

The varicella zoster virus vasculopathies

Clinical, CSF, imaging, and virologic features

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Background: Varicella zoster virus (VZV) vasculopathy produces stroke secondary to viral infection of cerebral arteries. Not all patients have rash before cerebral ischemia or stroke. Furthermore, other vasculitides produce similar clinical features and comparable imaging, angiographic, and CSF abnormalities.

Methods: We review our 23 published cases and 7 unpublished cases of VZV vasculopathy. All CSFs were tested for VZV DNA by PCR and anti-VZV IgG antibody and were positive for either or both.

Results: Among 30 patients, rash occurred in 19 (63%), CSF pleocytosis in 20 (67%), and imaging abnormalities in 29 (97%). Angiography in 23 patients revealed abnormalities in 16 (70%). Large and small arteries were involved in 15 (50%), small arteries in 11 (37%), and large arteries in only 4 (13%) of 30 patients. Average time from rash to neurologic symptoms and signs was 4.1 months, and from neurologic symptoms and signs to CSF virologic analysis was 4.2 months. CSF of 9 (30%) patients contained VZV DNA while 28 (93%) had anti-VZV IgG antibody in CSF; in each of these patients, reduced serum/CSF ratio of VZV IgG confirmed intrathecal synthesis.

Conclusions: Rash or CSF pleocytosis is not required to diagnose varicella zoster virus (VZV) vasculopathy, whereas MRI/CT abnormalities are seen in almost all patients. Most patients had mixed large and small artery involvement. Detection of anti-VZV IgG antibody in CSF was a more sensitive indicator of VZV vasculopathy than detection of VZV DNA ($p < 0.001$). Determination of optimal antiviral treatment and benefit of concurrent steroid therapy awaits studies with larger case numbers.