Central Nervous System Vasculitis

General information

AKA isolated angiitis of the CNS. Rare (≈ 20 cases reported [1] as of 1983); limited to vessels of CNS. Small vessel vasculitis is ≈ always present → in segmental inflammation and necrosis of small leptomeningeal and parenchymal blood vessels with surrounding tissue ischemia or hemorrhage [2].

Clinical features

Combinations of H/A, confusion, dementia, and lethargy. Occasionally seizures. Focal and multifocal brain disturbance occurs in > 80%. Visual symptoms are frequent (secondary either to involvement of choroidal and retinal arteries, or to involvement of visual cortex → visual hallucinations).

Diagnosis

ESR & WBC count is usually normal. CSF may be normal or have pleocytosis and/or elevated protein.

CT may show enhancing areas of low density.

Angiography (required for diagnosis): characteristically shows multiple areas of symmetrical narrowing (“string of pearls” configuration). If normal, it does not exclude the diagnosis.

Histological diagnosis (recommended): all biopsy material should be cultured. Brain parenchyma biopsy infrequently shows vasculitis. Leptomeningeal biopsy invariably shows involvement.

Treatment and outcome

Reportedly fatal if untreated, but may smolder for years.

The rarity of this condition makes treatment uncertain. Recommended: cyclophosphamide (Cytoxan®) 2 mg/kg/d and prednisone 1 mg/kg/d qod therapy.

NB: this condition is thought to be T-cell mediated, but prednisone causes more B-cell suppression, therefore breakthrough during prednisone therapy is not uncommon.

Case reports

A 74-year-old woman with a history of allergic rhinitis, but not asthma, presented with slowly progressive left hemiparesis. Magnetic resonance imaging of the head revealed a heterogeneously enhancing mass involving the right internal capsule and corona radiata. Histological examination of

the resected specimen revealed eosinophil-rich non-granulomatous small vessel vasculitis with no neutrophil infiltration or foci of microbial infection. Epstein-Barr virus in situ hybridization was negative, and polymerase chain reaction tests for both T-cell receptor gamma and immunoglobulin heavy-chain variable region genes did not show rearrangements, excluding the possibility of lymphoma and lymphoproliferative disorders. Blood hypereosinophilia and elevated erythrocyte sedimentation rate were observed; however, anti-neutrophil cytoplasmic antibodies were not detected. A biopsy of the erythema in the hips and thighs revealed perivasculitis with eosinophilic infiltration within the dermis. Chest computed tomography revealed multiple small nodules in the lungs. Her symptoms, aside from hemiparesis, disappeared after corticosteroid administration. The clinicopathological features were similar to eosinophilic granulomatosis with polyangiitis but did not meet its current classification criteria and definition. This patient is the first reported case of idiopathic eosinophilic vasculitis or idiopathic hypereosinophilic syndrome-associated vasculitis affecting the small vessels in the brain. Further clinicopathological studies enrolling similar cases are necessary to establish the disease concept and unravel the underlying pathogenesis [3].

1)
Cupps TR, Moore PM, Fauci AS. Isolated angiitis of the central nervous system: prospective diagnostic and therapeutic experience. Am J Med. 1983; 74:97–105
2)
Moore PM, Cupps TR. Neurologic Complications of Vasculitis. Ann Neurol. 1983; 14:155–167
3)
Noro Y, Miyata H, Furuta T, Sugita Y, Suzuki Y, Kusumi M, Tanabe M, Shomori K. Tumefactive eosinophil-rich non-granulomatous small vessel vasculitis in the cerebrum in a patient with idiopathic hypereosinophilic syndrome. Neuropathology. 2022 Apr 11. doi: 10.1111/neup.12810. Epub ahead of print. PMID: 35411628.