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Adra et al[1] report on the shrimp sign, a highly sensitive and specific neuroimaging finding seen in patients with cerebellar progressive multifocal leukoencephalopathy (PML). We read their article with great interest because PML often poses a diagnostic challenge. This challenge is compounded for HIV-negative patients in whom an etiology of immunocompromise is not readily apparent. In particular, PML may occur in patients with systemic rheumatologic disorders, including those who are not on immunosuppressive or immunomodulatory medications.[2] In these patients, the shrimp sign described by Adra et al has the potential to avert misdiagnosis and obviate the need for invasive testing, such as brain biopsy.

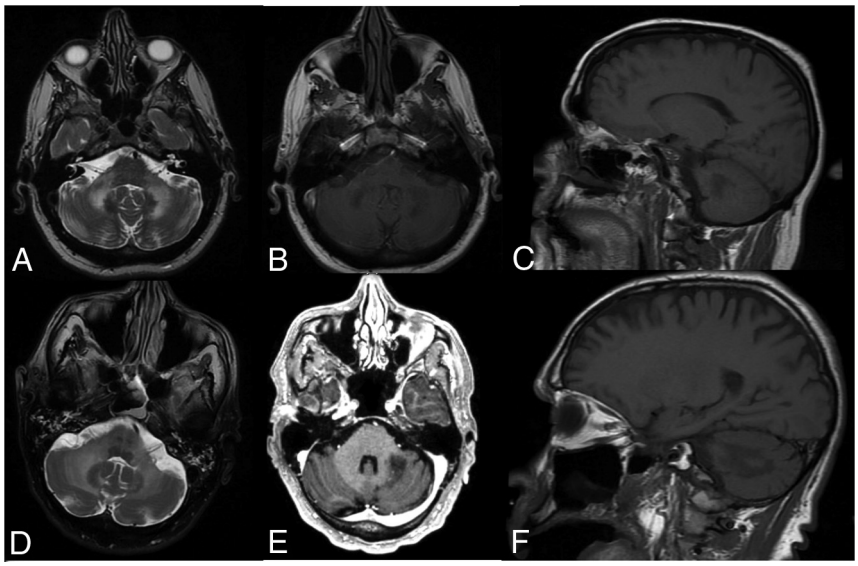

FIGURE. In both cases, brain MR imaging revealed well-defined, T2-hyperintense (patient 1 [*A*], patient 2 [*D*]) and T1-hypointense lesions (patient 1 [*B* and *C*], patient 2 [*E* and *F*]) of the cerebellar white matter abutting and sharply demarcating and outlining the dentate nucleus.

The association between PML and sarcoidosis was first reported in 1955, when PML was found at postmortem examination in 6 patients with sarcoidosis whose neurologic symptoms that had been mistakenly attributed to neurosarcoidosis. It is hypothesized that general T-lymphocyte dysregulation and anergy, as well as the redistribution of CD4+ T-lymphocytes to sites of granuloma formation, may reduce immune surveillance and allow the reactivation of the JC virus in the brain. Misdiagnosis remains common in patients with sarcoidosis who develop PML, which is particularly concerning given the risk of worsened outcomes in patients with PML who are inappropriately treated with immunosuppressive or immunomodulatory therapies for presumed neurosarcoidosis.[3] Patients with sarcoidosis and other systemic rheumatologic diseases may also be at higher risk for false-negative CSF JC virus testing results than more severely immunocompromised patients, requiring brain biopsy to facilitate diagnosis.[4]

Here, we report on 2 patients with sarcoidosis who presented with new neurologic symptoms and were successfully diagnosed with PML using the MR imaging shrimp sign of Adra et al.[1] Patient 1 is a 49-year-old man with a history of pulmonary sarcoidosis, not on immunosuppression, who presented with imbalance, dysphagia, dysarthria, and incoordination of his right hand. Patient 2 was a 62-year-old man with a history of pulmonary, splenic, and cutaneous sarcoidosis, not on immunosuppression, who presented with incoordination of his left hand, imbalance, and scanning speech. MR imaging of the brain for both patients demonstrated well-defined, T2-hyperintense, T1-hypointense lesions of the cerebellar white matter abutting and sharply demarcating and outlining the dentate nucleus on the axial view (Figure), meeting the inclusion criteria for the shrimp sign of Adra et al. Both patients were misdiagnosed with neurosarcoidosis at an outside hospital, and patient 1 was treated with high-dose steroids. Following review of neuroimaging by physicians familiar with the

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shrimp sign, the diagnosis of PML was confirmed by positive CSF JC virus PCR in both cases.

In light of these cases, we encourage physicians involved in the management of patients with sarcoidosis to familiarize themselves with the features of the shrimp sign of Adra et al,[1] which may help avert misdiagnosis and facilitate appropriate care of PML in this unique population.

Disclosure forms provided by the authors are available with the full text and PDF of this article at www.ajnr.org.

P. Anand and K.H. Vincent Lau shared first authorship.

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