Polyarteritis Nodosa-Type Vasculitis Isolated to the Central Nervous System: First Case Report of a Fulminant Primary CNS Vasculitis Variant

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Abstract
Primary angiitis of central nervous system (PACNS) is an inflammatory, disease of the blood vessels restricted to the central nervous system (CNS). The histopathology is heterogeneous, but in the majority of adult cases it is a segmental necrotizing granulomatous vasculitis with intramural giant cells (1, 2). Here we report a case of PACNS in which the dominant cell type was polymorphonuclear (PMN) cells.

Case Descriptions
A 57-year-old woman was initially diagnosed at a community hospital with aseptic meningitis after presenting with headache, vertigo, and bilateral eye pain. Her lumbar puncture revealed 100 WBCs with 29% neutrophils, 66% lymphocytes and 40 RBCs. She was discharged on a 10 day oral dexamethasone taper, but the day after completing her taper she presented to our hospital with symptom exacerbation. On admission, CT head with and without contrast revealed a right occipital hyperdensity, leptomeningeal enhancement in the posterior parietal and occipital regions, a small right parietal subdural hematoma, and a left parietal-occipital subarachnoid hemorrhage. CT angiogram and venogram did not reveal any abnormalities. Lumbar puncture on day of admission to our hospital showed 2 WBCs, 1478 RBCs, 174 glucose, 40.3 mg/dl protein. Repeat CSF analysis the next day showed 0 WBCs, 7310 RBCs, 118 glucose and protein of 78.9 mg/dl. ANA, ACE, anti-cardiolipin, C-ANCA, P-ANCA, C3, C4, SS-A, SS-B, anti-Smith, anti-dsDNA, RF, and CCP IgG were negative. ESR was 25 (normal range 0-20) and hs-CRP was 0.286 (normal range 0-3). HIV, RPR, and hepatitis panel, as well as CSF VDRL, arbovirus were negative. CT scan revealed bilateral ACA-PCA watershed infarcts and small subdural hematoma.

CNS biopsy with methylprednisolone 1 gram daily. On hospital day three the patient suddenly lost her vision despite a normal fundoscopic examination. MRI brain with and without contrast revealed bilateral corona radiata ischemia, left subdural hematoma, bilateral parietal-occipital and posterior temporal leptomeningeal enhancement, and left parietal-occipital subarachnoid hemorrhage. She received one dose of intravenous cyclophosphamide 750 mg/m2. A brain and meningeal biopsy were performed the next morning she was dead by brain criteria, confirmed by a cerebral blood flow study.

Pathology & Images
The patient received empirical treatment for HSV encephalitis with acyclovir, TB meningitis with standard four-drug therapy, and CNS vasculitis with intravenous methylprednisolone 1 gram daily. On hospital day three the patient suddenly lost her vision despite a normal fundoscopic examination. MRI brain with and without contrast revealed bilateral corona radiata ischemia, left subdural hematoma, bilateral parietal-occipital and posterior temporal leptomeningeal enhancement, and left parietal-occipital subarachnoid hemorrhage. She received one dose of intravenous cyclophosphamide 750 mg/m2. A brain and meningeal biopsy were performed two days later. At surgery, the patient’s blood vessels were hemorrhagic, and her arachnoid membrane was notably thickened, consistent with an inflammatory process. On hospital day 7, the patient became unresponsive and was intubated for airway protection. She had developed interval bilateral ACA-PCA watershed infarcts as well as bilateral ACA-MCA watershed infarcts and left putamen infarct. The next morning she was dead by brain criteria, confirmed by a cerebral blood flow study.

Discussion
The natural history, patient demographics and disease course of PACNS are highly heterogeneous, and cerebral cortex, white matter, and spinal cord have all been involved. Aggressive cases with rapid progression to coma and death as well as indolent, chronic forms had both been described (3).

Background
Considering the clinical heterogeneity, it is likely that PACNS covers a group of diseases entities, not a single disease. Further contributing to the confusion is the lack of tissue diagnosis on some of the published series. While angiography was often used as the diagnostic method of choice because it carried a low procedural risk, it had been well documented that the sensitivity and specificity of angiography in the diagnosis of PACNS was variable at best (2, 7). Brain and meningeal biopsies remained the gold standard for the diagnosis of PACNS. It served to confirm the diagnosis and to exclude other conditions mimicking CNS vasculitis, which could be up to 39% of cases (8).

References
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