Supplementary Online Content

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This supplementary material has been provided by the authors to give readers additional information about their work.

eTable 1. Clinical Manifestations and Frequency of Neurosarcoidosis

Clinical Manifestation	Approximate Frequency, % (95% CI)[e23]	Comment
Cranial neuropathy	55 (52-58)	
2	21 (18-24)	
7	24 (21-27)	
>1	28	
Meningeal disease	16 (13-19)	"meningitis"
Acute, aseptic meningitis	NA	
Chronic meningitis		
Mass lesion		
Hydrocephalus	9	
Communicating	NA	
Noncommunicating		
CNS disease	NA	
Endocrinopathy	9	
Mass lesion	NA	
Encephalopathy/micro-vasculopathy	NA	
Seizure	14	

Stroke		• Small or large artery in situ thrombosis
		• Cardiogenic emboli
		  o Cardiomyopathy
		  o Arrhythmia
		• Large artery compression from a granulomatous mass
		• Artery-to-artery emboli
		• Sinovenous thrombosis
		• Intracerebral hemorrhage

Vegetative dysfunction	NA	
Spinal cord	18 (15-21)	
Nerve roots, including cauda equina syndrome	NA	
Neuropathy	17 (14-21)	
Mononeuropathy	NA	
Axonal or demyelinating		
Sensory, motor, sensorimotor		
Myopathy	15 (9-11)	
Polymyositis	NA	
Nodule		
Paraneurosarcoidosis	NA	

Small fiber neuropathy	30[e12]	• Sensory neuropathy
		  o Impaired pin and temperature sensibility
		  o Length and non–length dependent
		  o Often painful
		• Autonomic neuropathy

Brain fog	NA	
Fatigue	NA	

Abbreviations: CNS, central nervous system; NA, not applicable.

eTable 2. Select Clinical Manifestations of Neurosarcoidosis and Associated Comments and Diagnostic Considerations

Clinical Manifestation	Comments	Diagnostic Considerations
Meningeal disease	• Subacute lymphocytic meningitis • Chronic lymphocytic meningitis • Dural involvement including pachymeningitis • Dural mass mimicking a meningioma	
Cranial nerves	• Any cranial nerve can be involved. • Unilateral or bilateral lower facial nerve palsy is the most common cranial nerve presentation. [e1] • Relatively frequent cranial neuropathies are optic neuropathy and eighth (acoustic/vestibular) nerve dysfunction, which can be unilateral or bilateral. • Optic nerve involvement typically presents as a subacute or chronic optic neuritis. [e2] • Vision loss can be rarely due to extrinsic compression of the optic nerve from granulomatous inflammation. • Isolated hearing loss or vertigo due to selective vestibulocochlear nerve involvement can occur; bilateral sensorineural hearing loss should prompt an evaluation for NS. [e3]	• Optic nerve involvement is usually manifest as swelling, T2 hyperintensity, and contrast enhancement of the optic nerve, optic nerve sheath, or chiasm. [e2]
Brain	• Small subcortical or periventricular white matter lesions may be asymptomatic and represent inflammatory changes or small ischemic lesions resulting from arteriolar or venous inflammation-associated thrombosis. • NS has a predilection for the hypophysis-hypothalamus, resulting in a range of disturbances including hypothyroidism, hypogonadism, and the syndrome of inappropriate antidiuretic hormone (SIADH). • Communicating and non-communicating hydrocephalus can develop.	• Leptomeningeal enhancement, especially along the skull base, is the most common area involved. • Dural thickening and enhancement can be localized or diffuse, and may be hypointense on T2-weighted sequences. • Intraparenchymal granulomas can be seen as subcortical white matter lesions mimicking multiple sclerosis or micro-ischemic lesions, or can involve the deep grey matter. • Large solitary aggregations of granulomas can masquerade as neoplasms. • Diffusion weighted imaging of parenchymal disease is not bright, except rarely when an infarct results from vascular involvement.

		• Stroke can be due to in situ thrombosi, cardiogenic emboli, compression of a large vessel by a granulomatous mass, sinovenous thrombosis, and intracerebral hemorrhage.

