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A Rare Case of Neurosarcoidosis Presenting as Severe Hypothermia

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Funds Collection G

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Patient: Male, 41
Final Diagnosis: Neurosarcoidosis
Symptoms: Fecal incontinence • hypothermia • loss of balance • loss of consciousness
Medication: —
Clinical Procedure: —
Specialty: Family Medicine

Objective: Unusual clinical course

Background: Sarcoidosis is a systemic disease of unclear etiology with an estimated prevalence of 10 cases per 100 000 population. Although sarcoidosis primarily affects the pulmonary system, it affects the nervous system in 5–10% of patients with the condition.

Case Report: This article describes a case of neurosarcoidosis with the primary presenting finding of hypothermia. A 41-year-old African-American man with a past medical history significant for hypertension, asthma, and anemia, presented to the Emergency Department after being found unresponsive at home. The patient was found to have a significantly low temperature of 27.0°C on presentation. He was subsequently diagnosed with sarcoidosis by mediastinal biopsy, which revealed non-caseating granulomas with variable stages of sclerosis, consistent with long-standing sarcoidosis. In a subsequent admission, the patient presented with hyperthermia, further suggesting hypothalamic involvement as part of the neurosarcoidosis.

Conclusions: Neurosarcoidosis is a rare variant of sarcoidosis that has varying presentations, including headaches, seizures, ataxia, visual disturbances, loss of sensation, and other neurologic abnormalities. By presenting this case, which to our knowledge is the first to report neurosarcoidosis presenting as hypothermia, our aim is to use it as a teaching moment and for further discussion on clinical presentations and management of such patients.

MeSH Keywords: Hypothalamus • Hypothermia • Sarcoidosis

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/901973

Background

Sarcoidosis is a systemic disease of unclear etiology with an estimated prevalence of 10 cases per 100 000 population. Sarcoidosis appears to affect African-Americans most, with a 2.4% lifetime risk compared to only 0.85% in whites [1,2]. Although sarcoidosis primarily affects the pulmonary system, it affects the nervous system in 5–10% of patients with the condition [3]. Neurosarcoidosis is a very rare variant of sarcoidosis involving structures within the central nervous system. Depending on the central nervous system structure involved, neurosarcoidosis can present with cranial nerve dysfunction, headaches, seizures, ataxia, vertigo, diplopia, or other neurologic disturbances [4,5]. In instances where pituitary structures are affected, patients can also present with polyuria, polydipsia, and menstrual irregularities [4].

Magnetic resonance imaging (MRI) is invaluable in confirming the diagnosis of neurosarcoidosis. Common MRI findings in patients with neurosarcoidosis include cortical lesions, cerebral white matter lesions, or involvement of the leptomeningeal, spinal, cortical, sellar, suprasellar, periventricular, or corpus callosum regions [6]. CT imaging remains far superior to X-ray in identifying subtle lesions. Biopsy of nerve tissues, palpable lymph nodes, or superficial skin lesions remains the criterion standard to confirm the diagnosis [7].

Neurosarcoidosis can follow different courses, including self-limiting, chronic, relapsing, and progressive ones, and the prognosis therefore varies according to individual case. Patients with leptomeningeal, spinal, or parenchymal involvement appear to have the worst prognosis [5, 8]. Although no cure exists for neurosarcoidosis, approximately 65% of patients respond to treatment [7]. Corticosteroids and immunomodulators remain the primary treatment. Despite therapy, neurosarcoidosis remains potentially fatal, with close to 10% mortality in affected individuals [7].

Case Report

A 41-year-old African-American man with a past medical history significant for hypertension, asthma, and anemia was brought into the emergency department (ED) after being found face down and unresponsive in a warm room at home. According to family members, the patient was complaining of “loss of balance” and fecal incontinence beginning 4–5 days prior to admission, but he attributed these symptoms to “a cold” and never visited a physician. Upon arrival in the ED, the patient’s vital signs were significant for a blood pressure=130/90, heart rate=56, respiratory rate=16, and a temperature=27.0°C. The patient’s Glasgow Coma Scale score was 3 and physical examination revealed lack of gag reflex, lack of oculocephalic reflex, and decreased deep tendon reflexes.

The patient was intubated, started on intravenous steroids, broad-spectrum antibiotics, and acyclovir, and transferred to the medical intensive care unit (MICU). He was treated medically as well as via passive and active warming techniques, including the Arctic Sun Temperature Management System. Initial laboratory studies revealed Sodium=134, Potassium=3.9, Chloride=95, Carbon dioxide=27, Glucose=184, BUN=9, Creatinine=0.8, WBC=2.7, CPK=56, Troponin <0.017, and TSH=0.8. Chest X-ray revealed no acute process and electrocardiogram (EKG) demonstrated Osborne waves but was otherwise unremarkable. Lumbar puncture revealed an opening pressure of 43 and cerebrospinal fluid (CSF) evaluation revealed moderate polymorphonuclear cells (PMNs), but was negative for infectious etiologies. Protein count was elevated, while glucose was decreased, likely reflecting the leptomeningeal involvement. CT-head showed cerebral edema and an enlarged 4th ventricle (Figure 1). A follow-up MRI revealed extensive leptomeningeal enhancement, associated with vasogenic edema and cortical swelling (Figure 2).

