

What is Central Nervous System Vasculitis?

Central nervous system (CNS) vasculitis is a rare condition that affects the central nervous system, which includes the brain and spinal cord. Vasculitis causes inflammation of the blood vessels. This restricts blood flow and can damage the brain and spinal cord, and can cause loss of brain function, stroke and even life-threatening conditions.

Vasculitis is classified as an autoimmune disorder, which occurs when the body's natural defense system mistakenly attacks healthy tissues.

CNS vasculitis is typically categorized as *primary* and *secondary*:

- Primary angiitis of the CNS (PACNS) is vasculitis confined specifically to the brain and spinal cord. It is not associated with any other systemic disease (affecting the whole body).
- Secondary CNS vasculitis usually occurs in the presence of other autoimmune diseases such as systemic lupus erythematosus or Sjögren's syndrome; and systemic forms of vasculitis, such as granulomatosis with polyangiitis, microscopic polyangiitis, or Behçet's syndrome or viral or bacterial infections.

Causes

The cause of CNS vasculitis is not fully understood. It is believed that several factors may cause CNS vasculitis:

- An infection may contribute to the onset of CNS vasculitis.
- Environmental and genetic factors may play a role as well.

Who Gets Vasculitis of the CNS?

In general, CNS vasculitis is considered rare. In the case of PACNS, the disorder can affect people of all ages but generally peaks around age 50. It most often occurs in males.

Symptoms

Many forms of vasculitis are accompanied by fever, fatigue, and unintentional weight loss. CNS vasculitis symptoms may also include:

- Severe headaches that don't go away

- Strokes or transient ischemic attacks (mini-strokes)
- Swelling of the brain (encephalopathy)
- Forgetfulness or confusion
- Muscle weakness or paralysis
- Difficulty with coordination
- Abnormal or loss of sensations
- Vision problems
- Seizures or convulsions

Diagnosis

Diagnosing CNS vasculitis is a challenge. It involves ruling out other conditions that have similar symptoms like other diseases and infections. There is no single diagnostic test for CNS vasculitis, so your doctor will consider a number of factors, including:

- **Lab work:** Blood tests are frequently normal in PACNS vasculitis, but may be abnormal in other diseases.
- **Examination of the spinal fluid:** A sample of the cerebrospinal fluid (surrounding the brain and spinal cord) is tested for infection and signs of inflammation. It is removed by inserting a needle in the lower section of the back, and is called a spinal tap.
- **Diagnostic imaging:** Computed tomography (CT) scans and magnetic resonance imaging (MRI) can help identify abnormalities of the brain, spinal cord, blood vessels, and other organs and tissues.
- **Cerebral angiogram:** An angiogram dye (contrast agent) is injected into the blood vessels. X-rays are taken that can detect narrowing and blockages of blood vessels in the brain.
- **Biopsy:** This surgical procedure removes a small tissue sample from a blood vessel or an affected organ. It is examined under a microscope for signs of inflammation or tissue damage. Because other conditions can cause brain abnormalities that are similar to CNS vasculitis, a brain biopsy may be necessary in trying to make a more definitive diagnosis.

Reversible Cerebral Vasoconstriction Syndrome (RCVS)

RCVS is a group of conditions that involve spasm of the blood vessels in the brain. The symptoms can be the same as seen in CNS vasculitis. RCVS causes sudden, severe headaches, as well as strokes or bleeding into the brain. It is important for your physicians to be aware of RCVS in order to tell if the symptoms are from CNS vasculitis or RCVS. Treatment and prognosis are different.

Treatment

The goal of treatment is to reduce the inflammation. This is done by using a high-dose corticosteroid such as prednisone. For more severe cases, prednisone is used in combination

with drugs that suppress the immune system's response, such as cyclophosphamide, mycophenolate mofetil or azathioprine.

Treatment may be more aggressive for the first three to six months and then tapered down as symptoms improve. In addition to medications, other forms of treatment may include physical, occupational and speech therapy. If memory is affected, brain activities that enhance memory may be recommended.

Side Effects of Treatment

The medications used to treat CNS vasculitis have potentially serious side effects, which include:

- Lowering your body's ability to fight infection
- Potential bone loss (osteoporosis), among others

Therefore, it is important to see your doctor for regular checkups. Medications may be prescribed to lessen these side effects.

Infection prevention is also important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

Relapse

Even with effective treatment, relapses are common for individuals with CNS vasculitis. If your initial symptoms return or you develop new ones, report them to your doctor as soon as possible.

Regular check-ups and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Your Medical Team

Effective treatment of CNS vasculitis may require the coordinated efforts and ongoing care of a team of medical providers and specialists.

In addition to a primary care provider, you may need to see the following specialists:

- Rheumatologist (joints, muscles and immune system)
- Neurologist (brain/nervous system)
- Physical, occupational or speech therapist
- Others as needed

The best way to manage your disease is to actively partner with your health care providers and get to know the members of your health care team.

It may be helpful to keep a health care journal to track medications, symptoms, test results and notes from doctor appointments in one place.

To get the most out of your doctor visits, make a list of questions beforehand and bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it is up to you to be your own advocate. If you have concerns with your treatment plan, speak up. Your doctor may be able to adjust your dosage or offer different treatment options. It is always your right to seek a second opinion.

Living with CNS Vasculitis

Living with a chronic disease such as CNS vasculitis can be overwhelming at times. Fatigue, pain, emotional stress and medication side effects can take a toll on your sense of well-being.

This can affect your relationships, work and other aspects of your daily life. Sharing your experience with family and friends, connecting with others through a support group, or talking with a mental health professional can help.

Outlook

There is no cure for CNS vasculitis at this time. However, it is treatable. Early diagnosis and treatment are essential to prevent loss of brain function or stroke.

Even with treatment, relapses are common with CNS vasculitis, so follow-up medical care is essential.

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