Woodlands Health
National Healthcare Group

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Adult Onset Still's Disease

What is adult onset Still's Disease?

Adult onset Still's Disease is a rare inflammatory condition in which patients experience daily high fevers, rash, swollen and achy joints. In some patients, the disease can progress and lead to long term arthritis. It may also involve other organs like the liver, blood system, heart and lungs.

What causes it?

The cause of adult onset Still's Disease is not known. It has been hypothesised that the condition may be triggered by a viral or bacterial infection. The resultant over-activity in the body's immune cells causes inflammation and damage which often does not abate unless treated.

What are the symptoms and signs?

Most patients will experience a combination of the following symptoms and signs:

- High spiking fever of at least 39°C daily for a week or longer. The fever usually peaks in the late afternoon or early evening.

- A salmon pink rash may come and go with the fever. The rash usually appears over the trunk, arms or legs. The rash is usually not itchy.

- Joint swelling, warmth and pain may occur in the hands, wrist, elbows, shoulders, knees and ankles. Usually, several joints are affected at the same time. There can be associated morning stiffness in the joints that lasts for several hours.

Other signs and symptoms may include:

- Sore throat
- Swollen lymph glands in the neck
- Abdominal pain and swelling (enlarged liver or spleen)
- Chest pain on deep breathing (inflammation in the lining of the heart and lungs)

What are some complications?

- Long term inflammation can damage joints leading to loss of range of movement, deformities and in severe cases, may require surgical joint replacement.

- Liver disease can occur as a result of liver inflammation.

- Inflammation of the heart can occur in the cells lining the heart (pericarditis) or in the heart muscle (myocarditis).

- Inflammation in the lining of the lungs may cause fluid buildup in the pleural cavity, a fluid filled space that surrounds the lungs.

- Inflammation in the bone marrow may cause low platelet count, low white cell count and anaemia.

How is it diagnosed?

There is no single test that can be used to diagnose the disease. Very often, many medical tests would have been performed before a final diagnosis is made.

This is because other diseases that may present similarly (e.g. blood disorders like leukaemia and lymphoma, infectious mononucleosis, autoimmune diseases like rheumatoid arthritis and systemic lupus erythematosus) often need to be first excluded.

The following blood tests can be helpful in diagnosis:

- Full blood count (FBC) may show a high white blood cell count, reduced red blood cells (anaemia), or abnormal numbers of platelets (high or low).
- Erythrocyte Sedimentation Rate (ESR), a measure of inflammation, will be high.
- C Reactive Protein (CRP), a measure of inflammation, will be high.
- Serum ferritin level will be very high.
- Liver function tests may show high levels of ALT and AST, liver enzymes that are released when the liver is inflamed.
- Antibody tests for other autoimmune diseases such as the Anti Nuclear Antibody and Rheumatoid Factor are usually negative.

The following imaging tests may be performed:

- X-rays of affected joints
- X-ray of the chest, electrocardiogram, and echocardiogram when inflammation of the lungs and heart is suspected.
- CT scan or ultrasound of the abdomen when enlargement of the liver and spleen is suspected.
- Another test that may sometimes be performed is a bone marrow examination. This may be necessary to help rule out other blood cell disorders like leukaemias and lymphomas which may present with similar symptoms and signs.

How is it treated?

A variety of drugs can be used to treat the disease. Each treatment regime is individualised to the patient, depending on the severity of symptoms, organs involved and the likelihood of developing drug side effects.

Non-steroidal anti-inflammatory drugs (NSAIDs) are usually used to reduce inflammation and can be the initial treatment choice in patients with mild arthritis. Patients on regular NSAIDs will require regular blood tests to monitor blood counts, kidney and liver function.

Steroids may be necessary when one suffers from high fever spikes, severe joint symptoms, or inflammation affecting the internal organs. Steroids work by suppressing the immune response which is overactive in adult onset Still's disease. Intravenous steroids may be utilised in life threatening situations or when rapid control of the disease is necessary.

Disease modifying anti rheumatic drugs (DMARDs) may be necessary when the use of steroid alone is insufficient to control the disease. When used in combination, patients require less steroids, thus reducing their risks for developing osteoporosis and other complications. Each individual DMARD has its own potential side effects for which regular blood tests for monitoring will be needed.

In cases where traditional DMARDs are not effective, biologic agents may be considered. These drugs, such as tocilizumab or anakinra, target specific pathways in the immune system to reduce inflammation.

Contributed by Nursing