Neurosarcoidosis

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**Sarcoidosis In General** → see [p. 2150-2150 (3) >>](http://WWW.NEUROSURGERYRESIDENT.NET/USMLE%202/Respiratory%20system%20%282101-2200%29/2150%20%283%29.%20Sarcoidosis.pdf)

Neurosarcoidosis – part of systemic granulomatous disease.

* commonly involved organs: lungs, skin, lymph nodes, bones, eyes, muscles, parotid glands.
* incidence of sarcoidosis is 3-50 cases/100,000 population.
* 5% sarcoidosis patients have CNS involvement.
* 1-3% neurosarcoidosis patients have no systemic manifestations.

Etiopathophysiology

* cause is unknown.
* exaggerated cellular immune response.

Pathology

* primarily involves **leptomeninges** (parenchymal invasion also often occurs) - ***adhesive*** arachnoiditis with ***nodule*** formation (nodules have predilection for posterior fossa), meningitis or meningoencephalitis - at the base of the brain (**basal** meningitis) and in the subependymal region of the **third ventricle**.
* ***noncaseating granulomas with lymphocytic infiltrates***; Langhans giant cells may or may not be present.

Clinical Features

1. Diabetes insipidus (hypothalamic involvement) - most common neurologic manifestation!
2. Multiple cranial nerve palsies (esp. facial diplegia)
3. Intracranial hypertension - common
4. Hydrocephalus - from adhesive basal arachnoiditis
5. Seizures occur in 15%
6. Peripheral neuropathy
7. Myopathy
8. Low grade fever
* median age of onset of neurologic symptoms - 44 years.

Diagnosis

Laboratory

1. Mild **leukocytosis** and **eosinophilia**.
2. ↑**Angiotensin-converting enzyme (ACE)** in serum:
3. in 83% of patients with active pulmonary sarcoidosis, but in only 11 % with inactive disease
4. in 55% of cases with neurosarcoidosis
5. **CSF** (similar to any *subacute meningitis*): elevated pressure, mild pleocytosis (10-200 cells) mostly lymphocytes, elevated protein (up to 2,000 mg), mild hypoglycorrhachia (15-40 mg/dl).

Imaging

CXR

Characteristic findings of sarcoidosis (hilar adenopathy, mediastinal lymph nodes).

FLAIR-MRI

* gadolinium enhancement of **leptomeninges** and/or **optic nerve**
* lesions may be *solitary* *or* multiple
* lesions may be located *intra- or extraparenchymal, periventricular*, in *basal cisterns*.
* **hydrocephalus** may occur.

Gallium Scan

- with **67Ga citrate**:

1. **Panda sign**: uptake in lacrimal glands, parotid glands & nasopharynx (normal). Not specific for sarcoidosis.
2. **Lambda distribution**: uptake in hilar lymph nodes.
3. **Leopard man sign**: diffuse dappled pattern due to uptake in soft tissues, skin, muscles, mediastinum, and lacrimal glands

Biopsy

- in uncertain cases.

* biopsy should include all layers of meninges and cerebral cortex.
* cultures and stains for ***fungus*** and ***acid-fast bacteria (TB)*** should be performed in addition.

Diffuse cerebral neurosarcoidosis mimicking gliomatosis cerebri:



Treatment

**Corticosteroids** - beneficial for systemic as well as neurologic involvement.

* initiated with prednisone 60 mg PO qd in adults, and tapered based on response.
* cyclosporine may allow a reduction in steroid dosage in refractory cases.

Treatment for unresponsive cases: methotrexate, cytoxan, cyclophosphamide, azathioprine, low dose XRT.

Prognosis

* benign disease.
* peripheral and cranial nerve palsies recover slowly.

Bibliography for “Neurosarcoidosis”:

Mark S. **Greenberg** “Handbook of Neurosurgery” 7th ed. (2010); Publisher: Thieme Medical Publishers; ISBN-10: 1604063262 ISBN-13: 978-1604063264 [>>](http://www.amazon.com/gp/product/1604063262)

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