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CASE REPORT

Neurosarcoidosis Presenting as Normal Pressure Hydrocephalus in a Patient with Pulmonary Sarcoidosis

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Abstract

A gentleman in his early 50's with past medical history significant for biopsy confirmed pulmonary sarcoidosis presented with acute onset confusion, gait disturbance and bowel/urinary incontinence. Brain imaging was consistent with normal pressure hydrocephalus and CSF studies supported the suspicion of neurosarcoidosis. He received treatment with a prolonged course of steroids with good effect. We discuss a rare case of normal pressure hydrocephalus as a presenting feature of neurosarcoidosis and the rationale of starting empiric treatment for probable diagnosis of the same.

Keywords: Neurosarcoidosis, Normal pressure hydrocephalus, Extra-pulmonary sarcoidosis, Hydrocephalus, Pachymeningeal enhancement

1. Background

Sarcoidosis is a multi-system granulomatous inflammatory disorder of unknown etiology.[1] 5% of the patients with sarcoidosis can develop neurosarcoidosis, however only half of them present with neurological complaints.[2] Neurosarcoidosis can affect both the central and peripheral nervous system. Manifestations are based on the type of structure affected. Hypothalamic or pituitary involvement causes neuroendocrine manifestations; brain involvement results in seizures, cognitive or behavioral changes; spinal cord involvement as radiculopathy or myelopathy, meningeal involvement as aseptic meningitis; and neuronal involvement as peripheral neuropathy. Neurosarcoidosis can also cause communicating or non-communicating hydrocephalus.[3] There is a lack of data about hydrocephalus as a presenting feature of neurosarcoidosis. Diagnosis of neurosarcoidosis is considered in patients with sarcoidosis who develop neurological symptoms after excluding infectious and malignancy related causes.

2. Case presentation

A 52 year old gentleman with past medical history significant for stage III chronic kidney disease, type 2 diabetes mellitus, hypertension, pulmonary sarcoidosis, renal cell carcinoma status post radiation, gastroesophageal reflux disease, obstructive sleep apnea, attention deficit hyperactivity disorder, anxiety presented to the hospital with confusion for two days. His home medications included losartan, alprazolam, methylphenidate, and dexamethasone. Patient carries a diagnosis of biopsy proven pulmonary sarcoidosis on dexamethasone 6 mg daily and was last seen by his pulmonologist two months ago. He notably ran out of dexamethasone a few days prior to presentation. Most of the history at presentation was obtained by his mother whom he lives with and who brought him to the hospital. Patient was apparently doing well until two days

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ago when the mother noticed him to be confused, unaware of his surroundings and endorsing bizarre behavior like opening the doors of all the cupboards in the house and leaving the lights on in every room he entered. His mother also mentioned that he had changes in gait which she described as 'short stepped gait' resulting in three falls. She also endorsed two episodes of bowel incontinence, occasional urinary incontinence, episodes of diffuse headache associated with nausea and vomiting and increased lethargy since the last few weeks.

On presentation he was afebrile, blood pressure 156/107 mmHg, heart rate 96/minute and saturating at 96% on room air. On examination, the patient was alert and oriented to self and place, involving in tangential talks. On exam, head was atraumatic, normocephalic, Montreal cognitive assessment score of 21 and no focal neurological deficits. Neck examination was normal and clear breath sounds were auscultated bilaterally.

Laboratory results were significant for sodium of 125 mEq/L and CT of brain without contrast (Fig. 1) was suggestive of interval enlargement of ventricular system concerning for normal pressure hydrocephalus. This was followed by MRI brain with contrast (Figs. 2 and 3) revealing new ventricular system dilatation with mild transependymal edema consistent with hydrocephalus with pachymeningeal enhancement. Due to his imaging findings, he underwent lumbar puncture and cerebrospinal fluid (CSF) was sent for cell count, protein, glucose, IgG, angiotensin converting enzyme (ACE) levels, gram stain, bacterial, viral and fungal cultures. Initial CSF fluid analysis revealed normal opening pressures with elevated white blood cells and elevated protein. Patient was evaluated for placement of ventriculo-peritoneal shunt, but a consensus was reached against the same due to normal CSF opening pressure. Hence, he was started on a guideline directed high dose of prednisone 60 mg for probable diagnosis of neurosarcoidosis. Patient showed dramatic clinical improvement within a week and was discharged home on the same prednisone dose.

