Neurosarcoidosis

ETIOPATHOPHYSIOLOGY

**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 meninges) and in the subependymal region of the third ventricle.

**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

**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 *medially* or *extraparenchymal, periventricular, in basal cisterns.*
- *hydrocephalus* may occur.

**GALLIUM** SPECT

**BRAIN**
- 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.

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.

**SARCOIDOSIS IN GENERAL**
- see p. 2150-2150 (3)
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**

**Terminally stage**

- METHOTREXATE, CYTOXAN, CYCLOPHOSPHAMIDE, AZATHIOPRINE, low dose XRT.

**PROGNOSIS**

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

**BIBLIOGRAPHY** for “Neurosarcoïdosis”:


**Viktor’s Notes℠ for the Neurosurgery Resident**

Please visit website at www.NeurosurgeryResident.net