

Isolated CNS vasculitis

General information

AKA isolated angiitis of the CNS. Rare (\approx 20 cases reported³² as of 1983); limited to vessels of CNS. Small vessel vasculitis is \approx always present \rightarrow segmental inflammation and necrosis of small leptomeningeal and parenchymal blood vessels with surrounding tissue ischemia or hemorrhage.

Presentation

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 \rightarrow visual hallucinations). Evaluation ESR & WBC count are 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 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. 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.

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