Vasculitis

Inflammation of the blood vessels, also known as **vasculitis**, occurs as a part of several **autoimmune** disease states. Many autoimmune diseases are caused by production of **antibodies** (proteins made by the immune system) against the body's own tissues. Because vasculitis can occur in many forms, individuals with vasculitis may have different signs and symptoms. Sometimes vasculitis affects the smallest blood vessels in the body (**capillaries**). Other vasculitis syndromes cause large blood vessel disease, such as **aortic aneurysms** (weakness and bulging of part of the large blood vessel from the heart). Vasculitis can also cause loss of blood flow to the extremities or organs, including the heart, the kidneys, and the brain. The August 8, 2007, issue of *JAMA* includes an article about vasculitis.

TYPES OF VASCULITIS

- Behçet syndrome—usually consists of painful sores in the mouth and is more common in men, especially from the Middle East.
- Buerger disease—related to cigarette smoking. Stopping tobacco use is the only way to treat this type of vasculitis.
- Churg-Strauss syndrome—related to allergic rhinitis and asthma, this can progress to severe whole-body vasculitis.
- Giant cell arteritis (temporal arteritis)—common in older women, this vasculitis causes headache, jaw pain, and visual loss.
- Henoch-Schönlein purpura—more common in children, this type of vasculitis may cause kidney failure or may resolve on its own.
- Kawasaki disease—uncommon type of vasculitis that usually affects children. It may cause a swollen, red tongue. Inflammation of the heart and aorta makes this a particularly dangerous type of vasculitis if not treated rapidly.
- Microscopic polyarteritis—affects the smaller blood vessels of the body, causing kidney failure and lung hemorrhage. A type of antibody called **ANCA** (antineutrophil cytoplasmic antibody) is commonly found.
- Polyarteritis nodosa—affects skin, joints, peripheral nerves, and blood vessels of the kidney.
- Polymyalgia rheumatica—a disease of adults, this vasculitis often causes joint pain and stiffness.
- Rheumatoid vasculitis—associated with rheumatoid arthritis, this type of vasculitis affects small blood vessels, especially in the skin.
- Wegener granulomatosis—usually affects the lungs, the kidneys, and the skin. **Granulomas** (compact, solid inflammatory lesions) may occur.

DIAGNOSIS

A detailed medical history and physical examination may lead to diagnosis of a specific type of vasculitis. Vague symptoms may occur at the beginning of a vasculitis-related illness; these may include fever, malaise (feeling ill), nausea, weakness, weight loss, poor appetite, and fatigue. Each type of vasculitis has specific signs and symptoms associated with it. Based on a person's symptoms, testing for vasculitis may include blood tests, x-rays, angiograms (x-ray dye studies of blood vessels), biopsies of involved tissue, or urine testing.

TREATMENT

Medications commonly used to treat all types of vasculitis include

- Steroids
- Nonsteroidal anti-inflammatory drugs (NSAIDs) that may be helpful for some individuals
- Cytotoxic drugs (including cyclophosphamide, often used as part of cancer chemotherapy)
- Immune suppressants (such as azathioprine, mycophenolate mofetil, and cyclosporine)

Each of the vasculitis syndromes has its own best method of treatment. Intravenous immune globulin can be helpful in certain types of vasculitis, particularly Kawasaki disease. Surgical treatment may be offered for aortic and other types of aneurysms.

FOR MORE INFORMATION

- American Heart Association
  www.americanheart.org
- National Heart, Lung, and Blood Institute
  www.nhlbi.org
- Vasculitis Foundation
  www.vasculitisfoundation.org

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Sources: American Heart Association; National Heart, Lung, and Blood Institute; Vasculitis Foundation

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