

# Neurosarcoidosis

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ETIOPATHOPHYSIOLOGY.....	1
PATHOLOGY.....	1
CLINICAL FEATURES .....	1
DIAGNOSIS.....	1
LABORATORY .....	1
IMAGING .....	1
TREATMENT.....	2
PROGNOSIS.....	2
SARCOIDOSIS IN GENERAL → see p. 2150-2150 (3) >>	

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:
  - a) in 83% of patients with active pulmonary sarcoidosis, but in only 11 % with inactive disease
  - b) in 55% of cases with neurosarcoidosis
3. **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 **<sup>67</sup>Ga citrate**:

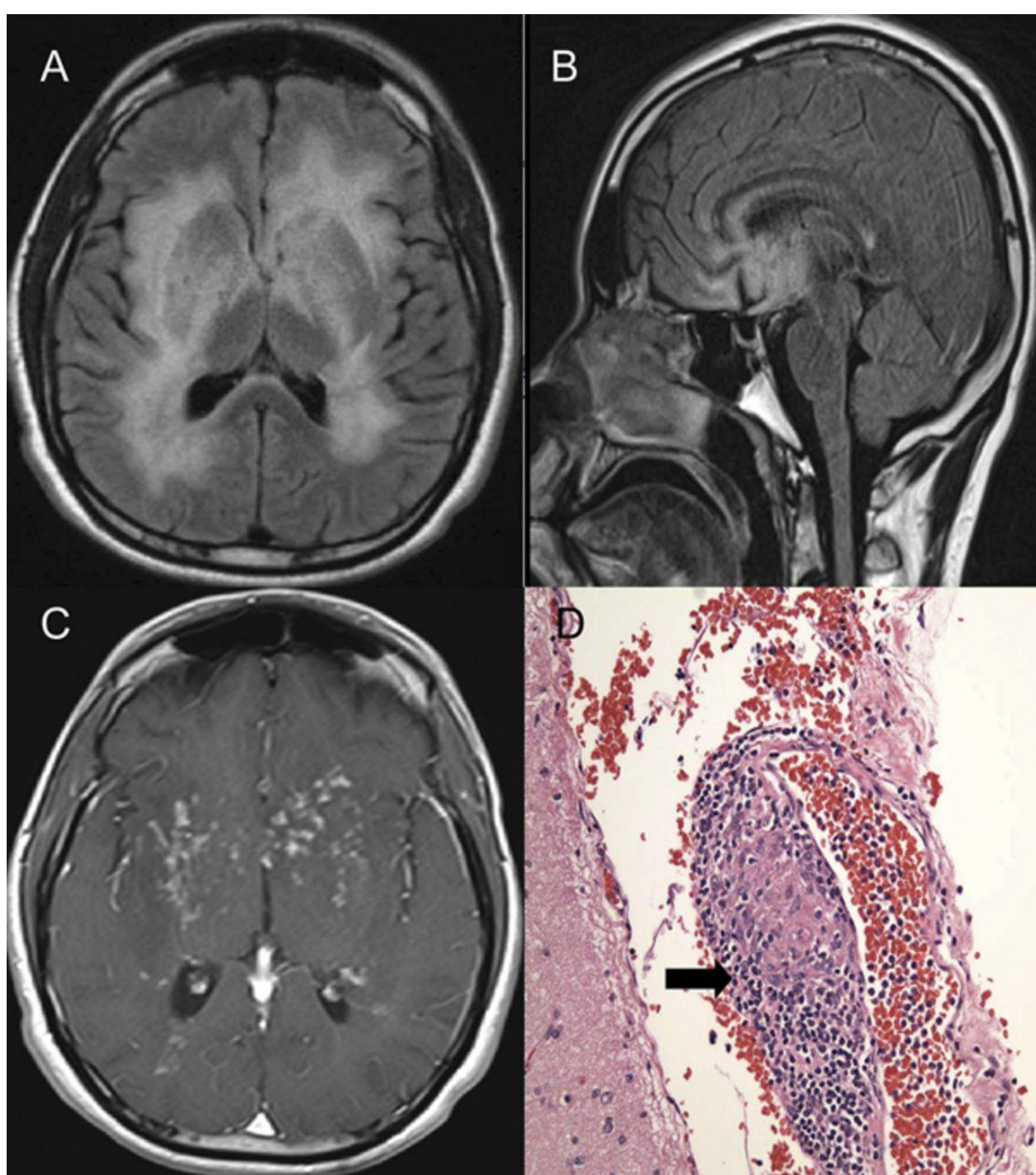
1. **Panda sign**: uptake in lacrimal glands, parotid glands & nasopharynx (normal). Not specific for sarcoidosis.
2. **Lambda distribution**: uptake in hilar lymph nodes.
3. **Leopard man sign**: diffuse dappled pattern due to uptake in soft tissues, skin, muscles, mediastinum, and lacrimal glands

#### 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 **PREDNISON** 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" 7<sup>th</sup> ed. (2010); Publisher: Thieme Medical Publishers; ISBN-10: 1604063262 ISBN-13: 978-1604063264 >>