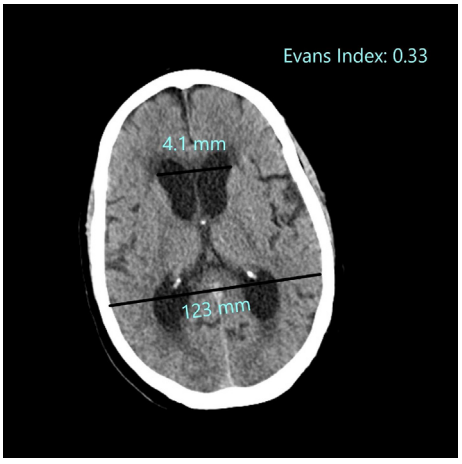

Fig. 1. Axial brain CT showing interval enlargement of the ventricular system concerning for normal pressure hydrocephalus. Evans index 0.33.

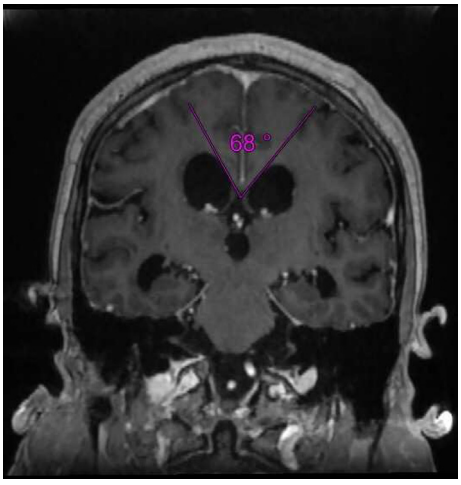

Fig. 2. Coronal brain MRI with contrast showing ventricular system dilatation with mild transependymal edema consistent with hydrocephalus. Callosal angle of 68°.

3. Differential diagnoses

Differential diagnoses for our patient have been discussed in (see Table 1).

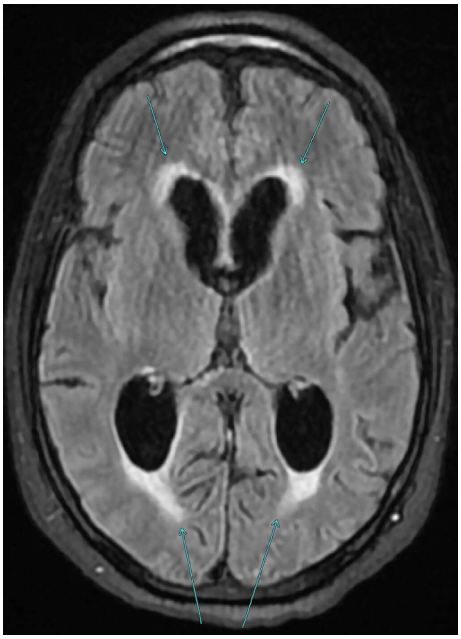

Fig. 3. T2 FLAIR axial brain MRI showing ventricular system dilatation with pachymeningeal enhancement, and no evidence of PRES.

Table 1. Differential diagnoses.

Differential diagnosis	Supporting features	How diagnosis was excluded
Posterior fossa lesion	Gait abnormalities and signs of raised intracranial pressure at presentation.	No evidence of the same on imaging.
Autoimmune encephalitis	Confusion, bizarre behavior. CSF modest elevation of protein (<100 mg/dL), mild to moderate lymphocytic pleocytosis, elevated IgG index, and/or the presence of oligoclonal bands.	No evidence of signal hyperintensities on fluid-attenuated inversion recovery (FLAIR) or T2-weighted images in affected brain regions.
Acute disseminated encephalomyelitis	Multifocal neurological abnormalities with CSF white blood cell count <100 cells/mL, lymphocytic pleocytosis, and a mildly elevated CSF protein.	Absence of one or more supratentorial or infratentorial demyelinating lesions on brain MRI. Presence of oligoclonal bands in CSF. It is a diagnosis of exclusion.
Wernicke's encephalopathy	Presence of confusion, gait abnormalities.	No history of chronic alcohol use and did not fulfill the Caine criteria for diagnosis.

CSF- Cerebrospinal fluid, MRI- Magnetic resonance imaging, IgG- Immunoglobulin G.