Spinal cord	• Most patients have subpial intramedullary lesions and meningeal involvement is present in up to half the patients. [e4] • Patients may have gradually worsening myelopathic symptoms developing over months. [e5] • Myelitis may span more than 3 vertebral segments (predilection cervicothorac); NS is a diagnostic consideration of a longitudinally extensive myelitis, including neuromyelitis optica spectrum disorder. [e6]	• Spinal cord involvement can be intramedullary and indistinguishable from other causes of transverse myelitis. • Lesions are typically longitudinally-extensive. [e6] • The characteristic location of increased T2 signal in sarcoidosis is in the dorsal cord, and contrast enhancement is typically subpial, dorsal, and irregular or nodular in appearance. • A characteristic finding on axial views is the “trident” sign of contrast enhancement extending from the dorsal subpial surface circumferentially bilaterally with a central spoke extending into the midline of the cord. [e7] • Meningeal involvement, with both dural thickening and enhancement, suggests NS.
Neuropathy	• The pattern of large fiber nerve involvement varies widely, but most commonly presents as either an axonal distal sensorimotor polyneuropathy with stocking-glove sensory symptoms manifesting as pain and paresthesias or an asymmetric polyradiculoneuropathy in which proximal and distal nerve segments are equally affected in a non-length dependent distribution. [e8,9] • Other neuropathy manifestations include mononeuritis multiplex, pure sensory or pure motor neuropathy, and isolated plexus, root or peripheral nerve involvement (e.g. focal mononeuropathy). Less frequent are demyelinating neuropathies, which include chronic inflammatory demyelinating polyneuropathy, a Guillain-Barre syndrome (GBS)-like presentation, and multifocal motor neuropathy with conduction block.	• Electromyography (EMG), and nerve conduction study (NCS) findings are nonspecific and often indistinguishable from primary immune-mediated neuromuscular disorders. • EMG and NCS usually demonstrate changes consistent with axon loss; primary demyelinating abnormalities are occasionally found. • While imaging studies are not routinely part of the neuropathy evaluation, MRI may demonstrate gadolinium enhancement or enlargement of the affected nerve roots or plexus. • CSF findings are nonspecific and may be normal, but elevated protein is most commonly seen followed by pleocytosis. • Histological confirmation of the affected tissue or evidence of systemic disease is required for a definite or probable diagnosis of sarcoidosis neuropathy, respectively. • Biopsy of both muscle and nerve tissue increases the diagnostic yield as granulomas between muscle fibers tend to be larger and more developed than those found within the epineurium or endoneurium. [e8,10,11]

Para-neurosarcoidosis	• Small fiber neuropathy
	  o Sensory
	  o Autonomic
	• “Brain fog”
	• Fatigue
		• Small fiber neuropathy (SFN)
		  o The diagnosis of sarcoidosis-associated SFN should only be made in patients in whom the diagnosis of extra-neural sarcoidosis or NS is certain.
		  o Patients usually present with prominent somatic nerve dysfunction (e.g., pain, numbness, dysesthesias, and allodynia) [e12] that is migratory and intermittent but can become constant and slowly progressive. Distribution is often in a non-length dependent pattern. [e13]
		  o Dysautonomia manifesting as orthostatic hypotension, gastrointestinal disturbances, sweating changes can occur. [e14]
		  o Skin biopsy with evaluation of intra-epidermal nerve fiber density (IENFD) can confirm the presence of a SFN, with reduced sweat gland density and innervation supporting an autonomic neuropathy.
		  o Analysis of sweat production on quantitative sudomotor axon reflex testing (QSART) and corneal nerve fiber density on corneal confocal microscopy can also be helpful in confirming the clinical impression.
		  o For patients with prominent symptoms of dysautonomia, tilt table testing, heart rate variability measurements, and thermoregulatory sweat testing can be considered.
		• When evaluating a patient for “brain fog” consider infection, disordered sleep, hypothalamic pituitary dysfunction, depression, etc.
		• When evaluating a patient for fatigue consider pulmonary or cardiac disorders, depression, hypothyroidism, testosterone deficiency, corticosteroid myopathy, sleep disorders, painful neuropathy, etc.

eTable 3. Differential Diagnosis of Sarcoidosis and Neurosarcoidosis

Potential multi-system disorders to consider based on clinical context	**Comments**
Systemic and CNS Infections	
Lyme disease [neuroborreliosis] [e25]	Obtain Lyme serologic studies in blood and CSF* [26]
Neurosyphilis [e27]	Obtain syphilis serologic studies in blood and CSF*
CNS Fungal Infection including coccidiomycosis, histoplasmosis, cryptococcus, spirotrichosis, blastomycosis	Appropriate histologic stains and culture.* If chronic meningitis, CSF cryptococcal antigen should be performed. Consider fungal serology (Histoplasmosis, Blastomycosis, and Coccidiomycosis) depending on epidemiology, from serum and CSF. CSF Sporotrichosis serology in selected cases should be considered.
Tuberculosis [e28]	PPD*, Quantiferon gold assay*; Appropriate histologic stains and culture including from the CSF
Bacterial meningitis, especially subacute and chronic phenotypes	Culture*, PCR, serologic testing
Whipple's disease [e29]	PCR assay on CSF; small bowel or brain biopsy
Atypical mycobacterium (very rare in the absence of prior neurosurgery or hardware)	Appropriate histologic stains and culture*
Progressive multifocal leukoencephalopathy	CSF PCR assay for JC virus, brain biopsy if high suspicion and CSF negative per guidelines. [e30] Note that sarcoidosis,

