

Resource

Felty's syndrome

Felty's syndrome is a very rare complication of RA, in which those affected have low white blood cell count, infections and leg ulcers. It more commonly occurs in patients who have severe RA.

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Rheumatoid arthritis (RA) is undoubtedly a disease of the joints. Hence, the word “arthritis” (which means ‘inflammation of the joint’) in its name, but there are variants that have severe manifestations outside of the joints. Thus, patients with active RA may have narrowing of the arteries due to atheroma (a fatty deposit that can build up inside arteries) with an increased risk of heart attacks and strokes, which occur if the atheroma grows enough to block the flow of blood. Another recognised but

very rare extra-articular feature of RA is Felty's Syndrome (FS). About 1-3% of patients with RA may develop Felty's. However, this incidence rate may be declining. ?

What is Felty's Syndrome?

Felty's syndrome is a rare complication of RA, first described in 1924. Its features are low neutrophils (white blood cells) in the blood, infections and leg ulcers in a patient who usually has severe RA.

Features of Felty's Syndrome

1. Rheumatoid arthritis
2. Low white blood cells
3. Large spleen
4. Often serious and repeated infections

How is it diagnosed?

The RA is usually severe. Examination of the blood of patients with Felty's syndrome shows a very low white blood cell count due to a reduction of specialised white blood cells called neutrophils. Neutrophils are vital for defending us from bacterial infections. Hence a significant proportion of patients with Felty's syndrome have recurrent infections of the lungs, the urinary tract and even blood infection (septicaemia). There is also enlargement of the spleen which can be detected either clinically or by ultrasound.

What is the cause of Felty's syndrome?

This is not known. The neutrophils accumulate in and are destroyed by the spleen. ?

Why me?

All the factors that go into producing Felty's syndrome are not known, so it is not possible to diagnose FS in advance. ?

Why do only some patients develop infections?

There is, at best, only a rough relationship between the low neutrophils and infection. ? Two patients can have the same very low numbers of circulating neutrophils, and one may have infections, and the other may not. Conversely, patients with higher numbers of neutrophils may still have infections.

Treatment

The treatment is satisfactory in the vast majority of patients.

1. Adequate treatment with disease-modifying drugs, especially methotrexate, may improve the number of circulating neutrophils and reduce infections. However, in some patients, if the methotrexate has a toxic effect on the bone marrow, the neutrophils may fall, which can make monitoring difficult, as it is hard to determine whether this fall is caused by the drug treatment or the condition.

2. Biologics may be used when there has been no response to drugs. A low neutrophil count on its own does not warrant therapy. If there are infections, then treatment is indicated. Usually, DMARDs, particularly methotrexate, are tried. In some patients but not, all the neutrophil count may increase. If the low neutrophil count is associated with severe infections, then rituximab has been successfully used to correct the low neutrophils. No clinical trials have been undertaken so this is an experimental, off-label use of this biologic. In some cases, drugs to stimulate white cell production may be used (granulocyte colony-stimulating factor).
3. In extreme cases, removal of the spleen (splenectomy) may be carried out.

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