
Central Nervous System Vasculitis Factsheet

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CNS Vasculitis is caused by inflammation of the blood vessels supplying the brain and spinal cord. Several different types of vasculitis which affect other areas of the body can cause Central Nervous System (CNS) Vasculitis. These include the ANCA-associated vasculitides, Takayasu Arteritis and Giant Cell Arteritis. CNS vasculitis can also occur without any associated cause. This is called Primary Angiitis of the CNS (PACNS). The symptoms of CNS Vasculitis can include confusion, problems with eyesight, headache and personality change. Sometimes it is mistaken for a stroke. Steroids and medicines to suppress the action of the immune system such as cyclophosphamide are usually given to treat the condition.

Who gets CNS vasculitis?

Men over the age of 50 are most commonly affected. PACNS is a rare condition. At any one time there are about 2 in 10,000 people in Europe suffering from CNS vasculitis. Only about 700 cases have been reported in the medical literature worldwide.

What causes CNS vasculitis??

CNS vasculitis is caused by inflammation of the blood vessels of the brain and spinal cord. Small, medium or large blood vessels can be affected. It is thought that the inflammation is caused by auto-immunity, a process in which the person's immune system attacks their own body tissues. It can sometimes be triggered by an infection with a germ or a virus.

What are the symptoms of CNS vasculitis?

You may notice severe long-lasting headaches, memory loss or confusion, weakness, numbness, eyesight problems or dizziness. Your friends or family may notice changes in your personality. Seizures occur and in rare cases, coma. Symptoms such as speech problems, numbness and paralysis of the arms or legs can lead to the condition being confused with a stroke.

How is CNS vasculitis diagnosed?

CNS vasculitis is an extremely difficult condition to diagnose. Your doctor may suspect the disorder from your symptoms and the findings of a physical examination. Blood tests may reveal anaemia, a high white blood cell count (these cells are often raised when there is inflammation in the body), and kidney or liver problems. Tests for over activity of your immune system may show positive. Inflammatory markers (chemicals in the blood which are produced in excess when inflammation occurs) may be found at high levels. X-rays and biopsies (samples of tissue removed for examination under the microscope) are sometimes required. You may need a magnetic resonance imaging (MRI) scan, lumbar puncture or an angiogram which involves injecting dye to show up the blood vessels of the brain.

What is the treatment for CNS vasculitis?

You may need a steroid medicine such as prednisolone to control the inflammation. This may need to be taken in a high dose to start with. It is either given alone or, commonly, in combination with cyclophosphamide, a medicine which helps to suppress the activity of the immune system. Treatment needs to be given for a long time and in some people for life.

Does CNS vasculitis get better?

The course of CNS vasculitis is very variable. Many people find their symptoms come and go over time but are generally controlled providing they take their medicine. CNS vasculitis can be a serious disease. It can cause brain function to deteriorate over time and can on occasion be life-threatening.

Further reading

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