Neurosarcoidosis

Last updated: May 8, 2019

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

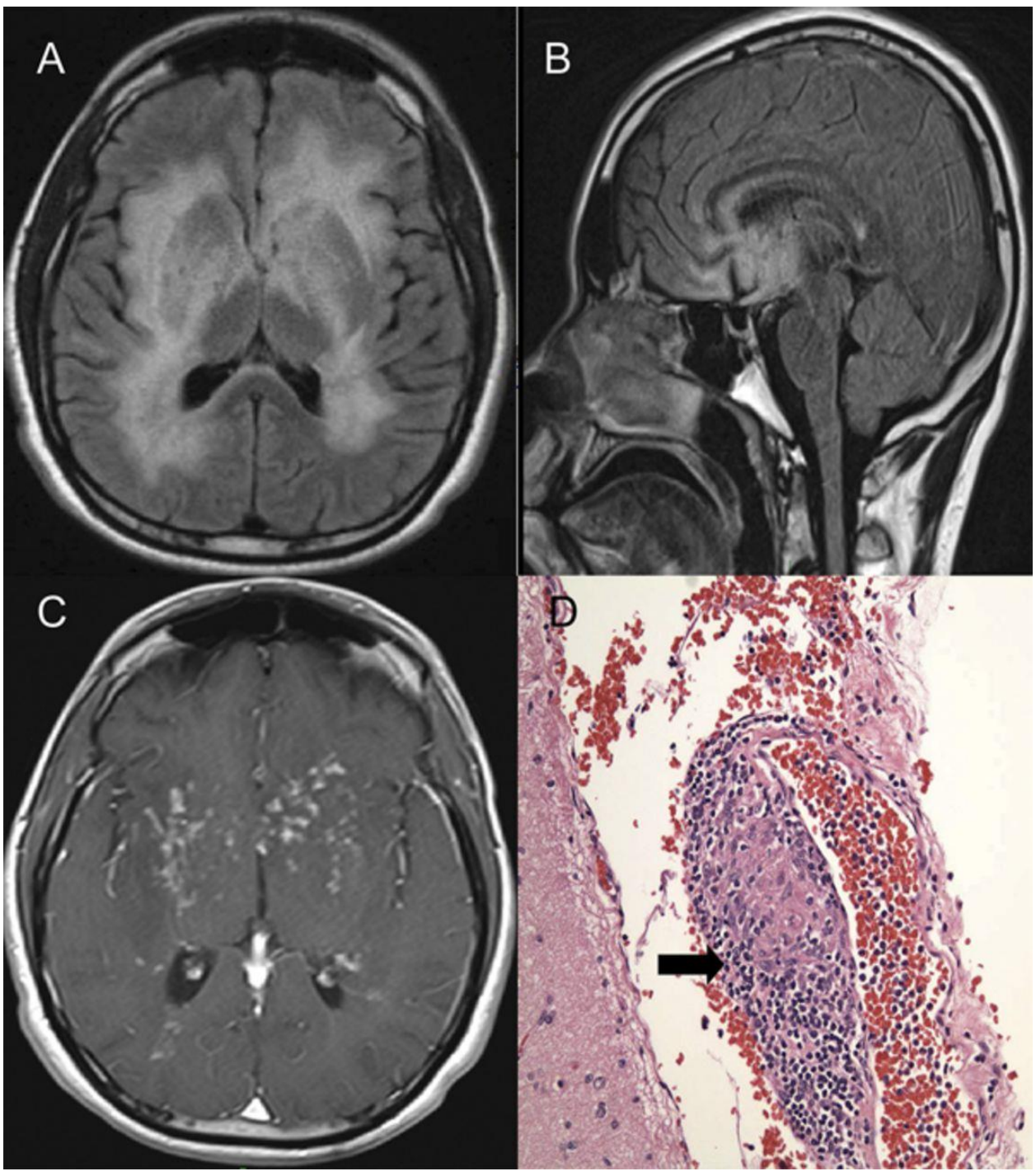
- with ^{67}**Ga citrate**:
    1. **Panda sign**: uptake in lacrimal glands, parotid glands & nasopharynx (normal). Not specific for sarcoidosis.
    2. **Lambda distribution**: uptake in hilar lymph nodes.
    3. **Leopard man sign**: diffuse dappled pattern due to uptake in soft tissues, skin, muscles, 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 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 >>