Neurosarcoidosis

Pathology

Gross involvement

CNS sarcoidosis primarily involves the leptomeninges; however, parenchymal invasion often occurs. Adhesive arachnoiditis with nodule formation may also occur (nodules have a predilection for the posterior fossa). Diffuse meningitis or meningoencephalitis may occur and may be most pronounced at the base of the brain (basal meningitis) and in the subependymal region of the third ventricle (including the hypothalamus). Spinal involvement may include arachnoiditis, and lesions that may be intramedullary, extramedullary intradural and extramedullary extradural.

Microscopic features

Constant microscopic features of neurosarcoidosis include noncaseating granulomas with lymphocytic infiltrates. Langhans's giant cells may or may not be present.

Neurosarcoidosis generally occurs only in cases of sarcoidosis with substantial systemic involvement, and signs of neurologic involvement usually are seen in patients known to have active disease. Strictly neurologic forms are seen in fewer than 10% of patients; a subset has predominantly neuromuscular involvement.

A definitive diagnosis of neurosarcoidosis requires the exclusion of other causes of neuropathy and the identification of noncaseating sarcoid granulomas by histologic analysis of nerve and muscle biopsy specimens (see Workup).

Clinical features

Clinical findings include multiple cranial nerve palsies in 50–70% (particularly facial n., including diplegia), peripheral neuropathy, and myopathy [1)].

Occasionally the lesions may produce mass effect, [2)] and hydrocephalus may result from adhesive basal arachnoiditis. Patients may have low grade fever. Intracranial hypertension is common and may be dangerous. Hypothalamic involvement may produce disorders of ADH (diabetes insipidus, disordered thirst). Rare involvement of the pituitary may produce pituitary insufficiency. Seizures occur in 15%. Spinal cord involvement may produce myelopathy.

Treatment

Neurosarcoidosis Treatment.

Outcome

Neurosarcoidosis has no known cure. Spontaneous remission has been observed, but long-term therapy often is required. Immunosuppression is the principal method of controlling the disease, and corticosteroids are the cornerstone of therapy.

Case reports

A 62-year-old woman was transported to our hospital for management of generalized clonic seizures. Cerebrospinal fluid examination showed an increased monocyte-dominant cell count, high protein concentration, and low glucose concentration that was 17% of the plasma glucose concentration. Contrast-enhanced cranial magnetic resonance imaging revealed diffuse leptomeningeal enhancement with multiple nodular lesions. She underwent examinations that ruled out the following conditions: tuberculous meningitis, systemic sarcoidosis, malignant lymphoma, carcinomatous meningitis, and central nervous system vasculitis. On hospital day 13, dural and brain biopsies revealed neurosarcoidosis, for which steroid therapy was administered. Thereafter, imaging examinations showed marked improvement. Because isolated neurosarcoidosis is difficult to diagnose, early pathologic diagnosis may be essential [3].

1)
Oksanen V. Neurosarcoidosis: Clinical Presentation and Course in 50 Patients. Acta Neurol Scand. 1986; 73:283–290 [75] de Tribolet N
2)
de Tribolet N, Zander E. Intracranial Sarcoidosis Presenting Angiographically as a Subdural Hematoma. Surg Neurol. 1978; 9:169–171
3)
Deguchi I, Osada T, Suzuki T, Tabata S, Arai E, Uchino A, Hayashi T, Takahashi S, Momoshima S, Takao M. [Case of isolated neurosarcoidosis requiring differentiation from tuberculous meningitis]. Rinsho Shinkeigaku. 2020 Feb 26. doi: 10.5692/clinicalneurol.cn-001387. [Epub ahead of print] Japanese. PubMed PMID: 32101846.