On hospital day 2, the patient’s condition improved and his vitals stabilized, with temperature in the 36.9–37.8°C range. He was extubated on hospital day 3 and was oriented only to person. Physical examination after extubation revealed no focal deficits. An EEG was conducted and revealed changes suggestive of moderate-to-severe encephalopathy. A CT-chest demonstrated enlarged lymph nodes throughout, as well as bilateral hilar and mediastinum in the pretracheal, precarinal, subcarinal, and pre-vascular spaces. Nodularity along bilateral perihilar fissures was also noted. The patient underwent mediastinal biopsy, which revealed non-caseating granulomas with variable stages of sclerosis, consistent with long-standing sarcoidosis (Figure 3A, 3B). The patient slowly recovered and on hospital day 6 he was alert and oriented times 3. He was diagnosed with neurosarcoidosis and discharged in stable condition. He presented to our facility with heat stroke secondary to hyperthermia 3 months after this admission, but was subsequently lost to follow-up.

Discussion

As the epicenter of primary temperature control in the body, involvement of the hypothalamus can lead to labile temperature, as was noted in our patient’s persistent hypothermia during initial presentation and hyperthermia during subsequent presentation. In both cases, our patient regained the ability to regulate bodily temperature control after his initial presentation, suggesting a partial insult of the hypothalamus rather than complete involvement of the structure. The insult may have been secondary to ischemic injury to the hypothalamus, as vasogenic edema can lead to compression of the vessels supplying the hypothalamus. The apparent cortical edema

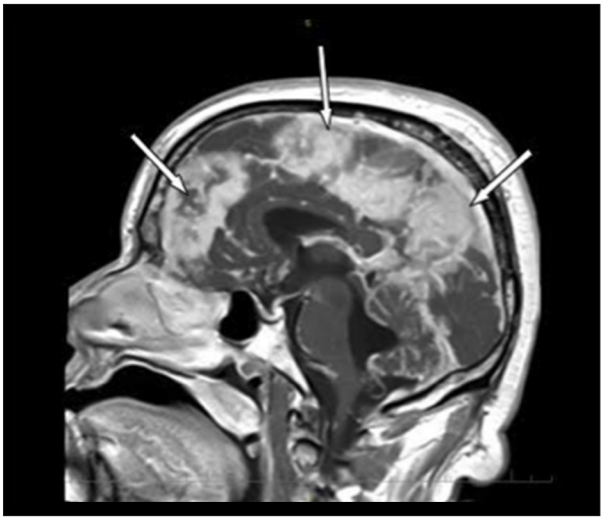


Figure 1. CT head showing cerebral edema and enlarged 4th ventricle.

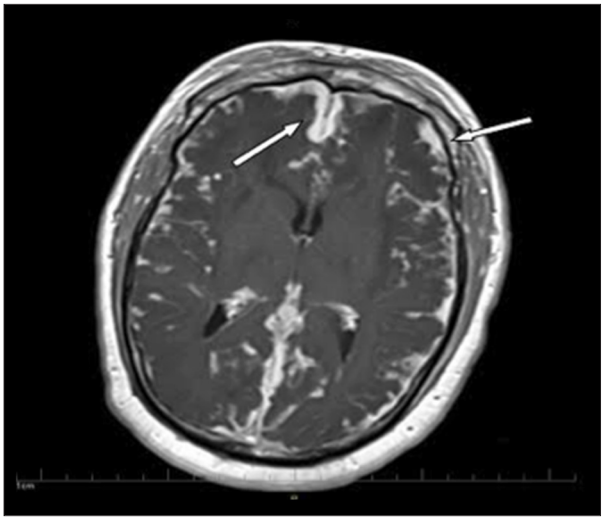
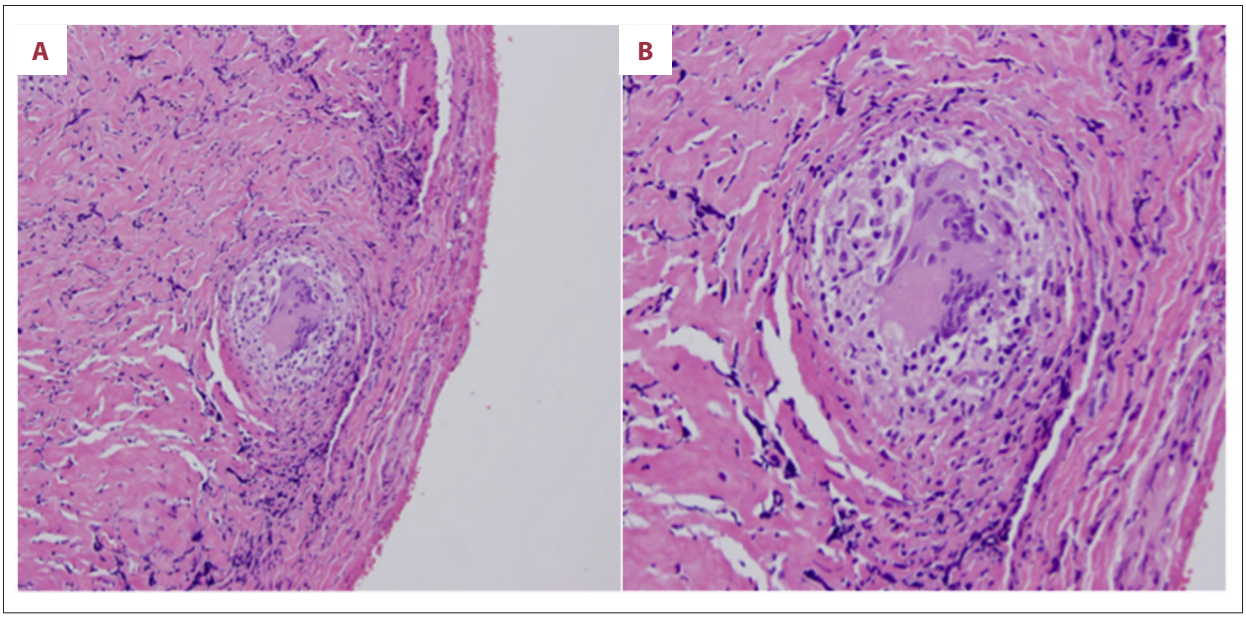


