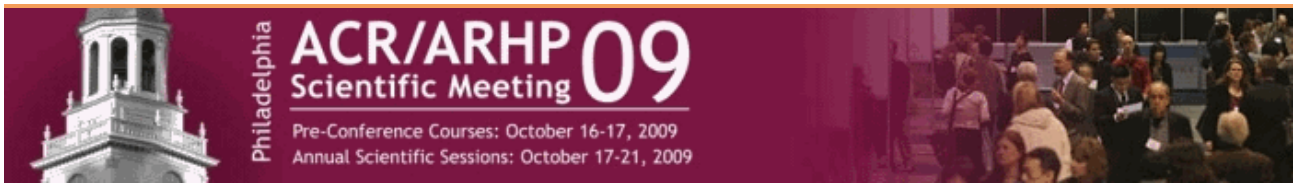

1825 - Catastrophic Primary Central Nervous System Vasculitis

Tuesday, October 20, 2009: 9:00 AM - 11:00 AM
Hall D (Pennsylvania Convention Center)

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Presentation Number: 1825
Poster Board Number: 558

Purpose: Primary Central Nervous System Vasculitis (PCNSV) is an uncommon and heterogeneous condition that affects the brain and spinal cord. Recently, we reported 101 patients with PCNSV seen over a 21-year period (1983-2003). Although most patients responded to therapy, an increased mortality was observed. This study was undertaken to identify the subset of patients who appear to have a catastrophic clinical course in the context of the largest cohort of consecutive patients with PCNSV studied to date.
Methods: The present study extends the previous cohort and includes 131 patients with PCNSV seen over the 25 year period of 1983 to 2007. PCNSV diagnosis was based on brain/spinal cord biopsy or angiograms. Intracranial internal carotid artery and proximal anterior, middle, and posterior cerebral arteries were considered large cerebral vessels. The modified Rankin Scale was used to identify catastrophic disease: patients with Rankin 5 (severe disability) or 6 (stroke death) at diagnosis and/or Rankin 5 or 6 at last follow-up (patients with Rankin < 5 at diagnosis must develop a score of 5 or 6 within 6 months after the diagnosis). We compared patients with catastrophic disease to those without.
Results: 11 cases had catastrophic PCNSV. Cerebral angiography was performed in 10 patients and showed bilateral, multiple, large-vessel changes in 9. Three cases had positive CNS biopsy. Compared with the 120 patients without, the 11 patients with catastrophic vasculitis more frequently had paraparesis/quadriparesis at presentation (36.4% vs 2.5%, $p < 0.001$), angiographic presence of bilateral, large-vessel vasculitis (90% vs 52.9%, $p = 0.04$), and MRI evidence of cerebral infarctions (100% vs 51.4%, $p = 0.004$); those infarctions were more frequently multiple and bilateral (77.8% vs 37.6%, $p = 0.03$) and more frequently involved both the cortex and subcortex on initial MRI (66.7% vs 25.7%, $p = 0.02$). Parenchymal and meningeal gadolinium-enhancing lesions occurred less frequently (0% vs 42.2%, $p = 0.01$). Other differences in those with catastrophic PCNSV that were not statistically significant included a higher frequency of persistent neurological deficit or stroke at presentation (63.6% vs 36.7%) and more patients were treated initially with cyclophosphamide or azathioprine (63.6% vs 47.5%). Cerebrospinal fluid abnormalities and the frequency of relapses/recurrences were similar in the two groups. No patients with catastrophic PCNSV showed a lymphocytic histopathological pattern. Survival of the patients with catastrophic PCNSV was significantly reduced ($p < 0.001$).

Conclusion: Catastrophic PCNSV appears to form a clinical subset of PCNSV characterized by bilateral, multiple, large-cerebral vessel lesions and multiple cerebral infarctions.

Keywords: vasculitis

Disclosure: C. Salvarani, None; **R. D. Brown**, None; **K. T. Calamia**, None; **T. J. H. Christianson**, None; **J. Huston**, None; **J. F. Meschia**, None; **C. Giannini**, None; **D. V. Miller**, None; **G. G. Hunder**, None.
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