

URTICARIAL VASCULITIS

WHAT IS URTICARIAL VASCULITIS?

Urticarial Vasculitis is a form of cutaneous vasculitis characterised by inflammation of the small blood vessels. It can be classified into three subtypes. All are defined by a measure of the "complement" levels in the blood. The complement system is a set of proteins that contribute to and amplify immune responses. They play a role in some, but not all, autoimmune disorders including some forms of Urticarial Vasculitis.

TYPE OF URTICARIAL VASCULITIS

Normocomplementemic Urticarial Vasculitis (NUV).

NUV is diagnosed where a patient has the main symptoms of Urticarial Vasculitis combined with normal levels of C1q complements. NUV is generally the least severe form of Urticarial Vasculitis. It is less likely to be associated with any other symptoms.

Hypocomplementemic Urticarial Vasculitis (HUV)

HUV is diagnosed where the patient has the main symptoms of Urticarial Vasculitis combined with lower than normal levels of C1q complements and raised levels of anti-C1q antibodies (antibodies that attack the C1q complements). HUV is a more severe form of Urticarial Vasculitis and is likely to include symptoms such as purple or dark red spots or rash on the skin (a typical vasculitic rash); arthritic joint pain breathing difficulties such as asthma, and stomach pains.

Hypocomplementemic Urticarial Vasculitis Syndrome (HUVS)

HUVS is a very rare and severe form of Urticarial Vasculitis. HUVS patients have more extensive complement abnormalities (low circulating 3rd and 4th complement components). As well as the symptoms of HUV, patients will suffer from systemic problems such as: episcleritis or uveitis (bloodshot or inflamed eyes) ; pleuritis (inflammation of the membrane surrounding the lungs); angioedema (swelling of the tissues under the skin (breathing difficulties) and cardiac involvement such as myocardial infarction (heart attack).

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CAUSE

The cause of Urticarial Vasculitis is not known in 50 per cent of cases. However it can be associated with or triggered by autoimmune/connective tissue diseases like Systemic Lupus Erythematosus (SLE or simply Lupus) Rheumatoid Arthritis, or occur in the context of a systemic vasculitis syndrome, such as Churg Strauss syndrome (Eosinophilic Granulomatosis with Polyangiitis); infections or viruses such as hepatitis; a reaction to certain drugs such as ACE inhibitors; certain types of diuretics; penicillin and Non-Steroidal Anti-Inflammatory Drugs (NSAIDs); cancer; and Glandular problems, such as Graves' Disease (overactive thyroid).

SYMPTOMS

The main symptom is a recurring urticarial ("stinging nettle") rash that lasts for longer than 24 hours and "burns" rather than "itches", leaving marks behind on the skin when it clears.

TREATMENT

Milder cases of Urticarial Vasculitis may simply be treated with antihistamines and NSAIDs such as Ibuprofen. Corticosteroids such as Prednisone might be used for more persistent cases. However for the more severe forms of Urticarial Vasculitis, steroids such as Prednisone are usually prescribed to reduce inflammation as well as immunosuppressants such as Azathioprine, Cyclophosphamide or Mycophenolate Mofetil (CellCept). In cases where a patient is unresponsive to treatment, intravenous immunoglobulin and anti-cytokine monoclonal antibodies or rituximab may have a role. If the disease is very severe large doses of Methylprednisolone or Plasmapheresis (plasma exchange) may also be given. When the disease becomes quiet less toxic drugs are used to keep control and these include: Azathioprine, Methotrexate and Mycophenolate Mofetil usually in combination with low dose prednisone