VASCULITIS

Blood Vessel Inflammation and its Consequences

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Vasculitis includes more than 20 organ specific and systemic disorders that cause inflammation in the veins and arteries as well as the organs that these vessels supply.

What is Vasculitis?

Vasculitis, which is sometimes called angiitis, is a condition of blood vessel inflammation. Blood vessels include the veins, arteries, capillaries and their branches that make up the circulatory system. Blood vessels carry blood to and from the heart, nourishing and oxygenating the body's organs and tissues.

Conditions of vasculitis cause inflammation of veins and arteries along with their smaller tributaries, the venules and arterioles. More than 20 distinct conditions cause vasculitis, with most disorders of vasculitis having an autoimmune origin. When arteries are affected, the disorder may be called arteritis, and when the skin is primarily affected the disorder is known as cutaneous vasculitis. When the veins and arteries of multiple organs and systems are affected, the condition is known as systemic vasculitis. The kidney is one of the most common organs to be damaged in systemic vasculitis.

Conditions of Vasculitis

The specific conditions of vasculitis are distinguished by the size and type of blood vessel affected and also its location in the body. For instance, central nervous system vasculitis affects the small vessels of the brain and central nervous system.

Conditions of autoimmune vasculitis include:

* Temporal arteritis
* Wegener granulomatosis, which primarily involves vessels of the upper respiratory tract, lungs, and kidneys
* Polyarteritis nodosa
* Kawasaki's syndrome and disease, which primarily affects children and involves the medium vessels of the coronary arteries and eyes, causing rash and febrile illness
* Behcet's disease
* Buerger's disease (Thromboangiitis obliterans), which primarily causes a peripheral vascular obstructive disease in the hands and feet of patients older than 50 years with a history of smoking
* Churg-Strauss syndrome, a form of eosinophilic vasculitis that is usually preceded by severe allergic rhinitis and asthma

* Cryoglobulinemia
* Giant cell arteritis, which primarily occurs in patients younger than 50 years with a recent history of headaches
* Henoch-Schonlein purpura, which often occurs in children following streptococcal infection and causes bruising, arthralgia and abdominal pain.
* Microscopic polyangiitis, which affects small to medium vessels in the skin, lungs, gastrointestinal track and kidneys.
* Polymyalgia rheumatica
* Takayasu's arteritis, which occurs primarily in young Asian women and affects large blood vessels of the aorta and sometimes other internal organs.
* Central nervous system vasculitis

Association with other Disorders

In addition, many other autoimmune conditions such as rheumatoid arthritis, Sjogren's syndrome, and systemic lupus erythematosus (SLE) are accompanied by vasculitis. Vasculitis can also occur in other conditions such as non-Hodgkin's lymphoma, Hodgkin's disease, myelogenous leukemia, adenocarcinoma, and carcinoma of the bronchus.

Causes of Vasculitis

Vasculitis can occur as an inflammatory reaction to many different medications; by direct invasion of vessels by infectious organisms or tumor cells; or as an inflammatory reaction in other systemic autoimmune disorders. Drugs known to cause vasculitis include antibiotics and heart medications. Organisms known to trigger vasculitis include hepatitis viruses, human immunodeficiency virus and parvovirus in panarteritis nodosa; streptococci, salmonella, yersinia, mycoplasma, parainfluenza, rotavirus, and human herpes virus 6 (HHV-6) in Kawasaki's syndrome, and streptococci, staphylococci, yersinia, mycobacteriae, varicella zoster virus, hepatitis viruses B and C, cytomegalovirus and influenza virus in leukocytoclastic vasculitis including Schoenlein-Henoch's purpura.

Drugs that may mimic systemic vasculitis include: cocaine, amphetamines, antibiotics, and ergotamine derivatives. Certain recreational drugs may also trigger autoimmune conditions, including vasculitis.

Symptoms

Symptoms of vasculitis, especially systemic vasculitis, resemble influenza with night sweats, joint pain, weight loss and malaise often occurring. Skin changes in systemic vasculitis include purpura, urticaria, ulceration, livedo reticularis, papules, pustules, nodules, and erythema nodosum.

Cutaneous vasculitis causes a skin rash and sometimes pain and itching.

A characteristic discoloration known as livedo reticularis may also occur. When the small blood vessels of the hands and feet are affected, numbness and tingling may occur. When specific organs such as the kidney are affected, organ-specific symptoms such as flank pain in kidney disease are commonly seen.

Treatment

Vasculitis is usually treated with medications that reduce inflammation such as corticosteroids, cyclophosphamide, and azathioprine. In severe cases, plasma exchange via plasmapheresis or injections of intravenous immunoglobulins may be used.

Resources:

Thomas Bush, Systemic Vasculitis, Diagnostic Clues to this Confusing Array of Diseases, vol 103(2) Postgraduate Medicine, Feb 1998, accessed Feb 26, 2007.

Elaine Moore, Autoimmune Diseases and Their Environmental Triggers, Jefferson, NC: McFarland Publishers, 2002.

Vasculitis, Renal Unit of the Royal Infirmary of Edinburgh, July 2006, accessed Jan 3, 2007.

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