

NEUROSARCOIDOSIS

Dr. Varvara Avdeeva
Russian University of Medicine
HMC – Dr. Muhammad Faisal Khan

Eyes
12-23%



Nervous system
3-9%



Parotid glands
3-4%

Upper respiratory tract
3-10%

Lymph nodes
12-15%

Heart
2-5%



Calcium dysregulation
4-7%

Lung
89-95%



Kidney
1%

Bone marrow
4-8%

Liver
12-20%



Bones/joints
1-7%

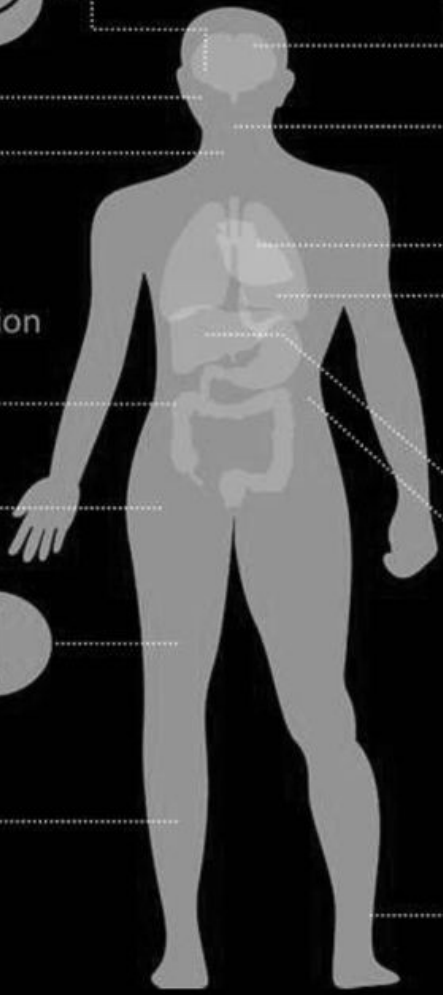


Spleen
7%



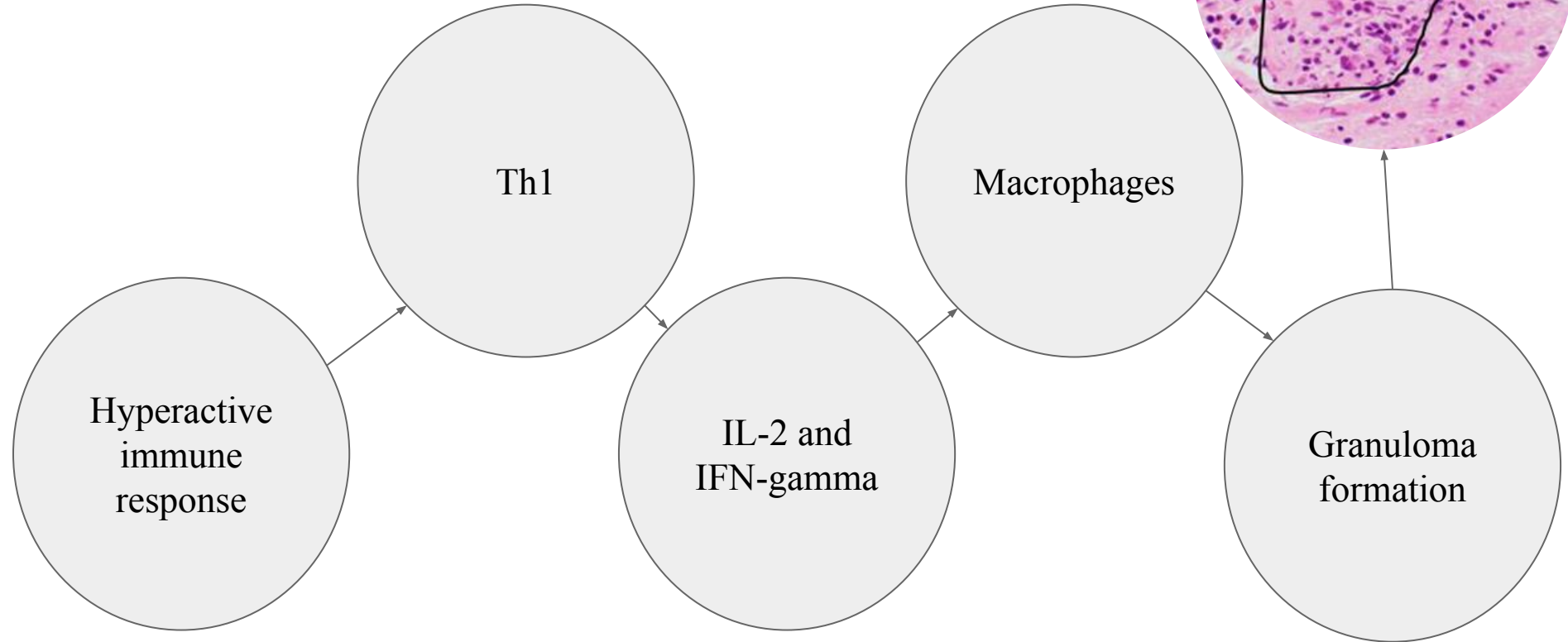
Muscles
