Neurosarcoidosis and Psychosis, a Case Study

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Introduction

Sarcoidosis is a systemic disease characterized by noncaseating granulomatous inflammation with an estimated incidence of 35.5 per 100,000 person-years in African Americans and 10.9 per 100,000 person-years in Americans of European descent.[1] Nervous system involvement occurs in 5-16% of cases and common presenting signs often involve the optic nerve, other cranial nerves, the spinal cord, or brainstem. However, less common presentations include meningeal signs, cognitive decline, hydrocephalus, and hypothalamic-pituitary-adrenal axis dysfunction. Delirium and psychosis are estimate to manifest in 1% of individuals with sarcoidosis. [4]

Case

We describe an African-American male in his 30's with a history of obstructive hydrocephalus, seizure disorder, obstructive sleep apnea, and morbid obesity who presented with a two month history of intermittent paranoia and visual hallucinations progressing in intensity, as well as daytime drowsiness. Though previously high-functioning and living independently, he began experiencing headache and blurry vision three years prior to our encounter and was subsequently diagnosed with hydrocephalus. A ventriculoperitoneal shunt was placed, and thought it required several corrective interventions over that period, the patient experienced no remote perceptual disturbances until two months prior to presentation.

Family collateral information indicated that the patient had become progressively lethargic over this time period, often waking only to eat meals. He was subsequently assessed by a sleep specialist and diagnosed with sleep apnea and obesity hypoventilation syndrome, for which at-home CPAP was initiated.

His psychotic symptoms were reported to onset shortly after a ventriculoperitoneal shunt revision at an outside institution, though outside clinicians had since determined that the patient's shunt continued to function normally.

On exam, he patient exhibited disorientation and drowsiness, though unit staff reported that the patient was intermittently paranoid and overtly agitated. Thought processes were organized, though cognitive assessment was somewhat limited by poor attention and concentration.

A neurological assessment identified no focal neurological deficits. EEG showed only diffuse slowing of waves, and CT head revealed a functioning shunt without signs of hydrocephalus.

In addition to the patient's psychiatric symptoms, he exhibited signs of hypercapnic respiratory failure and subsequent CT of the thorax revealed significant mediastinal and hilar adenopathy consistent with sarcoidosis vs malignancy. Mediastinal lymph node biopsy was obtained and showed granulomatous processes indicative of sarcoidosis. As systemic sarcoidosis had been confirmed, the index of suspicion that the patient's inflammatory pathology was involving the central nervous system was high, and so intravenous corticosteroids were administered for a presumptive diagnosis of neurosarcoidosis (NS) immediately. Improvements in orientation and psychosis could be observed within two days following initiation of intravenous methylprednisolone, and after five days the patient was transitioned to oral steroids and discharged to home.

Discussion

Neurosarcoidosis, though rare in the general population is an important and occasionally overlooked neuropsychiatric condition that leads to significant morbidity and mortality. In one postmortem study, only 50% of the patients with observable nervous system involvement had been diagnosed with NS by the time of death. [4] Another study reviewed 268 patient diagnosed with first-episode schizophrenia, of which 2 patients had NS. [2] Due to the varied clinical presentations of NS and the lack of specificity of commonly used imaging and laboratory studies, the diagnosis of NS is often "presumptive" or "probable" in nature. [4] Our patient's presentation lacked the more typical neurological signs often associated with NS, and so excluding alternative diagnoses and obtaining evidence of systemic inflammatory involvement led to the probable diagnosis in this case. Our patient's history was significant for obstructive hydrocephalus and seizure disorder, both of which are associated with NS. [3]

While further diagnostic studies such as MRI and CSF analysis were considered, the multidisciplinary treatment team elected to initiate treatment without delay. Though MRI is sensitive in identifying white matter disease, the white-matter changes observed in NS are nonspecific and difficult to distinguish from other vascular, infectious, or inflammatory conditions. MRI findings have not been shown to correlate with either the severity of disease or the response to treatment. Similarly, estimated specificity and sensitivity of CSF angiotensin converting enzyme vary greatly, and other findings of pleocytosis and elevated protein are nonspecific. [2]

Works Cited

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