Imaging features consistent with Neurosarcoidosis

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Background: Neuroimaging has emerged as an important tool in the diagnosis of neurosarcoidosis and MRI is the preferred imaging technique with high sensitivity but low specificity.

Objective: To describe diagnostic challenges that can be encountered in the diagnosis of neurosarcoidosis

Design: Case report
Setting: The University of Rochester Medical Center

Patient:
A 53-year-old otherwise healthy man presented with seven years of slowly progressive left greater than right leg weakness and numbness, causing him to use bilateral assistive devices. He also experienced urinary urgency.
Neurologic exam was notable for bilateral leg spasticity with distal greater than proximal leg weakness, 3+ patellar reflexes with absent Achilles reflexes, vibratory sense of 5 seconds at bilateral toes and mild cognitive impairment (Montreal Cognitive Assessment score 25/30). MRI lumbar spine showed a T2 intramedullary hyperintensity spanning T10 to the conus medullaris that heterogeneously enhanced. There were scattered enhancing nodularities of the cauda equina nerve roots. These findings were unchanged compared with an MRI from 7 years earlier, before the patient was lost to follow-up. MRI brain showed extensive T2 hyperintensities in the bilateral cerebral white matter and brainstem with multiple areas of punctate and linear enhancement.
CSF showed 22 nucleated cells/mm3 (lymphocytic predominance), protein 291 mg/dL, and glucose 58 mg/dL. CSF angiotension-converting enzyme was elevated at 4.4 U/L. Negative CSF testing included cytology, oligoclonal bands, AQP-4 IgG and viral/fungal cultures. Serum tests were unremarkable including negative AQP-4 IgG, HIV, RPR, antinuclear antibodies, double stranded DNA, anti-SSB/SSA, and HTLV 1/2. Chest CT showed scattered sub-centimeter mediastinal lymph nodes and pulmonary nodules. A gallium scan showed abnormal increased uptake in bilateral hilar lymph nodes and left cerebellopontine region.

Intervention: Abnormal neuro-imaging, elevated CSF/serum ACE levels and evidence of increase metabolic activity on gallium scan or FDG-PET are all supportive for a diagnosis of neurosarcoidosis in the absence of tissue biopsy. Once diagnosis is made, immunosuppressive therapy can be started.

Main Outcome Measures: Clinical improvement

Results: The linear and punctate enhancement, the elevated ACE and gallium scan were most consistent with neurosarcoidosis.

Conclusions: The case described above shows the diagnostic challenges that can be encountered in the diagnosis of neurosarcoidosis. Punctate or linear enhancement on neuro-MRI in the right clinical setting should raise the suspicion for neurosarcoidosis and prompt appropriate workup.