Figure 2. MRI showing leptomeningeal enhancement, cortical swelling, and vasogenic edema.



Figures 3. (**A, B**) Mediastinal biopsy revealing non-caseating granulomas and variable sclerosis, confirming long-standing sarcoidosis.

suggested by imaging involves multiple regions of the cortex and possibly the brainstem.

The hypothalamus receives its vascular supply from anterior and posterior regions of the cerebrum. Anteriorly, it is mostly supplied by branches of the anterior cerebral and anterior communicating arteries, while the tuberal and mammillary regions mostly receive blood supply from the posterior circulation, including the posterior cerebral, basilar, and posterior communicating arteries [9,10]. To the best of our knowledge, our patient is the first reported case in the literature with neurosarcoidosis presenting as hypothermia.

Conclusions

Neurosarcoidosis is a rare variant of sarcoidosis, which has varying presentations, including headaches, seizures, ataxia, visual disturbances, loss of sensation, and other neurologic abnormalities. Although most cases are treatable, neurosarcoidosis can have mortality as high as 10%. Imaging studies and biopsy remain integral to confirming the diagnosis. Corticosteroids, immunomodulators, and close follow-up are critical to managing the symptoms and reducing long-term sequelae of the condition. Our patient's symptoms improved with intravenous steroids, but long-term response to treatment was not determined because the patient was lost to follow-up.

To the best of our knowledge, no reported case in the literature has described hypothermia as the presenting symptom of a patient with neurosarcoidosis. We aim to use this case as a teaching moment and for further discussion on clinical presentations and management of such patients.

Acknowledgements

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Conflict of Interest

None declared. None of the aforementioned authors have any financial interest in this report.

References:

1. Voorter CE, Drent M, Hoitsma E et al: Association of HLA DQB1 0602 in sarcoidosis patients with small fiber neuropathy. Sarcoidosis Vasc Diffuse Lung Dis, 2005; 22(2): 129–32
2. Desestret V, Didelot A, Meyronet D et al: Neurosarcoidosis with diencephalitis and anti-Ma2 antibodies. Neurology, 2010; 74(9): 772–74
3. Stern BJ, Krumholz A, Johns C et al: Sarcoidosis and its neurological manifestations. Arch Neurol, 1985; 42(9): 909–17
4. Motta M, Alongi F, Bolognesi A et al: Remission of refractory neurosarcoidosis treated with brain radiotherapy: A case report and a literature review. Neurologist, 2008; 14(2): 120–24
5. Joseph FG, Scolding NJ: Neurosarcoidosis: A study of 30 new cases. J Neurol Neurosurg Psychiatry, 2009; 80(3): 297–304
6. Allen RK, Sellars RE, Sandstrom PA: A prospective study of 32 patients with neurosarcoidosis. Sarcoidosis Vasc Diffuse Lung Dis, 2003; 20(2): 118–25
7. Kobayashi D, Rouster-Stevens K, Harper A: La crosse virus encephalitis preceding neurosarcoidosis diagnosed by brain biopsy. Pediatrics, 2011; 127(4): e1091–96
8. Petropoulos IK, Zuber JP, Guex-Crosier Y: Heerfordt syndrome with unilateral facial nerve palsy: A rare presentation of sarcoidosis. Klin Monbl Augenheilkd, 2008; 225(5): 453–56
9. Rhoton AL: The supratentorial arteries. Neurosurgery, 2002; 51(4 Suppl.): S53–120
10. de Souza DG, Prevedello DM, Ditzel Filho L.F. et al: Superior hypophyseal artery anatomy: A ventral perspective. J Neurol Surg B, 2014; 75: A116