0.4-1%

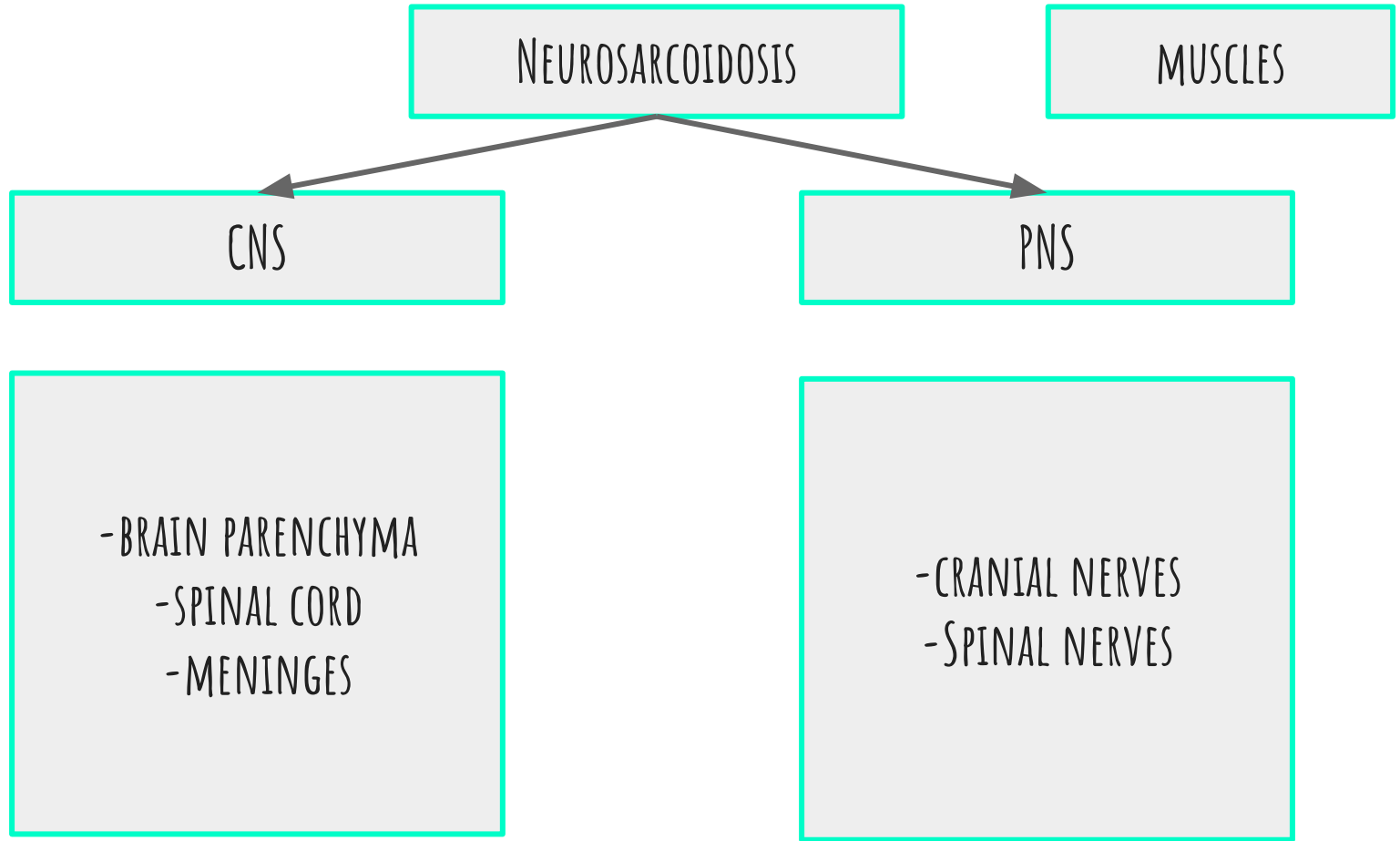
Skin
16-32%

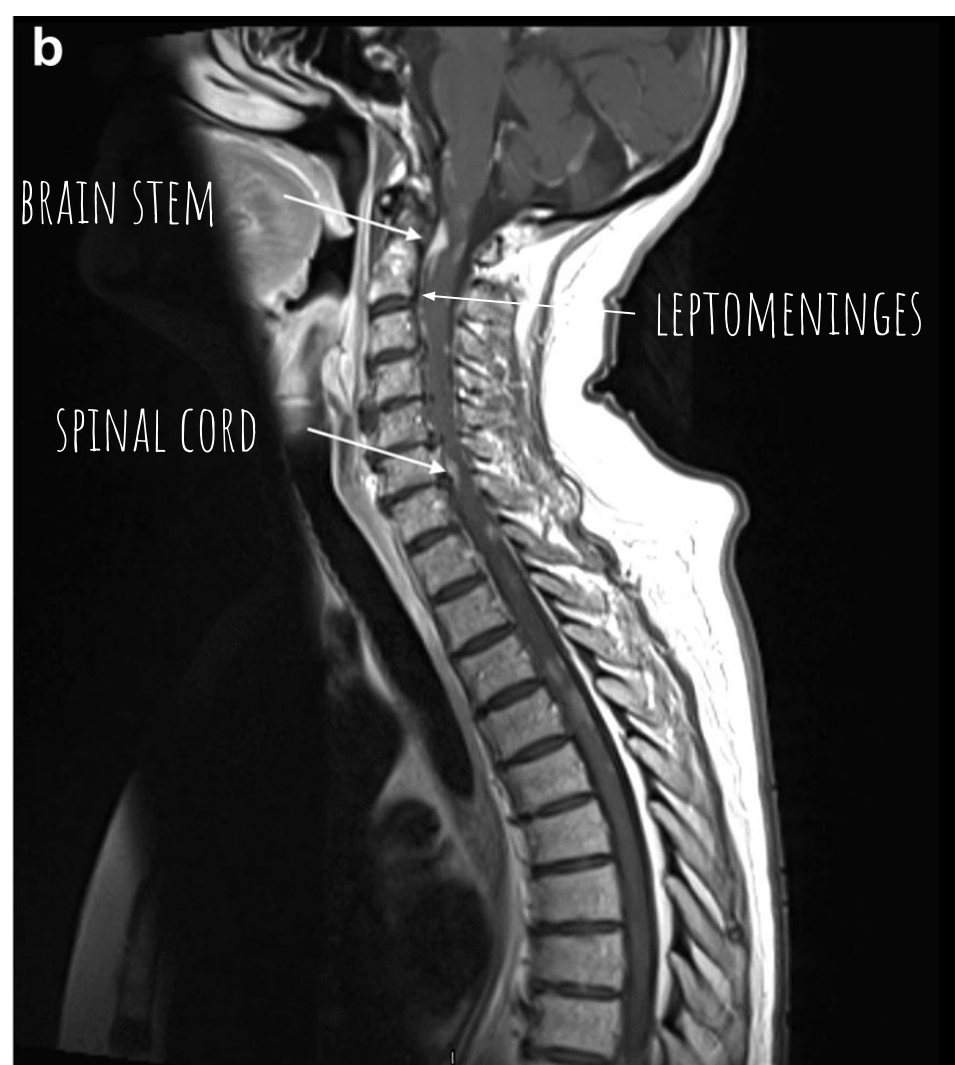
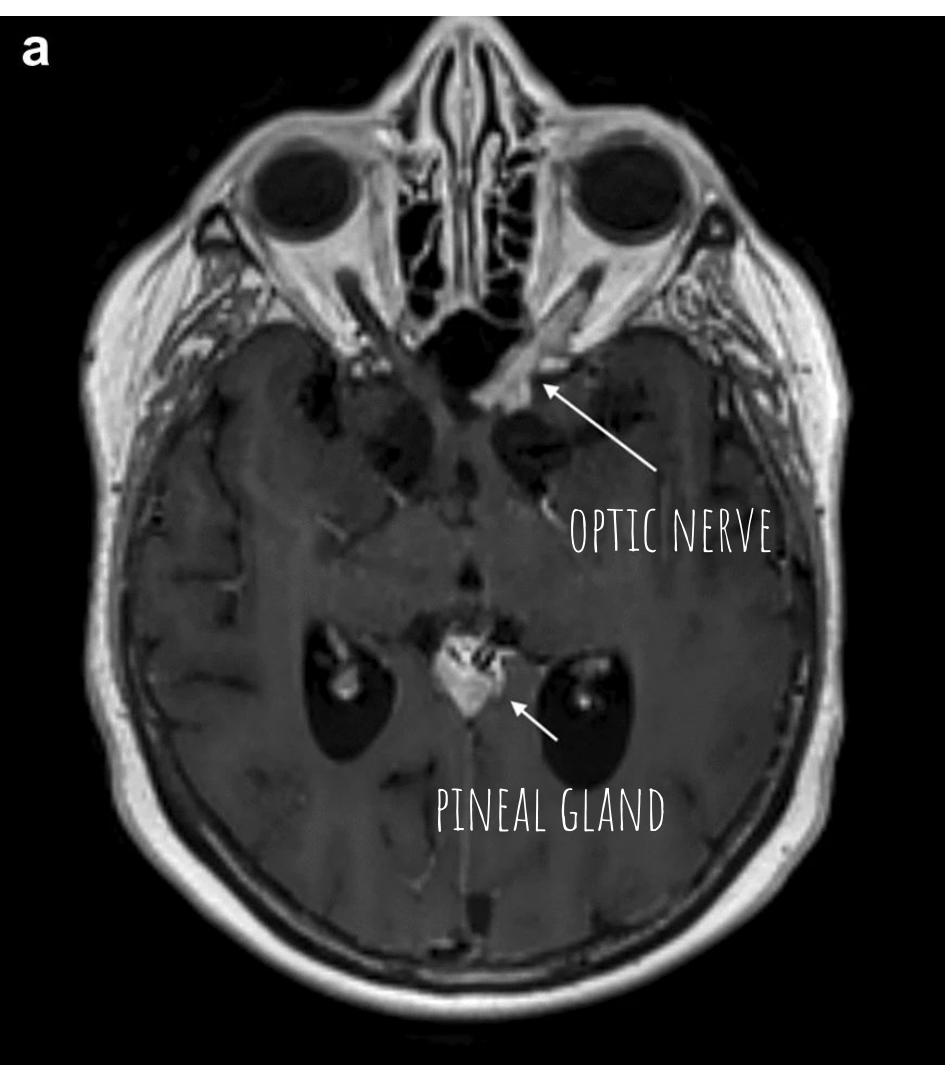