The definitive diagnosis of neurosarcoidosis requires biopsy proven non-caseating granulomas in the affected nervous system tissue. However, probable diagnosis can be made in patients with evidence of systemic sarcoidosis who have CSF studies or MRI findings to support the same. In our patient, MRI showed pachymeningeal enhancement indicating inflammation and CSF analysis was significant for elevated protein, white blood cells, IgG and presence of oligoclonal bands.

4. Treatment

The first-line treatment for the neuro-ophthalmologic manifestations of sarcoidosis is systemic corticosteroid therapy. Our patient was started on prednisone of 60 mg daily for 4 weeks in keeping with recommended guidelines of 0.5−1 mg/kg/day.

5. Outcome and follow up

Patient demonstrated dramatic clinical improvement following systemic corticosteroid therapy. He continued to remain on prednisone 60 mg daily for a period of 4 weeks, followed by a taper regimen of 5 mg/day every two weeks. At 3 month follow up visit, he had tolerated his medications without adverse effects. He had begun to have noticeable changes in behavior and energy levels after 2 weeks. His gait had significantly improved, although he occasionally endorsed minimal lower extremity weakness. Resolution of bowel and bladder incontinence was also relatively delayed. At the end of 6 months, he continues to remain in remission with complete return to baseline functionality in terms of mental status, gait and bowel/bladder control.

6. Discussion

The neurological manifestations of sarcoidosis comprise a spectrum of diverse manifestations. Increased awareness is needed regarding the less common presentations of neurosarcoidosis like hydrocephalus. Keeping a low index of suspicion for its diagnosis can help initiate early treatment and decrease associated morbidity and mortality. Defining the certainty of diagnosis as-possible, probable or definite may aid in formulating a plan of action.

In a case report of 61-year-old female presenting with idiopathic normal pressure hydrocephalus, neurosarcoidosis was diagnosed based on abnormalities in CSF and abnormal enhancement of caudal equine, leptomeninges of brainstem and spinal cord on MRI. Mediastinal lymph node biopsy confirmed sarcoidosis.[4] In another case of cryptogenic hydrocephalus, neurosarcoisosis was diagnosed by caudal equine biopsy due to lack of systemic involvement and it required multiple ventriculoperitoneal shunt placements due to its recurrent course. Patient was also managed with steroids and mycophenolate mofetil, due to lack of significant improvement with empirical trial of steroids.[5] A case of sensory manifestation of neurosarcoidosis in a middle aged man has been reported who presented with progressive inability to walk. He had clinical and electrophysiological features of chronic sensory lumbosacral polyradiculopathy with nerve root biopsy characteristic of sarcoidosis. It was a rare case of cauda equina neurosarcoidosis with evidence of involvement of sensory fibers only. He showed early clinical improvement with oral corticosteroids.[6]

Neurosarcoidosis is a rare and difficult diagnosis. Corticosteroids are the mainstay of treatment, despite the absence of randomized controlled trials of treatment and recommendations being based on retrospective case series, anecdotal experience, and randomised controlled trials in pulmonary sarcoidosis.[7] For treatment of neurosarcoidosis, prednisone, 0.5−1 mg/kg/day, is initially prescribed for 2−4 weeks, before a slow taper is begun as the patient's

symptoms and examination are monitored. One useful approach is to allow the patient to stabilize or improve with four weeks of treatment and then to decrease the prednisone dose by 5 mg every two weeks as tolerated by symptoms. Methotrexate or hydroxychloroquine may then be added, especially if the response to steroids is inadequate.

Learning points

- Neurosarcoidosis develops in 5% of the patients with sarcoidosis and although rare can cause communicating or non-communicating hydrocephalus.
- A possibility of neurosarcoidosis should be considered in patients with history of pulmonary sarcoidosis presenting with nonspecific neurological manifestations including behavioral changes.
- Prednisone 0.5–1 mg/kg/day over 2–4 weeks should be started for patients with definitive or probable diagnosis of neurosarcoidosis and they should be followed up to monitor for clinical improvement.

Conflict of interest

No conflict of interests to report. No data needed for data availability statement. No external funding. No COIs to disclose. No ethics approval statement requirement. No patient information involved. No clinical trial involved.

No consent was needed since subject's identity has been masked.

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