

Resource

# Rheumatoid vasculitis

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## Introduction

The word 'vasculitis' means that blood vessels are inflamed, just as appendicitis indicates the appendix is inflamed and arthritis that the joints are inflamed. The consequences of vasculitis depend on the size, site and number of blood vessels involved. When small or medium-sized arteries are involved, they can become blocked, and this can result in infarction (death) of the tissue the blood

vessel supplies. If, for example, a coronary artery in the heart is involved (fortunately rare) then this may result in a heart attack and, potentially, death. When very small blood vessels such as capillaries are involved this is rarely serious except when lots of blood vessels close by each other are involved, and there is associated inflammation, such as can occur in the kidney, resulting in glomerulonephritis (a type of kidney disease). Arteries can also cause problems if part of the wall is involved. Under those circumstances, because the pressure inside the artery is high, the wall can become weak due to inflammation, resulting in the formation of a blood-filled sac known as an 'aneurysm' which can potentially rupture with severe haemorrhage (bleeding).?

## Classification of Vasculitis

Vasculitis can occur as a primary event (out of the blue) in diseases such as polyarteritis nodosa, GPA – (Granulomatosis with PolyArteritis, previously known as Wegener's granulomatosis) etc., but also secondary to (or after) a number of infections, malignancies and connective tissue diseases. The best described of these is vasculitis occurring as a complication of rheumatoid arthritis (see below).?

Vasculitis can also be classified according to the size of the blood vessel involved. In patients with rheumatoid arthritis, aortitis (inflammation of the aorta, the largest artery in the body, which is connected to the heart) can occur (rarely), leading particularly to a leaking aortic valve (aortic incompetence). Very occasionally patients have a vasculitis involving medium-sized arteries (as seen in polyarteritis nodosa) with potentially serious infarction and haemorrhage.?

The most common type of vasculitis in rheumatoid arthritis is a small vessel vasculitis which can also involve small arteries and arterioles (small branches of arteries). When very small blood vessels only are involved, this usually affects the nail edges and nail folds, so-called nail fold vasculitis, which occurs in patients with severe arthritis but in itself is not serious. When a small artery is involved, this is usually associated with a systemic illness (weight loss, fever, etc., called systemic rheumatoid vasculitis) which more often has serious consequences.?

## Systemic Rheumatoid Vasculitis

Systemic vasculitis (i.e. widespread vasculitis causing symptoms of a general illness) complicating rheumatoid arthritis appears to be on the decline. This is probably as a consequence of modern and better treatments of underlying arthritis. There appeared to be an association between severe systemic vasculitis complicating rheumatoid arthritis and the uncontrolled (excessive) use of steroids in the 1950s and 1960s, but we still see patients with systemic vasculitis in the absence of steroid treatment. There is no evidence that the low doses of steroids currently used increase the risk of developing vasculitis. Data from Norwich suggests that systemic vasculitis now only affects around 3 patients per million of the population per year.?

Studies from the 1970s and 1980s have shown that this type of vasculitis is associated with a poor outcome and a high risk of early death in the absence of effective treatment. More recent studies in the 2000s have shown that although the frequency of this disease has declined, the clinical presentation has not changed, and the outcome is still poor despite aggressive treatment. Typical clinical features include weight loss, fever, numbness or weakness due to damaged nerves and leg ulcers, but it is important to recognise that leg ulcers occur in some patients with chronic arthritis in the absence of vasculitis.?

Vasculitis is also associated with most of the extra-articular (meaning 'outside of the joints') manifestations described in rheumatoid arthritis. These include inflammation of the eyes (iritis), inflammation of the lining of the heart and lung (pericarditis and pleurisy) and other lung and heart manifestations including inflammation of the bases of the lung (fibrosing alveolitis) and irregular heartbeat, including heart block when the heart beats very slowly.?

Neuropathy can also occur and describes damage to peripheral nerves which could just mean numbness (as mentioned previously) but could also be a condition called mononeuritis multiplex, where specific nerves are damaged due to lack of blood supply, which can present with symptoms including foot drop and wrist drop (i.e. difficulty lifting the foot or wrist). Vasculitis also occurs more frequently in patients who have Felty's syndrome (a low white cell count, a large spleen and rheumatoid arthritis) and is more common in patients who have nodules within the skin (intra-cutaneous nodules) in the skin of the hands as well as nodules under the skin (subcutaneous nodules) elsewhere, such as over the elbows.?

There are no diagnostic laboratory tests for systemic vasculitis, but patients usually have high levels of rheumatoid factor in their blood, frequently have subcutaneous nodules, and the systemic vasculitis is also frequently accompanied by small, brown spots around the nails (commonly referred to as nail fold infarcts), indicating the combination of both small and larger sized blood vessel involvement.?

## Sub-Clinical Vasculitis

Vasculitis is thought to be one of the major processes involved in rheumatoid arthritis. These comprise of:?

1. Serositis: inflammation of lining surfaces, including that of the joints (arthritis), the tendon sheaths (tendonitis), but also the lining of the heart and lung (pericarditis and pleurisy).
2. Nodules are a discreet process which is seen under the skin but under the microscope show characteristic features. Although these mainly occur on areas of the body that are subject to repeated knocks and under the skin, they can occasionally occur internally such as in the lung when they can look, to all intents and purposes, like cancer (though are not cancerous).?
3. Finally, the third process is vasculitis. Vasculitis can occur without displaying characteristic clinical symptoms (known as 'sub-clinical' vasculitis) It has been found in some studies that very minor inflammatory changes around blood vessels (as seen in sub-clinical vasculitis) are quite common, and the link between this and the more systemic vasculitis described in this article is not fully understood.

It is important also to recognise that subclinical inflammation in the walls of larger blood vessels is thought to be an important process in the development of atheroma/atherosclerosis (hardening of the arteries) and it is important when understanding this process that the type of vasculitis I have described in this article is a very extreme form and fortunately quite rare.?

## Treatment

Treatment of systemic rheumatoid vasculitis is with immunosuppressive drugs, particularly

cyclophosphamide accompanied by corticosteroids. Cyclophosphamide was originally given orally but, because of concerns regarding bladder toxicity (damage), more recent studies support the use of cyclophosphamide by intravenous infusion, along with corticosteroids.?

Once remission has been achieved, usually, within 3-6 months, patients may be switched to alternatives such as methotrexate or azathioprine.?

In resistant cases,? plasma exchange (where blood is removed, the plasma separated from the red cells and then the red cells given back) or immunoglobulin given by infusions can also be effective. Newer biologics have been tried with varied success, but there may be a role particularly for drugs which deplete B cells such as rituximab.?

## Conclusion

Systemic vasculitis is a very rare but serious complication of rheumatoid arthritis and may be considered one of the most serious extra-articular consequences of this disease. Early recognition and treatment with immunosuppressive drugs are usually effective.?

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