

8. Behçet's Disease and Blood Vessels

How are blood vessels affected by Behçet's disease?

One important feature of Behçet's disease is inflammation in the walls of blood vessels (vasculitis). The underlying cause is not known. Behçet's disease is unusual in that it can cause inflammation in both arteries and veins, and can involve vessels of any size.

Inflammation in a vessel means that many white blood cells, designed to fight infection, accumulate within the thickness of the vessel wall. This can lead to several outcomes:

- The inner lining of the vessel becomes more 'sticky', leading to a risk of a blood clot forming within the vessel and blocking it (thrombosis).
- Sometimes, the vessel wall slowly becomes thicker as the cells accumulate and the inside of the vessel – the lumen – becomes narrower so less blood can flow through it (stenosis).
- The wall may also become weaker, as the white blood cells can disrupt the small layer of muscle that keeps the strength of the vessel wall. This can result in expansion of the vessel (aneurysm).
- In rare cases, the vessel wall of an aneurysm can tear (rupture), causing internal bleeding.

Complications

Inflammation in veins

About 25% of patients with Behçet's disease experience inflammation in a vein in the leg at some time. This is usually caused by inflammation of a superficial vein (that is, a vein near the skin surface), causing painful thickening of vessels visible and touchable just under the skin (superficial thrombophlebitis). In about 10% of patients, a blood clot forms in one of the deep veins of the leg (deep vein thrombosis).

Very occasionally, larger central veins in the abdomen and chest become inflamed and occlude. This may manifest as swelling in the head and neck (superior vena cava obstruction) or legs (inferior vena cava obstruction).

Inflammation in small veins can cause some of the unusual skin problems that are seen in Behçet's disease, including skin ulcers (vasculitic ulcers) and tender nodules under the skin (erythema nodosum).

Involvement of arteries is much less common than that of veins, but when it occurs, it may be more serious. Involvement of arteries in the arms or legs can lead to ballooning (aneurysm) or occlusion (arterial thrombosis). Occlusion of a femoral artery that supplies a leg can cause a pale, painful leg. The pulmonary arteries, which take blood from the heart to the lungs, can be affected by the vascular inflammation. This may cause shortness of breath on exertion and sometimes causes a cough with small amounts of blood (haemoptysis).

Investigations

Doppler ultrasound is used to detect clots in the deep veins of the legs or other veins. Investigation of abnormalities in arteries usually involves MRI or CT scanning after intravenous injection of a contrast agent (MR/CT angiography).

Treatment

The treatment of vascular inflammation usually involves steroids and immunosuppressive drugs. The intensity of the treatment is tailored to the severity of the problem. If a blood clot has formed, this may be treated with an anticoagulant such as warfarin. Whether an anticoagulant is indicated and if so for how long anticoagulation needs to be continued varies between individuals.