SARCOIDOSIS IS A RARE SYSTEMIC INFLAMMATORY DISEASE CHARACTERIZED BY THE FORMATION OF NONCASEATING GRANULOMAS

ETIOLOGY







EPIDEMIOLOGY

- Prevalence of sarcoidosis in USA is 152-215 per 100 000
 - Greatest occurrence among African American females 20-40 years of age
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- Neurosarcoidosis occurs in 5%–25% of all patients with sarcoidosis
 - Neurologic manifestations are the presenting syndrome in 50%–70% of patients with neurosarcoidosis

MANIFESTATION

Peripheral neuropathy	86%
Cranial neuropathies	75%
Brain parenchymal disease	50%
mass lesion	15%
seizure	10%
encephalopathy	10%
Myelopathy	26%
Aseptic meningitis	20%
Myopathy	10%
Hydrocephalus	10%
Hypothalamic/pituitary involvement	8%

CN 2	35%
CN 3	2%
CN 4	4%
CN 5	9%
CN 6	9%
CN 7	25%
CN 8	17%
CN 9	2%
CN 12	2%

PERIPHERAL NEUROPATHY

- Pure motor, sensory, or mixed sensorimotor
- Large and small fiber polyneuropathies
 - Large f. p. - loss of joint position and vibration sense, sensory ataxia
 - Small f. p. - impairment of pain, temperature and autonomic functions
- Symmetric chronic sensorimotor neuropathy with axonal features on electromyogram (EMG) is reported to be the most common

CRANIAL NEUROPATHY (CN 2,7,8)

- Granulomatous infiltration of cranial nerve nuclei, fascicles or nerves
- Typically multiple concurrent or serial cranial neuropathies
- Typically a subacute, progressive course

- Optic neuritis is slightly more bilateral > unilateral
 - Visual recovery even with treatment can be poor