	even in the absence of therapeutic immunosuppression, can be sufficient as an illness to predispose to PML. [e31]
HIV	HIV serology*. CDC recommendations for diagnosis. [e32]
Other CNS viral infections	CMV PCR (most commonly ventriculitis or radiculomyelitis in a severely immunosuppressed patient); CSF VZV PCR and VZV antibodies; other viral PCRs and serologies as appropriate based on clinical context and local epidemiology
Parasitic infection (in the appropriate neuroimaging, CSF exam or epidemiological context)	Toxoplasma antibodies in serum and CSF (a negative serum test result usually rules out the infection) and biopsy of the lesion; cystercercosis antibodies; schistosomiasis (especially for spinal cord lesion)
Malignancy	
Lymphoma (Primary CNS or Systemic with neurological involvement)	SPEP/IFE, LDH, CSF flow cytometry and cytology*, body imaging (CT, FDG PET), histology
Meningeal carcinomatosis	MRI, CSF cytology, evidence of metastatic carcinoma on examination and body imaging.
CNS tumors (glioma, germ cell tumor, craniopharyngioma)	MRI, biopsy
Histiocytosis	Systemic involvement, Body imaging, Biopsy
Other Neuro-inflammatory Disorders	
Multiple sclerosis	MS diagnostic criteria [e33] based on clinical course and MRI findings; CSF IgG index and oligoclonal bands can be helpful para-clinical markers.

Neuromyelitis optica	NMO diagnostic criteria [e34], AQP4 IgG*; MRI findings (the pattern of enhancement in myelitis from NMO is typically intraparenchymal versus subpial involvement in the myelitis of neurosarcoidosis). [e5]
Tumefactive / Fulminant Demyelinating Process	Imaging features, biopsy
Acute Disseminated Encephalomyelitis	Initial diagnosis based on clinical features and routine testing. [e35]
Primary CNS Vasculitis [e36]	Clinical course, MRI findings, Angiography (including digital subtraction angiograph), Biopsy
CNS or PNS involvement from systemic vasculitis	Anti-neutrophil cytoplasmic and proteinase 3 antibodies (particularly for PNS manifestations), Cryoglobulins, Rheumatoid Factor, Complement, ANA, SSA, Hepatitis C antibody, hepatitis B serologies, renal markers (Creatinine, urinalysis). Evidence for systemic involvement, which may include involvement of skin, kidneys, lungs, sinuses, and other sites of injury. Granulomatosis with polyangiitis can manifest with pachymeningitis. [e37]
Lymphocytic hypophysitis	Isolated pituitary involvement; biopsy with demonstration of a mixed cellular inflammatory infiltrate and fibrosis. Granulomatous hypophysitis may be considered a manifestation of sarcoidosis versus an organ-specific granulomatous process depending on clinical context.
Chronic lymphocytic inflammation with pontine	Pontine perivascular enhancement on MRI responsive to corticosteroids; biopsy

perivascular enhancement responsive to steroids (CLIPPERS) [e38]	
Autoimmune astrocytopathy [e39]	Inflammation of the meninges, brain, and spinal cord with peri-vascular enhancement and longitudinally extensive spinal cord lesions on MRI. Glial fibrillary acidic protein (GFAP) IgG positive.
Myelin oligodendrocyte glycoprotein (MOG) antibody-associated disease [e40]	MOG IgG antibody positive. MRI lesions tend to be bilateral, large and “fluffy”, and involve subcortical and deep structures as well as the brainstem.
Neurologic Involvement from a Systemic Disorder	
Systemic lupus erythematosus	SLE diagnostic criteria [e41] ANA, double-stranded DNA and associated assays*
IgG4-related hypertrophic pachymeningitis [e42 43]	Serum IgG subsets for IgG4 elevation may be suggestive but is not specific. IgG4-related disorders may include pachymeningitis as part of systemic disease involvement. Tissue biopsy is definitive.
Sjogren’s syndrome	(For evaluation of peripheral nervous system involvement) SSA and SSB serologies, sicca symptoms, lip biopsy, sialography or salivary flow. [e44] Caution is advised in correlating CNS findings with Sjogren’s syndrome , particularly in the absence of histology. Note that sicca symptoms in isolation are not specific for Sjogren’s syndrome and can also be seen in sarcoidosis.

