Neurosarcoidosis Diagnosed After Three Decades of Evolution
A Case of Perseverance and Insight

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Abstract
A lymphoma survivor presented for reassessment of his long-standing neurological symptoms more than three decades following a diagnosis of systemic T-cell lymphoma. Despite our desire to establish the diagnosis promptly, time is often necessary to reach conclusions based on the patient’s clinical evolution. We recount the chronology of clinical and radiological findings that, until recently, were largely inexplicable. We then report on the challenging diagnosis of neurosarcoiodosis and suggest a known, yet rare, continuity linking it with lymphoma. Finally, we emphasize on the clinical evolution as a determining factor in the quest for a complex diagnosis. Insight, judgment, and perseverance are featured as key virtues of a resilient Neurologist.

Key Words: sarcoidosis, neurosarcoiodosis; t-cell lymphoma; anti-tr paraneoplastic syndrome; granulomatous disease

Case Report
In 1990, a 31-year-old male known for thoracic T-cell lymphoma presented for new-onset ataxia. He underwent chemotherapy and achieved remission within a year. Six years after oncological presentation, this gentleman developed pancerebellar symptomatology. A brain MRI revealed numerous bi-hemispheric white matter lesions consistent with lymphoma recurrence to the CNS, surprisingly sparing the posterior fossa, thus a paraneoplastic cerebellar syndrome (anti-Tr1) was suspected. The patient was treated with prednisone according to the clinical presentation, in an era of laboratory limitations for paraneoplastic antibodies. Following the curative treatment of the underlying neoplasia and steroid exposure, his symptoms resolved. Unexpectedly, several attempts to taper the oral prednisone failed over the subsequent three decades, leading to deleterious complications from prolonged steroid exposure.

Based on this patient’s medical history, failure of steroid tapering was unjustified, and we were motivated to revisit every detail. We reattempted an extremely slow tapering of oral prednisone, decreasing the initial dose of 60 mg OD by 2.5 mg every two weeks. The purpose was to document the nature of the previous failures, considering the possibility of asthenia due to adrenal insufficiency versus focal neurologic symptoms due to distinct CNS lesions. At half of the prednisone daily dose, the patient developed fatigue, dysphagia, and diffuse pyramidal weakness. Concomitant respiratory distress associated with pleural thickening, lobar consolidation and PET-FDG uptake further complicated the picture. Clinical examination was hindered by severe steroid-induced proximal muscular atrophy confining our patient to a wheelchair. Orthopedic procedures were required, resulting in a significant deconditioning affecting his global function.

A brain MRI showed numerous subacute enhancing white matter lesions scattered throughout the white matter of bilateral cerebral hemispheres (Figure 1).
that the patient’s neurological deterioration and peculiar sensitivity to prednisone was attributed to neurosarcoidosis.

The co-occurrence of lymphoma and sarcoidosis is a rare and intriguing phenomenon described as the sarcoidosis-lymphoma syndrome. It is theorized that granulomatous lesions act as a microenvironment of aberrant immune function. Elevated CD4+ activity increases the risk of developing dysplastic changes, while diminished CD8+ presence impairs immunoregulatory capacity. Unrestricted T-cell activation is promoted, ultimately leading to lymphoma transformation. We have presented the work-up of a steroid-dependent neurologically impaired lymphoma survivor leading to the diagnosis of neurosarcoidosis. This case exemplifies the fundamental and variable relationships between lymphoma and its numerous comorbidities. Furthermore, we demonstrate that the manifestations of cancer may be represented in figurative balance oscillating between immunosuppression and immunoreactivity, looking for its equilibrium. Finally, this case emphasizes the basic tenet of neurological investigation: do not dismiss clinical evolution or leave any stone unturned in challenging or inexplicable circumstances; the answer is often in front of us, buried under a pile of confounding factors.

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Authors Contributions
Dr. Jason Jia was responsible for conceptualization, redaction, editing, and revision.
Dr. Catherine Maurice was responsible for conceptualization, redaction, editing, figures, supervision and revision.

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