- Typically facial nerve palsy is recurrent, bilateral

HEERFORDT SYNDROME

parotitis

facial
nerve palsy

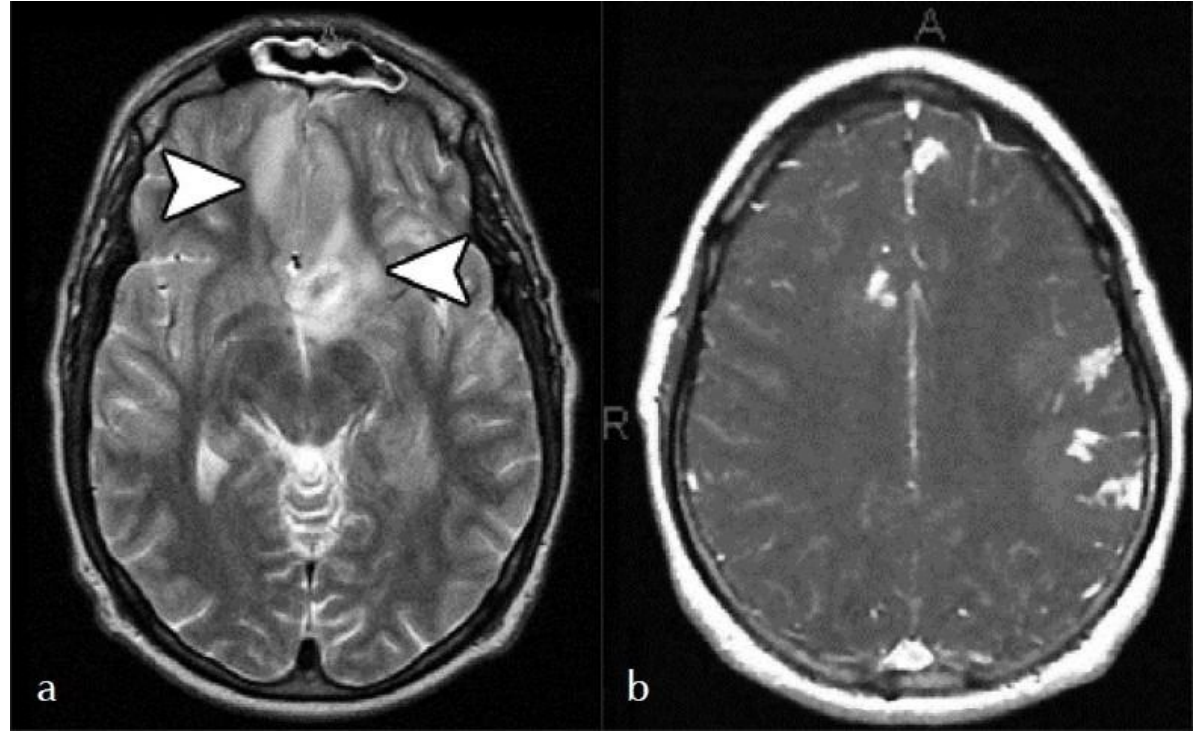
anterior
uveitis

low-grade
fever



BRAIN PARENCHYMAL DISEASE

- Mass lesion
- Seizure
- Encephalopathy (dementia)
- Cerebrovascular disease (vasculitis, vascular compression, venous sinus thrombosis)
- Neuropsychiatric disease (depression, psychosis)



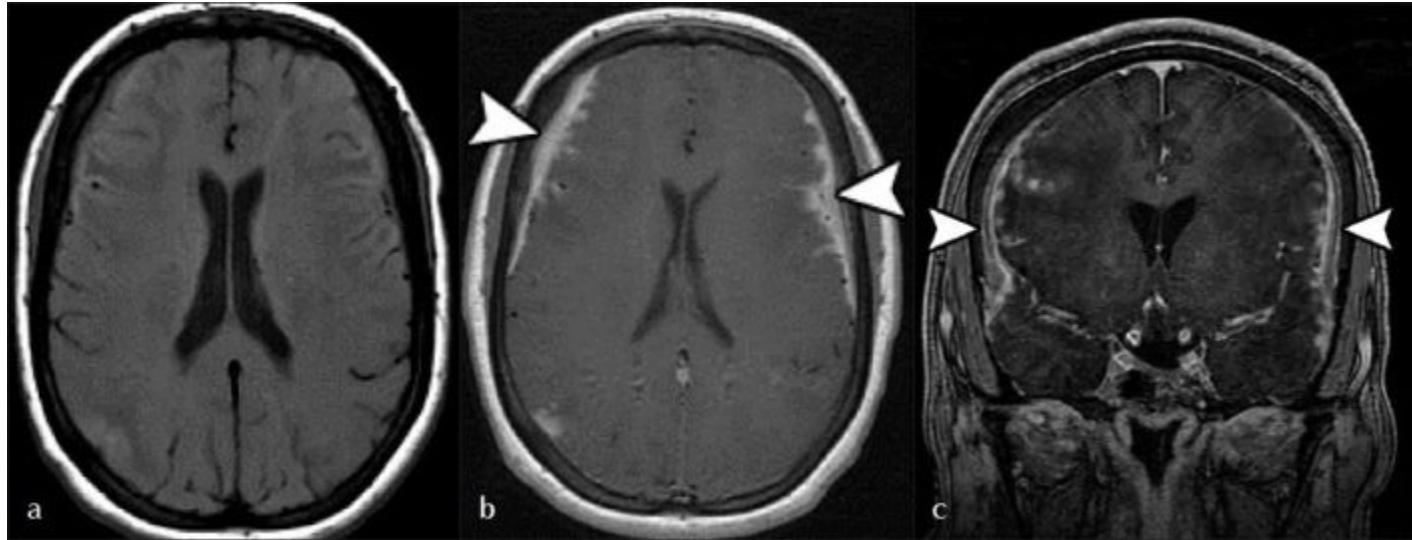
(a) T2 (b) post-contrast T1

MYELOPATHY

- Longitudinally extensive myelitis (LETM)
≥3 vertebral segments
- Common (75% of neurosarcoidosis myelitis)



