Clinical Challenges in the Diagnosis and Treatment in Neurosarcoidosis and Neurosyphilis

Mackenzie Kramer MD[1], Melissa Margolis[2], Neelima Manda MD[1], Rehan Saeed MD[1], Stephanie Tzarnas MD[1], Nathaniel Rosal DO[1], Daniel Puga MD[1], Rebecca Fallis MD[3]

1. Department of Internal Medicine, Abington Jefferson Health, Abington, PA
2. Drexel University College of Medicine, Philadelphia, PA
3. Department of Infectious Disease, Abington Jefferson Health, Abington, PA

Introduction

Syphilis and sarcoidosis, both known as the “great mimickers,” cause a wide range of symptoms making clinical differentiation between the two difficult.

We present a case in which both neurosarcoidosis and neurosyphilis were considered in the differential diagnosis, posing a unique challenge in decision making.

Case

A 54-year-old African American male with a medical history of anterior uveitis presented with new onset diplopia, 15lb weight loss over one-month, lower extremity paresthesia, and chronic sinusitis. Physical examination revealed new lateral gaze palsy and a left facial droop. Given the concern for stroke, a CT angiogram of the head and neck was obtained which showed no evidence of stroke, but mediastinal/cervical lymphadenopathy and severe mastoiditis. Subsequent MRI brain showed radiologic evidence correlating with cranial nerve VI palsy.

Case

Follow up CT chest/abdomen/pelvis showed multiple enlarged mediastinal lymph nodes and two lung nodules.

HIV test was negative, syphilis screen was positive with reflex RPR 1:8, and ACE level upper limit of normal at 67 U/L. Of note, the patient was previously treated for syphilis with anterior uveitis in 2018 with the last documented RPR of 1:16 prior to treatment.

We proceeded with a right inguinal lymph node fine needle aspiration, which revealed noncaseating granulomas. An axillary lymph node excisional biopsy showed noncaseating granulomas with negative stains for fungus, mycobacterium, and spirochetes.

As the patient refused to undergo a lumbar puncture for definitive diagnosis, he was empirically treated for both neurosarcoidosis with high dose steroids,as well as neurosyphilis with two weeks of parenteral penicillin G followed by a final 2.4 million unit injection at week 3. His wife was also counseled that she should be tested and treated for syphilis.

Discussion

Both neurosarcoidosis and neurosyphilis may be difficult to differentiate due to the overlapping and varied presentations. Additionally, there is no definitive test to diagnose neurosarcoidosis.

Our patient exhibited cranial nerve VI palsy, diffuse lymphadenopathy, noncaseating granulomas, and positive RPR without fourfold decrease since his prior syphilis treatment. Cranial nerve palsies may occur in both neurosarcoidosis and neurosyphilis.

Noncaseating granulomas are characteristic of sarcoidosis but can occur in rare cases of tertiary syphilis. A lumbar puncture would have proven useful to help differentiate between the two processes, although lumbar puncture may be a significant barrier to patients.

In neurosarcoidosis, we expect to see elevated CSF total protein and pleocytosis with mononuclear cell predominance. In neurosyphilis, we expect to see elevated CSF protein and positive CSF-VDRL, though the latter is poorly sensitive and only positive in about 50% of cases.

Conclusion

This case demonstrates the difficulty in distinguishing two vastly different diseases, the importance of practicing patient-centered care through education, and counseling regarding the diagnosis and treatment of the disease entity encountered.

References

1) Terushkin V, Stern BJ, Judson MA, Hagiwara M, Pramanik B, Sanchez M, Prystowsky S. Neurosarcoidosis: presentations and management. Neurologist. 2010 Jan;16(1):2-15. doi: 10.1097/NRL.0b013e3181c92a72. Erratum in: Neurologist. 2010 Mar;16(2):140. PMID: 20065791.

2) Pawate S, Moses H, Sriram S. Presentations and outcomes of neurosarcoidosis: a study of 54 cases. QJM. 2009 Jul;102(7):449-60. doi: 10.1093/qjmed/hcp042. Epub 2009 Apr 20. PMID: 19383611

3) Mehta, A. C., & Ali, S. R. (2017). Mnemonic for the differential diagnosis of non-caseating granulomas. Sarcoidosis, vasculitis, and diffuse lung diseases : official journal of WASOG, 34(2), 200–207. Edmonds LC, Stubbs SE, Ryu JH. Syphilis: a disease to exclude in diagnosing sarcoidosis. Mayo Clin Proc. 1992 Jan;67(1):37-41. doi: 10.1016/s0025-6196(12)60277-8. PMID: 1732690

Abington Jefferson Health®