CENTRAL NERVOUS SYSTEM VASCULITIS

Inflammation of CNS Blood Vessels

Vasculitis is a condition of inflammation affecting blood vessels, including veins, arteries, and capillaries. Most conditions of vasculitis have an autoimmune origin and are characterized by inflammation, bleeding under the skin, ruptured surface (cutaneous) blood vessels, and bruising. Vasculitis of the central nervous system (CNS) is a rare autoimmune disorder characterized by inflammation of the blood vessels that supply the tissues that make up the brain and spinal cord.

Consequences

Vasculitis in these areas leads to blood flow blockage, causing a condition of ischemia or oxygen deprivation. Consequently, the affected blood vessel wall and the tissue supplied by the blood vessel die.

Autoimmune Involvement

In CNS vasculitis the immune system produces antibodies that react with protein antigens in cerebrovascular (blood vessels of the nervous system and brain) tissue. These antibodies form a lattice-like antigen-antibody complex that lodges within the blood vessel walls. In addition, the presence of these complexes causes an immune response in which the immune system's white blood cells infiltrate the blood vessel wall. Immune system chemicals and enzymes are released during this infiltration, which causes swelling and cellular destruction.

Symptoms

Symptoms of CNS vasculitis are variable. Most patients will have a cluster of several symptoms rather than all associated symptoms. Headache is one of the earliest symptoms of CNS vasculitis and it may be accompanied by cognitive changes or symptoms of depression or seizures. Other symptoms include weakness, gait disturbances, seizures, ataxia (an inability to coordinate muscle reflexes), stroke-like symptoms and coma.

Diagnosis

Several procedures are used to diagnose CNS vasculitis. Because CNS vasculitis is an inflammatory disorder, patients typically have an elevated erythrocyte sedimentation rate and C-reactive protein. These are the usual screening tests for CNS vasculitis. Negative results suggest other causes.

Diagnosis of CNS vasculitis usually requires a lumbar puncture and a cerebrospinal fluid (CSF) examination. CSF surrounds the spinal cord. In CNS vasculitis, levels of protein are typically elevated although there is no evidence of bacteria, which are typically seen in conditions of meningitis, and the oligoclonal band pattern seen in multiple sclerosis is
absent. Magnetic resonance imaging (MRI) studies typically show evidence of multiple small blood vessel infarcts or signs of inflammation and breakdown of the normal blood-brain barrier. Cerebral angiography, an imaging procedure used to study the blood vessels of the brain after injection of a contrast dye, may show irregularities or "beading" of blood vessels. Although these procedures may all be used to help diagnose CNS vasculitis, a definitive diagnosis requires a brain biopsy. A brain biopsy involves removing a small piece of the brain for microscopic examination.

**Treatment**

Immunosuppressant medications such as dexamethasone and corticosteroids are the primary therapies used for CNS vasculitis. Patients with severe cerebral edema and inflammation may also require monitoring with an intracranial pressure monitor. Some patients may also require therapies such as plasmapheresis or intravenous immunoglobulins to quickly reduce the number of immune complexes in an effort to reduce inflammation quickly.