one dose. Four days later, patient noted to be confused when contacted by phone to discuss results and family advised to bring her to the ED.

In the ED, patient was childlike and fearful with impairment in attention, concentration and memory. History revealed fatigue for 1 month and confusion for 1 week. Lumbar puncture was negative for oligoclonal bands, markers of autoimmune encephalitis and infection. Brain MRI showed multifocal abnormal signal, most prominently centered in the bilateral hypothalamic region, and numerous areas of white matter hyperintensity. EEG was normal.

Throughout the admission, the presentation varied from nonverbal, brady-phrenic and inattentive to childlike, despairing and anxious with bizarre vocalizations. Patient displayed tremulousness that sometimes evolved into myoclonic jerking with facial grimacing and tongue protruding. Neurology postulated that the decline in mental status, somnolence, and tremulousness were likely secondary to the hypothalamic lesions, but that the other symptoms could not be explained by MRI findings.

The patient received 5 days course of intravenous steroids. She returned to baseline mental status after the first dose and a family prayer session, followed by abrupt return to the aforementioned mental status. Acute anxiety was managed with clonazepam and supportive psychotherapy and psychoeducation were provided. Mental status improved after 14 days and she was transferred to acute rehabilitation prior to discharge home on day 35. While on rehab, her processing speed, memory anxiety improved.

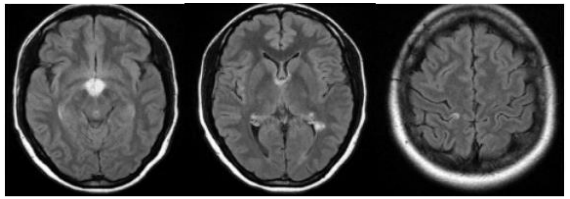
Outpatient course

Patient reported significant improvement in functionality on the second follow up. She tolerated well tapering and being of the Prednisone. Repeat brain MRI showed improvement and no new changes.

From the last follow up, patient continues to improve and is back to work as a researcher. Patient follows up with Rheumatologist due to swollen joints (ankles and shoulders) and is on Celebrex.

Latest diagnostic impression is “Inflammatory demyelinating syndrome of uncertain etiology, with significant improvement”

MRI brain 2/2/24: bilateral hypothalamic demyelination



Discussion

This was an unusual presentation whose clinical phenotype suggested autoimmune encephalitis, however, imaging was more suggestive of a demyelinating disorder. Patient had numerous symptoms that could not be explained by the MRI and were thought to represent superimposed FND. MS and FND are diagnostically challenging conditions which often co-occur and share similar features but have been poorly studied. Young age, major life stressors, inadequate social support, and poor coping skills could make FND a hidden confounder, but it is also important to consider whether unusual or non-focal symptoms are an early manifestation of neurological illness. In this case, it was challenging to parse out which symptoms were secondary to hypothalamic lesions versus FND.

Reference: Walzl D, Solomon AJ, Stone J. Functional neurological disorder and multiple sclerosis: a systematic review of misdiagnosis and clinical overlap. J Neurol. 2022 Feb;269(2):654-663.