Behcet's disease	Behcet's diagnostic criteria [e45]: Recurrent oral aphthous ulcers, plus any two of the following: positive Pathergy test, recurrent genital ulcers, eye lesions, characteristic skin lesions
Vogt-Koyanagi-Harada disease	Can cause recurrent meningitis in the context of severe uveitis, usually with cystoid macular edema as a prominent feature; not usually a chronic meningitis. Slit-lamp exam, fluorescein angiography, examination of the CSF (pleocytosis, melanin-laden macrophages), magnetic resonance imaging (MRI), and electrophysiologic testing [e46]
Common variable immunodeficiency (CVID)	Hypogammaglobulinemia. CVID may be associated with granulomatous disease that resembles sarcoidosis. [e47] CNS and PNS manifestations can occur due to infection or inflammation. [e48]
CSF Pressure Dynamics	
Syndrome of Intracranial Hypotension (SIH) / CSF leak	Diffuse pachymeningeal enhancement, often with other signs of brain sagging; low CSF opening pressure; low pressure headache syndrome, search for CSF leak
Vascular	
Dural arterio-venous fistula (brain or spinal cord) or other vascular malformations such as AVM	CT angiography or MRI angiography. Digital subtraction angiography for definitive evaluation.
Stroke	Ischemic and hemorrhagic stroke (which may relate to vascular inflammation, of large or small arteries or veins).

	Also consider cardiac sources of emboli from arrhythmias or congestive heart failure.
Other Disorders	
Cancer-associated granulomas	Occasionally granulomas can be manifest in regional lymph nodes or tissue proximate to a malignancy. A mass lesion not responding to immunosuppressive treatment may need to be biopsied, at times repeatedly

*: These tests should be considered in all patients being evaluated for the diagnosis of neurosarcoidosis unless there is a specific contraindication to obtaining the test (e.g. CNS mass effect precluding a LP).

eTable 4. Representative Previous Diagnostic Criteria for the Diagnosis of Neurosarcoidosis

Author/Year	Possible	Probable	Definite or Highly Probable	Comment
Judson/2014 [e15] [Footnote 1]	Seizures, negative MRI. Cognitive decline, negative MRI.	Isolated facial palsy, negative MRI. Clinical syndrome consistent with granulomatous inflammation of the meninges, brain, ventricular (CSF) system, cranial nerves, pituitary gland, spinal cord, cerebral vasculature, nerve roots but without characteristic MRI or CSF findings.	Clinical syndrome consistent with granulomatous inflammation of the meninges, brain, ventricular (CSF) system, cranial nerves, pituitary gland, spinal cord, cerebral vasculature or nerve roots -plus- An abnormal MRI characteristic of neurosarcoidosis, defined as exhibiting abnormal enhancement following the administration of gadolinium or a CSF exam demonstrating inflammation	These criteria were designed to support research based classification of organ system involvement by sarcoidosis in patients with known sarcoidosis.
Marangoni/2006 [e16]	Absence of histological confirmation and alternate inflammatory pathologies have	Signs of inflammation in the central or peripheral nervous system, positive histology for a systemic lesion, and/or positive results for at least two of	Positive nervous system histology	Indirect confirmation of sarcoidosis may risk diagnostic error due to lack of specificity in this clinical context.

	been ruled out.	the following tests: Gallium scan, chest high resolution CT, bronchoalveolar lavage CD4:CD8 ratio > 3.5, and CSF CD4:CD8 ratio >5.		
Joseph/2007 [e17]	Neurosarcoidosis may be diagnosed if there are symptoms not due to other conditions but other criteria are not fulfilled.	Neurosarcoidosis can be diagnosed if the symptoms are suggestive, there is evidence of CNS inflammation (e.g. CSF and MRI), and other diagnoses have been excluded. A diagnosis of systemic sarcoidosis is not essential.	Neurosarcoidosis can only be diagnosed by plausible symptoms, a positive biopsy and no other possible causes for the symptoms.	This definition requires histologic confirmation of neural tissue for a high-likelihood diagnosis of neurosarcoido-sis.
Zajicek/1999 [e18]	Clinical presentation suggestive of neurosarcoidosis with exclusion of alternative diagnoses where	Clinical syndrome suggestive of neurosarcoidosis with laboratory support for CNS inflammation (elevated levels of CSF protein and/or cells, the	Clinical presentation suggestive of neurosarcoidosis with exclusion of other possible diagnoses and the presence of positive nervous system histology.	The Zajicek criteria are widely used, often with modifications.

