Cryoglobulinemia and Cryoglobulinaemic Vasculitis

What is Cryoglobulinemia?

Cryoglobulinemia means "cold antibody in the blood" and is the presence of abnormal antibodies that are soluble in the blood at body temperature but which precipitate out of the blood at lower temperatures in the laboratory. These antibodies are often present in patients with a wide variety of pre-existing diseases such as hepatitis C virus infection, autoimmune diseases such as rheumatoid arthritis or Sjögren's Syndrome or cancers such as lymphoma or multiple myeloma. In the very rare cases where an underlying disease is not identified the presence of cryoglobulin antibodies in the blood is called "essential cryoglobulinaemia". Often the antibodies do not cause any problems and then no treatment may be needed. In some patients they cause a vasculitis of the small and medium blood vessels called Cryoglobulinaemic Vasculitis. This condition may need treatment.

What are the symptoms?

Cryoglobulinaemic Vasculitis symptoms include rash on the lower limbs, joint pain or arthritis, nerve damage, abdominal pain or kidney failure.

What is the aetiology (cause)?

The commonest underlying causes of cryoglobulinaemia are hepatitis C infection, lymphoma and myeloma. The cause of essential cryoglobulinaemia is not known and can only be diagnosed once all the possible underlying causes have been excluded.

Making a diagnosis

Diagnosis is made by recognising the clinical symptoms and signs and performing the specific laboratory test for cryoglobulins. A biopsy of the affected tissue or organ may be necessary to confirm the diagnosis.

Treatment

The most important treatment is of the underlying disease and will depend on the correct diagnosis. Patients with severe forms of vasculitis may need additional treatment with steroids, immunosuppressant drugs or plasma exchange to control the vasculitis.
Drugs and Side effects
For information on the main drugs prescribed for Cryoglobulinemia and Cryoglobulinaemic Vasculitis see:

- Steroids
For information on other drugs used in the treatment of vasculitis see Glossary of drugs and side effects.

Treatment - Plasma exchange or plasmapheresis
This treatment is sometimes used in patients with severe vasculitis where antibodies in the blood are thought to be important in causing the disease. The treatment involves removing antibodies from the blood using a machine and returning the "cleaned" blood back to the patient. The treatment may necessitate giving blood products to the patient including plasma, albumin or immunoglobulin. It may also involve giving drugs to thin the blood and prevent it clotting in the machine.

For information on plasma exchange:

- Plasma exchange or plasmapheresis

Prognosis
Prognosis is determined by the underlying disease as well as other factors including age, and the severity of damage to any organs, especially the kidneys.

Key Points

- Cryoglobulins and cryoglobulinaemic vasculitis are often caused by underlying diseases including infections and cancers
- Treatment depends on the underlying disease and the severity of the vasculitis

Related Vasculitis Articles

- Fertility and Vasculitis - Dr David Jayne

Further reading

- Mixed Cryoglobulinemia - Clodoveo Ferri
- Cryoglobulinaemic vasculitis: classification and clinical and therapeutic aspects - Gerald S Braun, Sophia Horster et al