MENINGITIS



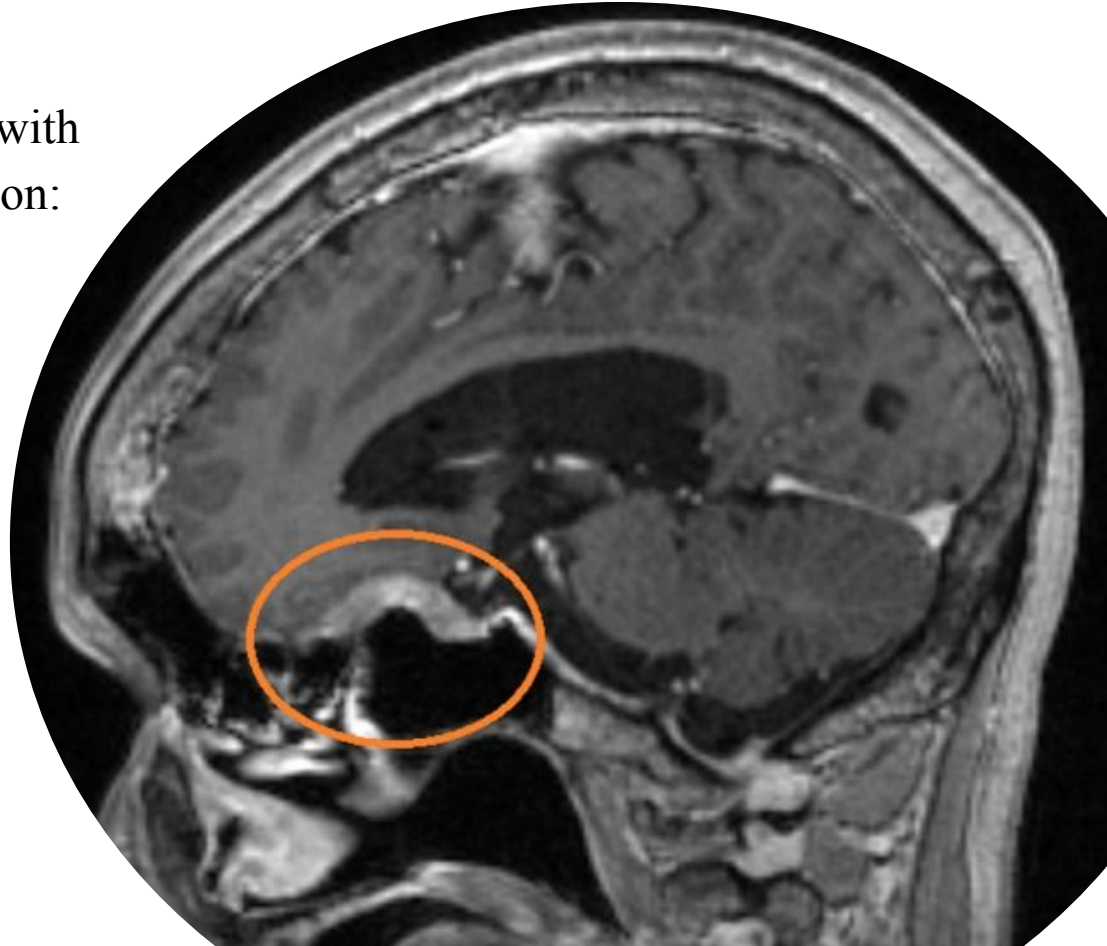
- Chronic meningitis → communicating hydrocephalus
- Predilection: base of the skull (basilar meningitis)
- Complications: cranial nerve dysfunction, seizures

