PATIENT FACT SHEET

Vasculitis

CONDITION DESCRIPTION

Vasculitis is a group of rare diseases that cause inflammation of small, medium and large blood vessels. There are many types of vasculitis, such as giant cell arteritis (GCA), Kawasaki disease, microscopic polyangiitis (MPA), and granulomatosis with polyangiitis (GPA). Symptoms, severity and duration vary. It affects all ages and both sexes.

Vasculitis may cause poor blood flow to tissues in the lungs, kidneys, skin or nerves. Vasculitis may be mild, disabling or even deadly. Patients may have one or repeated episodes throughout their life.

Most types of vasculitis have unknown causes. Genetic factors may be important. It may be an autoimmune disease, but some types of vasculitis are reactions to medications, cancer or viral infections like hepatitis B or C.

SIGNS/ SYMPTOMS

Vasculitis has many types, each with different symptoms. Signs of vasculitis include red spots (purpura), lumps (nodules) or sores (ulcers) on the skin; shortness of breath; cough; and numbness or weakness in a hand or foot. Vasculitis may seriously affect kidneys but have no symptoms. Some patients may have pain, fatigue, arthritis, or nose and sinus problems.

A rheumatologist can diagnose vasculitis. Symptoms or abnormal lab tests may suggest vasculitis. Diagnosis may include tissue biopsy, angiography to look for blood vessel abnormalities, and blood tests. A positive test for antineutrophil cytoplasmic antibodies (ANCA) helps detect vasculitis types like GPA, MPA and eosinophilic granulomatosis with polyangiitis (EGPA or Churg-Strauss). Other lab tests may show organ damage but do not help diagnose vasculitis.

Most patients with vasculitis do not have other diseases, but it may occur as a secondary disease for people with rheumatic diseases like systemic lupus erythematosus, rheumatoid arthritis or Sjögren's syndrome.

COMMON TREATMENTS

Glucocorticoids, such as prednisone (Deltasone, Orasone), are used to treat inflammation in many types of vasculitis. Dose and duration depend on disease severity and duration. Glucocorticoids or "steroids" may have serious side effects with long term use. Cyclophosphamide (Cytoxan, Neosar) is an immunosuppressant used for severe disease that threatens organs.

For milder vasculitis, methotrexate (Rheumatrex, Trexall, Otrexup, Rasuvo), azathioprine (Imuran) and other immunosuppressants may be used. Newer biologic drugs can treat certain types of vasculitis, such as rituximab (Rituxan) for GPA, MPA, and cryoglobulinemic vasculitis; tocilizumab (Actemra) for GCA; and mepolizumab (Nucala) for EGPA. Some patients with severe disease receive treatments like plasma exchange (plasmapharesis) or intravenous immunoglobulin (IVIG).

If severe vasculitis damages blood vessels or organs, surgery may be needed to repair them. This may include vascular bypass grafting, sinus surgery or kidney transplant.

CARE/ MANAGEMENT TIPS

Vasculitis may be short term or lifelong. Doctors may focus care on preventing permanent damage to organs like the lungs, kidneys or brain, or the nerves to protect from disability or death.

Patients with vasculitis may have other troubling issues, such as fatigue, pain, arthritis, or sinus problems. Side effects from medications, such as glucocorticoids, may also need careful monitoring and management from a rheumatologist. Patients on immunosuppressants need to take precautionary measures to reduce infection risk. With treatments, the outlook for patients with vasculitis is good.

Depending on their vasculitis type, patients may also need care from an ophthalmologist, dermatologist, otolaryngologist, neurologist, nephrologist or pulmonologist at times.

Updated March 2019 by Kwas Huston, MD, and reviewed by the American College of Rheumatology Committee on Communications and Marketing. This information is provided for general education only. Individuals should consult a qualified health care provider for professional medical advice, diagnosis and treatment of a medical or health condition.

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