Acute Hydrocephalus: Unsual Presentation of Neurosarcoidosis

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Citation

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Abstract

Objective: We report a case of hydrocephalus as manifestation of neurosarcoidosis in absence of pulmonary features. The diagnosis and management could be a challenge in such atypical presentation of hydrocephalus in previously healthy young adult. The goal is to emphasize on importance to recognize atypical presentation in timely manner as neurologic involvement is significant cause of mortality and morbidity.

INTRODUCTION

Sarcoidosis is a multisystemic disorder of unknown etiology characterized by an accumulation of noncaseating epitheloid granuloma. The most commonly involved organs are lungs in about 90 percent of patients. Other involved organs skin,eyes and lymphnodes.

Neurosarcoidosis has been described in 5% of patients with sarcoidosis. Neurosarcoidosis has wide clinical presentations; most comman being cranial neuropathy, but ranges from encephalopathy, aseptic/chronic meningitis, seizures, panhypopituitarism to spinal cord dysfunction. Hydrocephalus in patients with neurosarcoidosis is extremely rare especially in absence of any other systemic findings. The diagnosis can be challenging in such an atypical presentation of hydrocephalus in previously healthy young adult.

CASE REPORT

A 46-year old man of African origin was admitted for evaluation of altered mental status. He had past medical history of Hypopituitarism, seizures, left eye blindness. His hypopituitarism was secondary resection of pituatry gland year ago at different facility. He was on desmopressin, prednisone and thyroid supplements.

Physical examination: P 62, BP 90-110/50's, RR 10 with shallow breathing, temp 92.5.

Neurologic examination revealed unremarkable motor and sensory exam but DTRs were hyperreactive.

Labs: White cell count of 5 with 63% neutrophils, calcium 10.1, hemoglobin 10.8, sodium 153, ESR 34, ACE level 74(12-68 units/L).

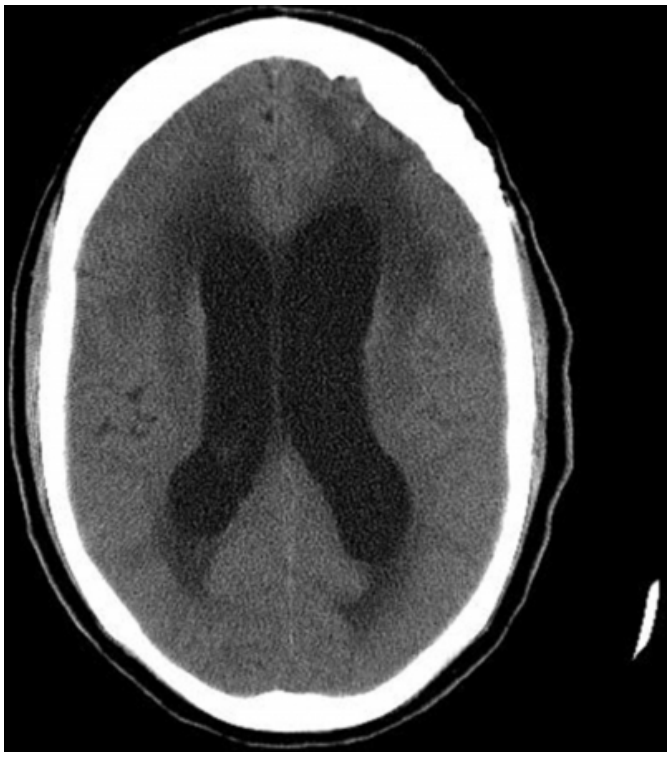
Lumbar puncture: protein >700, glucose 53, WBC 45 (48% PMN, 41% lymphs, 11% monocytes), protein 781 (15-45mg/dl).

Chest x-ray: No evidence of hilar adenopathy or prominent pulmonary markings.

CT head showed communicating acute hydrocephalus and areas of low attenuation in periventricular region.

Figure 1

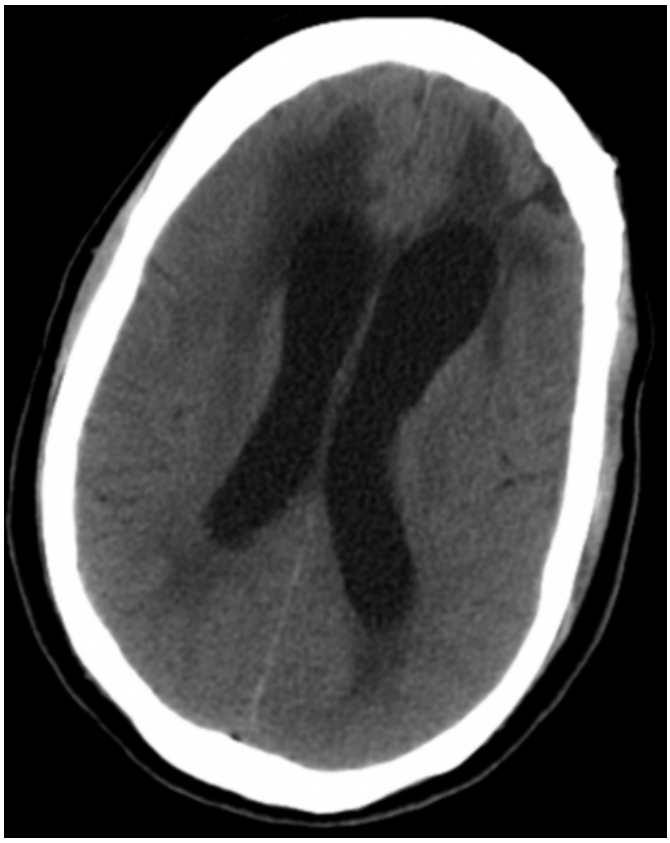
Panel 1: Severe Hydrocephalus at presentation



The follow up on a brain biopsy (pituitary gland) performed one year ago at a different facility showed multiple noncaseating granulomas. The sections of dura also revealed small noncaseating granulomas. The biopsy was negative for acid-fast bacilli, malignancy or fungal infection. The patient was treated with IV Corticosteroid and his mentation returned to the baseline the next day. The repeat CT scan of the head showed interval decrease in hydrocephalus, hence shunting procedure was not necessary.

Figure 2

Panel 2: Interval decrease after steroids.



DISCUSSION

This case is remarkable given its extreme rare occurrence. As review of the literature revealed only one prior report of acute hydrocephalus as presentation of neurosarcoidosis in absence of any systemic features. Secondly neurologic involvement is a significant cause of mortality and morbidity in patients with sarcoidosis and poses diagnostic conundrum of isolated neurologic and neuroendocrine symptoms as sole presenting abnormality in absence of pulmonary or other systemic findings. However with timely diagnosis, can be successfully managed with corticosteroid therapy. Corticosteroids remain the mainstay of treatment and patients may improve rapidly but can decompensate requiring emergent shunting.

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