	the above [probable] criteria are not met	presence of oligoclonal bands and/or MRI evidence compatible with neurosarcoidosis) and exclusion of alternative diagnoses together with evidence for systemic sarcoidosis (either through positive histology, including Kveim test, and/or at least two indirect indicators from Gallium scan, chest imaging and serum ACE).		
Sakuta/2006 [Footnote 2] [e19]	Positive clinical findings which suggest neurosarcoidosis. At least two of six examinations are positive.	Positive clinical findings which suggest neurosarcoidosis. Pathology proven in other organ. At least two of six examinations are positive.	Positive clinical findings which suggest neurosarcoidosis. Pathology proven case.	Same critiques as above.
Stern/2014 [e20]	The clinical syndrome and diagnostic	The clinical syndrome and diagnostic evaluation suggest	The clinical presentation is suggestive of neurosarcoidosis. Other	

	evaluation suggest neurosarcoidosis. However, alternate conditions, such as an infection or malignancy, are not excluded or there is no pathologic confirmation of systemic sarcoidosis.	neurosarcoidosis. Alternate diagnoses are excluded. There is pathologic confirmation of systemic sarcoidosis.	diagnoses are excluded. In addition, there is supportive nervous system pathology, or criteria for “probable neurosarcoidosis” are met, with the patient having a beneficial response to immunotherapy over a [6 months - 1] year or longer observation period.	
Gelfand/2014 [Footnote 3] [e21]	Clinical syndrome consistent with granulomatous inflammation infiltrating the nervous system but without confirmed sarcoidosis anywhere in the body.	Neurologic syndrome consistent with granulomatous infiltration of the nervous system in a patient with confirmed sarcoidosis outside the nervous system.	Histologic evidence of granulomatous inflammation consistent with sarcoidosis on a biopsy from the nervous system.	Includes criteria for “Para-neuro-sarcoidosis”: A neurologic syndrome possibly associated with having sarcoidosis but not known to result directly from granulomatous inflammation within the nervous system. Sarcoidosis- associated small fiber neuropathy would be in this category.

Spiegel/2012 [e22]	The criteria for Probable and Definite are not met.	[a] Laboratory evidence of CNS inflammation (elevated levels of CSF protein and/or cells, the presence of oligoclonal bands and/or MRI evidence compatible with NS), and [b] Evidence of systemic sarcoidosis (either through positive histology, including Kveim test, and/or at least two indirect indicators from Gallium scan, chest imaging and serum ACE	Positive CNS histology	
Fritz/2016 [e23]	Clinical suspicion and exclusion of other diagnoses, but criteria for Probable and Definite are not met	Evidence of nervous system inflammation on MRI or CSF (elevated protein, cells, immunoglobulin G indices, or presence of oligoclonal bands) in combination with evidence of systemic	Histologic confirmation of non-caseating granulomas of affected nervous system tissue	

		sarcoidosis with histological confirmation and/or at least two of the indirect indicators consisting on FDG PET, gallium scan, chest imaging, and serum ACE		
Gelfand and Bradshaw/2017 e24		Biopsy-confirmed sarcoidosis in an organ outside the nervous system and a neurological syndrome consistent with granulomatous inflammation along with rigorous exclusion of other causes	A CNS biopsy consistent with sarcoidosis and a neurological syndrome consistent with granulomatous inflammation along with rigorous exclusion of other causes	

Footnote 1. Two criteria were required to be fulfilled to apply the criteria: 1) histologic evidence of granulomatous inflammation of unknown cause … in at least one organ that was not being assessed; 2) the clinical manifestation being assessed required that all alternative causes other than sarcoidosis … had been reasonably excluded.

Footnote 2. At least two of six examinations need to be positive: bilateral hilar lymphadenopathy, abnormal uptake of 67 Ga scintigraphy, bronchoalveolar lavage fluid examination, elevated serum ACE, negative tuberculin reaction, and elevated serum or urinary calcium level.

Footnote 3. Assumes rigorous exclusion of other potential causes to the neurologic syndrome, particularly infection and malignancy.

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