Vasculitis Disorders
Frequently Asked Questions

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Q 1: What are vasculitis disorders?

Vasculitis disorders result from inflamed blood vessels. They are rare and can affect people of all ages. Granulomatosis with polyangiitis (also known as Wegener's granulomatosis) is the most common form of vasculitis, and it affects around five in a million people. They cause a wide range of symptoms that can affect the skin and internal organs. Treatment length varies, and some people need to use medications for long periods of time.

Vasculitis disorders result from inflammation of blood vessels. The types of blood vessels affected by vasculitis include arteries, arterioles, veins, venules, and capillaries. Inflammation causes a narrowing of blood vessels, which can result in blood flow obstruction (ischaemia). This may lead to tissue damage (necrosis), and blood clots (thrombosis).

Q 2: What causes vasculitis disorders?

There are three main underlying causes of vasculitis disorders:

- Autoimmunity.
- Allergy or hypersensitivity to medications, toxins, or other inhaled environmental irritants (where removing the medication, toxin or irritant usually stops symptoms).
- Viral or parasite infections.

Allergy, hypersensitivity, and infections should be ruled out before autoimmunity is considered as the cause of a vasculitis disorder.

Q 3: What is autoimmune related vasculitis?

The main role of the immune system is to defend against infections (such as bacteria, moulds and viruses) and other invaders (such as cancer cells), whilst protecting the body's own cells. Autoimmunity occurs when the body does not recognise its own cells and attacks them.

Antibodies produced by the immune system in vasculitis disorders cause inflammation in blood vessels that can lead to problems. Complications depend on which blood vessels, organs and other systems are affected. Vasculitis disorders may also occur in people with other autoimmune diseases such as rheumatoid arthritis, systemic lupus erythematosus (SLE) and dermatomyositis.

Q 4: What are the signs and symptoms of vasculitis disorders?

General signs and symptoms of vasculitis disorders include:

- Fever.
- Loss of appetite and weight loss.
- Fatigue, weakness and lethargy.
- General aches and pains.

Specific signs and symptoms of vasculitis disorders include:

- Skin - purple or red spots or bumps, clusters of small dots, splotches, bruises, urticaria (hives), itch.
- Joints - pain, arthritis in one or more joints.
- Lungs - shortness of breath, coughing up blood, signs that suggest pneumonia.
- Gastrointestinal tract - mouth ulcers (sores), stomach pain and in severe cases blood flow to the intestines can be blocked.
- Sinuses, nose, throat, ears - chronic (ongoing) sinus or middle ear infections, ulcers in the nose, and hearing loss.
- Eyes - red, itchy and burning, increased sensitivity to light, blurred vision.
- Brain - headaches, changes in mental function, stroke-like symptoms such as muscle weakness and paralysis.
- Nerves - numbness, tingling, and weakness in various body parts, loss of feeling or strength in hands and feet, shooting pains in arms and legs.

Q 5: How are vasculitis disorders diagnosed?

Due to the wide range of signs, symptoms and body systems involved, an extensive clinical history and physical examination is needed to diagnose vasculitis disorders. Blood tests are taken, and in some cases an x-ray or biopsy may be required. An exact diagnosis is needed to provide the right treatment. Referral to a clinical immunology/allergy specialist or other medical specialist is usually required.

Q 6: How are vasculitis disorders treated?

When vasculitis is due to an autoimmune disorder, immunosuppressive drugs are usually used.
In severe cases that do not respond to other treatment, plasmapheresis may be used, which filters the offending autoantibodies out of the blood plasma and returns the filtered blood back to the patient.

Q 7: What are examples of vasculitis disorders affecting small blood vessels?

- Granulomatosis with polyangiitis affects sinuses, lungs, kidneys, and skin.
- Eosinophilic granulomatosis with polyangiitis affects the lungs, skin, and nerves.
- Cryoglobulinaemia affects skin, kidneys, and nerves.
- Goodpasture's syndrome affects lungs and kidneys.
- Henoch-Schonlein purpura affects skin, joints, kidneys, and gut.
- Microscopic polyangitis affects skin, kidneys, and nerves.

Q 8: What are examples of vasculitis disorders affecting medium blood vessels?

- Behcet's disease affects mucous membranes, skin, and eyes.
- Central nervous system vasculitis affects the brain.
- Kawasaki syndrome affects skin, mucous membranes, lymph nodes, and blood vessels.
- Polyarteritis nodosa affects arteries, kidneys, gut, nerves, and skin.

Q 9: What are examples of vasculitis disorders affecting large blood vessels?

- Giant cell (temporal) arteritis affects arteries of the head and neck.
- Takayasu arteritis affects arteries of the head and neck.
- Polymyalgia rheumatic produces inflammation and swelling in joints and muscles.

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