

CNS Vasculitis Overview (2000)

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Several types of inflammatory vascular CNS disease can present in this way and we thought a brief review of these as they relate to headache might be useful to readers.

Most of the vasculitides involving the CNS affect small and/or medium sized arteries. When small arteries are involved, encephalopathy and seizures are common features. With larger arteries involved, ischemic syndromes are seen. Recurrent headache is a common feature. Intracerebral and subarachnoid hemorrhage can be seen in some varieties. Constitutional signs can be seen, such as fever and malaise. Diagnosis is often difficult, as serologic testing is non-specific and often misleading. Treatment generally involves glucocorticoids and/or immunosuppressants. On or off treatment, immune system dysfunction is generally present, so vigilance regarding infection is important.

Primary angiitis of the CNS (PACNS) presents with headaches, recurrent cerebrovascular ischemic events, and mental status changes, sometimes referred to as subacute encephalopathy. Intracerebral hemorrhage can occur. Spinal cord involvement can produce myelopathy. MRI shows periventricular white matter and other ischemic lesions, and with the use of contrast agents, leptomeningeal enhancement can be seen. Angiographically there is segmental narrowing and/or beading of vessels in the majority of cases. CSF is usually abnormal, with pleocytosis and/or increased protein. Lately there has been increasing discussion about the diagnostic standards to establish the diagnosis of PACNS, as the diagnosis is often solely based on clinical and radiological findings. Two studies examined the sensitivity, specificity and positive predictive value of diagnostic studies in PACNS (2,3). Both studies demonstrated a low positive predictive value for angiography (22%-37%). Sensitivity of tissue biopsy was between 53-83%. As a negative brain biopsy does not rule out vasculitis and bears a significant perioperative risk, brain biopsy is often reserved for cases not responsive to therapy. Treatment options include corticosteroids and cyclophosphamide,

Isolated CNS vasculitis can also be associated with AIDS, herpes zoster infections, meningeal infections of various types (fungal, viral, parasitic, treponemal, etc), drug abuse (amphetamines, cocaine), lymphomas, and amyloid angiopathy. Intravascular lymphoma is a rare disorder which mimics CNS vasculitis and can even respond transiently to corticosteroids. Amyloid angiopathy generally presents as lobar hemorrhage.

Giant cell arteritis involves predominantly external carotid branches, leading to the characteristic headache, but causes visual loss and ophthalmoplegia via involvement of ophthalmic and extraocular nerve arteries. Posterior circulation arteries can be involved rarely, leading to strokes.

Behcet's disease is characterized by oral and genital ulcers and iridocyclitis, but recurrent neurologic manifestations (resulting from a small vessel vasculitis) are prominent, with headache, stroke-like episodes, meningoencephalitis, cranial neuropathies, and ataxia. Erythema nodosum is common. CSF pleocytosis and elevated protein are common. The cause is still unknown.

Systemic vasculitides can also produce CNS syndromes with encephalopathy, headaches, seizures, and strokes, including polyarteritis nodosa (PAN), Churg-Strauss Syndrome (CSS), SLE, and Wegener's granulomatosis (WG). Cranial and peripheral neuropathies are typical of PAN. Upper and lower respiratory tract granulomatous disease is the hallmark of WG which differentiates it from PAN which spares the lungs. Glomerulonephritis is also common, and antibodies to nuclear cytoplasmic antigens (ANCA) are seen in most cases of WG. CSS is characterized by

eosinophilia, asthma, and gastrointestinal symptoms, and neurologic manifestations include encephalopathy, seizures, subarachnoid hemorrhage, chorea, and neuropathies. SLE commonly involves the CNS, with encephalopathy, seizures, cranial neuropathies most common, and strokes less common. When optic neuropathy is present the disease may mimic MS. CSF is usually acellular, but mild pleocytosis can be seen, and protein is generally elevated. Most CNS manifestations of SLE are thought to be due to the numerous microinfarctions seen in the cortex and brainstem

References

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