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Clinical and Magnetic Resonance Imaging Manifestations of Neurosarcoidosis

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OBJECTIVES To describe clinical and neuroimaging manifestations of neurosarcoidosis in a cohort of 21 patients.

PATIENTS AND METHODS We reviewed records of 21 patients with sarcoidosis and central nervous system (CNS) manifestations referred to Cooper University Hospital, with emphasis on neuroimaging findings and associated clinical and laboratory evidence of sarcoidosis. Nineteen patients were categorized as having "definite," "probable," or "possible" neurosarcoidosis, while 1 had associated CNS vasculitis and another had Hodgkin's lymphoma with cauda equina syndrome.

RESULTS The most common manifestations included myelopathy, cranial neuropathies, and encephalopathy. In 6 patients, CNS biopsy showed sterile, noncaseating granuloma (NCG), while in the remainder, the diagnosis was established through a combination of clinical, radiographic, and laboratory findings. Notably, 10 patients developed acute neurological emergencies, including seizures, spinal cord compression, and increased intracranial pressure. Findings on magnetic resonance imaging (MRI) included a variety of manifestations, including isolated mass lesion, diffuse intraparenchymal inflammatory lesions in the brain and spinal cord, leptomeningeal enhancement, hydrocephalus, and intracranial hemorrhage.

CONCLUSIONS Sarcoidosis is associated with diverse neurological manifestations and neuroimaging findings. The diagnosis of isolated CNS sarcoidosis requires a biopsy to document the presence of sterile NCG and to exclude neoplasms and other granulomatous diseases. When a biopsy of the CNS is not possible, a diagnosis of neurosarcoidosis can reasonably be supported in many patients by MRI findings and exclusion of other disorders.

RELEVANCE Optimum management of patients with neurosarcoidosis relies on the ability of clinicians to recognize the broad spectrum of clinical and neuroimaging manifestations of the disorder.

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KEYWORDS neurosarcoidosis, sarcoidosis, seizures, cranial neuropathy, encephalopathy, spinal cord compression

Sarcoidosis is a multisystem, granulomatous disease that affects the nervous system in 5-10% of patients (1-4). The diagnosis of neurosarcoidosis may be challenging, as the disease mimics other infectious, demyelinating, granulomatous, neoplastic, and connective tissue disorders affecting the nervous system. The diagnosis is particularly difficult when neurological complications precede other systemic manifestations of sarcoidosis affecting lungs, lymph nodes, skin, or eyes (5). No uniform, validated classification criteria for neurosarcoidosis were available until recently. The establishment of recent classification criteria (6) for neurosarcoidosis and the increasing availability of magnetic resonance imaging (MRI) may facilitate the identification of a wide spectrum of clinical and MRI manifestations of neurosarcoidosis.

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Table 1 Classification of Patients with Neurosarcoidosis and Summary of Findings

Patient No., Age/Race/Sex, Classification	Clinical Presentation	Imaging Findings	CSF	Biopsy	Other Manifestations of Sarcoidosis	Treatment and Outcome
1, 53/B/F Definite	Headaches, Encephalopathy	MRI: Enhancing mass left frontal lobe with subdural extension	ND	Brain and iliac crest lesions + NCG, uveitis	Gallium scan: abnormal lymph node uptake ACE = 136	Resection and CS; remains stable on low dose oral CS, methotrexate and hydroxychloroquine
2, 47/B/M Definite	Headaches, increased intracranial pressure	MRI: Enhancing mass 4th ventricle with hydrocephalus	Normal	4th ventricle, liver and spleen +NCG	Hilar LAD ACE = 48	Resolved with VP shunt; CS for pulmonary symptoms; died from gastric cancer 3 years
3, 45/B/F Definite	Trigeminal neuralgia, hearing loss, vertigo, encephalopathy, panhypopituitarism	MRI: suprasellar pachymeningitis extending into basilar cisterns and hypothalamus	Pleocytosis, protein = 308	Suprasellar biopsy +NCG	CXR = normal ACE = 14	Improved with CS and treatment of SIADH; rhizotomy for trigeminal neuralgia
4, 32/B/M Definite	Visual loss	MRI: Enhancement and swelling of optic nerve and chiasm, pachymeningitis	Normal	Optic nerve +NCG	Hilar LAD	Improvement with IV CS; maintained on oral CS developed avascular necrosis and osteoporosis
5, 41/W/F Definite†	Cauda equina syndrome	MRI: enhancing vertebral lesions with extension into spinal canal	ND	Vertebrae +NCG	Hilar LAD ACE = 111	Improvement following surgical decompression; Hodgkins lymphoma in remission following chemotherapy
6, 40/B/M Definite	Neck pain, gait disturbance, sensory loss	MRI: enhancing lesion in cervical spinal canal with abnormal signal in adjacent spinal cord	ND	Arachnoid mass +NCG	Hilar and systemic LAD	Minimal residual spasticity after surgical decompression
7, 25/H/F Probable	Paraparesis and visual loss	MRI: enhancing lesions in thoracic spinal cord and suprasellar meninges	ND	Lymph node +NCG	Hilar LAD ACE = 205*	Near-complete resolution after 2 days IV CS, oral CS taper
8, 65/W/F Probable	Chronic spastic paraparesis	MRI: multiple hyperintense lesions in brain and cervical spinal cord	Normal on two occasions	Skin +NCG	CXR: normal ACE = 49	Oral CS; increased weakness from steroid myopathy