Sarcoidosis is an idiopathic multisystem disorder characterized by an accumulation of noncaseating epithelioid granulomas in affected organs; neurological symptoms develop in approximately 5% of cases. Neurosarcoidosis may rarely present solely as an intracranial tumor mimicking more common disease processes, such as meningioma. We report on a case of isolated intracranial sarcoid granulomatosis.

This 24-year-old man presented with a 1-month history of recurrent seizures. Magnetic resonance (MR) imaging revealed two nodular homogenous enhancing masses in the left frontal and temporal lobes (Fig. 1). Abnormal leptomeningeal enhancement demonstrated confluence of the lesions. A left frontotemporal craniotomy was performed, and after dural incision a granulated fibrous mass was found closely attached to the leptomeninges and invading the superficial cortex. Complete tumor resection was attempted. Results of histopathological examination of the tumor specimen were consistent with neurosarcoidosis (Figs. 2 and 3). Necrosis and microorganisms were absent, and results of special staining for fungi and acid-fast bacilli were unremarkable. Findings of comprehensive laboratory studies, particularly measurements of angiotensin converting enzyme (ACE) and calcium levels in serum and cerebrospinal fluid (CSF), were unremarkable. Follow-up investigation, including thoracic computerized tomography, bronchoscopic biopsy, and abdominal ultrasonography, revealed no evidence of systemic disease. The patient received adjunctive corticosteroid therapy and was discharged without deficits.

Gadolinium-enhanced MR images yielded the highest sensitivity for detection of neurosarcoidosis. Enhancement of the affected brain parenchyma and meninges is a useful clue to the diagnosis. Intracranial neurosarcoidosis offers a wide spectrum of clinical presentation. Epithelioid cells produce ACE, but ACE levels in serum and CSF are neither specific nor reliably elevated in cases of neurosarcoidosis. It is important to consider neurosarcoidosis, both pre- and intraoperatively, because recognition of this condition may lead to appropriate treatment with steroids and avoid needlessly extensive surgery.

References


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Solitary sarcoid granulomatosis mimicking meningioma

Case illustration

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Fig. 1. Preoperative gadolinium-enhanced coronal MR images revealing two nodular extraxial lesions of the left frontal (1 × 1.5 cm, left) and temporal lobes (2 × 2.5 cm, right). In addition, diffuse leptomeningeal contrast enhancement is demonstrated.

Fig. 2. Photomicrograph of the tumor specimen showing noncaseating granulomas embedded within a network of fibrous stroma. Van Gieson’s, original magnification × 80.

Fig. 3. Photomicrograph of the tumor specimen showing characteristic noncaseating granulomas consisting of follicles with central multinucleated giant cells (some with polymorph conchoid inclusion bodies) surrounded by epithelioid macrophages and fibroblasts. H & E, original magnification × 200.