Leptomeningeal Involvement
(a) pre-contrast T1
(b,c) post-contrast

SELLAR DISEASE

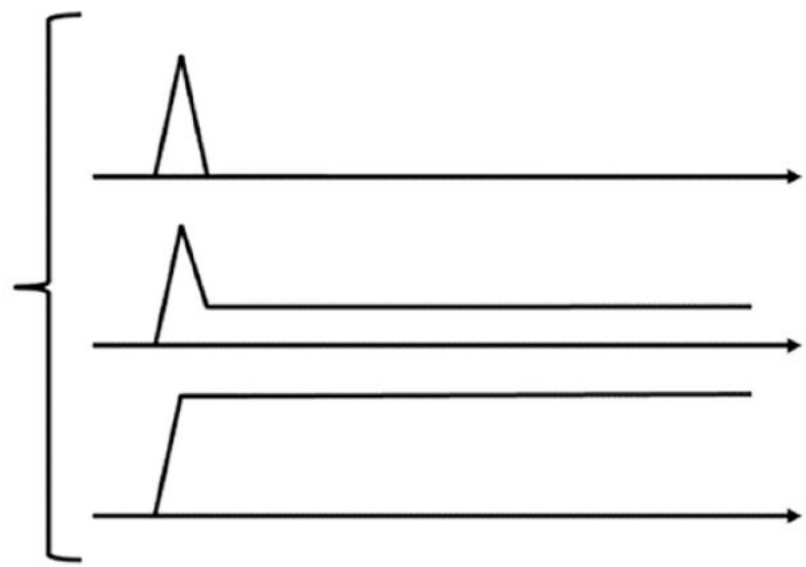
Hypothalamic/pituitary involvement with consequent neuroendocrine dysfunction:

- LH/FSH 89%
- TSH 67%
- Diabetes insipidus 65%
- GH 50%
- ACTH 49%
- Hyperprolactinemia 49%



TYPES OF DISEASE COURSES

Monophasic

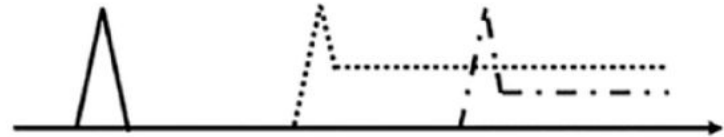


relapsing-remitting
or progressive

Polyphasic with
same symptoms



Polyphasic with
different symptoms



DIAGNOSTIC CRITERIA

1. **The clinical presentation and diagnostic evaluation suggest neurosarcoidosis**, as defined by the clinical manifestations and MRI, CSF, and/or EMG/nerve conduction study findings typical of granulomatous inflammation of the nervous system and **after rigorous exclusion of other causes.**

2. Pathologic confirmation of:

- No pathologic confirmation → **Possible** neurosarcoidosis
- Systemic granulomatous disease consistent with sarcoidosis → **Probable** neurosarcoidosis
- Neurosarcoidosis → **Definite** neurosarcoidosis

DIFFERENTIAL DIAGNOSIS WORKUP

1. Serum studies (CBC, HIV and syphilis serology, ANA, AQP4-Ig, ...)
2. CSF analysis (cell count, culture, ...)
3. Imaging (MRI, CT, PET/CT, CXR)
4. Biopsy (with acid-fast and fungal staining)
5. EEG
6. EMG/nerve conduction study
7. EKG
8. Skin examination
9. Dilated eye examination with slit lamp

CSF ANALYSIS

- Mild to moderate pleocytosis (<100 cells/ μ L)
- Lymphocyte predominance, neutrophils present acutely
- Elevated protein
- Hypoglycorrhachia (20-50 mg/dL)

Oligoclonal bands and elevated IgG index may be seen in 20–40%, nonspecific

TREATMENT

Medications:

- Glucocorticoids (Prednisone, Methylprednisolone)
- Immunosuppressant agents (Azathioprine, Methotrexate, Mycophenolate mofetil)
- Tumor necrosis factor inhibitors (Infliximab, Adalimumab)
- B Cell–Targeted Therapy (Rituximab)
- JAK inhibitor (Tofacitinib, Baricitinib)

Neurosurgery:

- Biopsy/excision procedures that involve the CNS can be associated with severe consequences, and remain a treatment of last choice
- Ventriculoperitoneal shunt for cases with hydrocephalus

PROGNOSIS

Clinical outcome varies substantially among patients with neurosarcoidosis depending on the severity, extent, and neuroanatomic localization of the underlying disease.

Typically:

- Cranial mononeuropathies: improvement
- Aseptic meningitis, meningeal/parenchymal mass lesions: chronic course
- Encephalopathy/vasculopathy: relapsing-remitting course with deterioration
- Peripheral neuropathy, myopathy: remissions, chronic and progressive course

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THANK YOU