Behçet's Disease

What is Behçet's Disease?

Behçet’s Disease (BD) is very rare in the UK. It is a vasculitis that can affect both large and small vessels including both arteries and veins.

Who are affected?

Behçet’s is rarely found in the UK. The disease can affect people of any age although it is commonest in the 20s-30s. Men and women are probably equally affected.

What is the aetiology (cause)?

The cause of Behçet’s is not yet known.

What are the symptoms?

Almost any organ can be affected. The commonest symptoms are recurrent crops of painful ulcers in and around the mouth and lips and the genitals. The joints may be affected with a painful arthritis. The most serious problems include inflammation in the front of the eye (anterior uveitis) which can result in cataracts and glaucoma and inflammation at the back of the eye (posterior uveitis) which can lead to blindness. Inflammation in the brain and nervous system may also occur. Inflammation may also occur in the aorta and major arteries leading to aneurysms and clots (thrombosis). Clots may also form in the veins of the skin (superficial phlebitis) or major veins (deep vein thrombosis).

Making a diagnosis

There are no specific tests for BD. As with other forms of vasculitis making the diagnosis depends on recognising the pattern of symptoms and findings on examination. Blood tests may show evidence of inflammation.

One test that is partly specific for BD is the "Pathergy test". This test involves using a sterile hypodermic needle to make a deep prick in the forearm skin of the patient. A positive test is where a small swelling (papule) possibly with pus (pustule) is present at the site of the needle prick 48 hours later. If positive, the test is useful but a significant proportion of patients with Behçet’s disease will have a negative test.

In cases where the brain is involved specialist scans such as magnetic resonance imaging scans (MRI) may provide useful information.
Treatment

High dose steroids possibly with cyclophosphamide are often used in serious cases of Behçet's disease such as those involving the brain or eyes. For less serious cases milder treatment with drugs such as colchicine, non-steroid anti-inflammatory drugs (eg ibuprofen), azathioprine, methotrexate or ciclosporin A may be used.

Drugs and Side effects

For information on the main drugs prescribed for Behçet's Disease see:

- Azathioprine
- Ciclosporin
- Colchicine
- Methotrexate
- NSAIDs

For information on other drugs used in the treatment of vasculitis see Glossary of drugs and side effects.

Prognosis

The overall prognosis is very variable and depends on the severity of the disease and the frequency of relapses. Most patients will require ongoing treatment to control the symptoms of the disease and disease relapses.

Key Points

- Behçet’s disease is a rare form of vasculitis
- It commonly causes ulcers on the mouth, lips and genitals
- May also cause inflammation in the eyes, brain and other organs
- Treatment depends on disease severity.

Related Vasculitis Articles

- Fertility and Vasculitis - Dr David Jayne

Further reading

- Behçet's Disease (Adamantiades-Behçet’s Disease) - Fumio Kaneko, Ari Togashi, Sanae Saito, et al
- Alemtuzumab (CAMPATH-1H) as Remission Induction Therapy in Behçet’s Disease

Video:

Behçet's Disease explained This video explains about Behçet's and patients speaking about their own journey with Behçet's

http://www.youtube.com/watch?feature=player_embedded&v=Txf7cWW8HwI

Behçet's - Sanya Richards – athlete

http://www.youtube.com/watch?feature=player_embedded&v=x2dIA_lr_ZQ
Behçet's - Centres of Excellence

In February 2012 three UK Behçet's Centres of Excellence, for the diagnosis and treatment of Behçet's Disease, were announced. These are to be at Bart's and the London Hospital, Birmingham City Hospital and at Aintree University Hospital in Liverpool. The centres will be lead by Prof Farida Fortune (London), Dr Deva Situnayake (Birmingham) and Prof Robert Moots (Liverpool). Further information from Chris Phillips (+44(0)8451307328. http://www.behcets.org.uk

Useful links

Our Useful Vasculitis Links page contains contact details for organisations offering help and support for patients with Behçet's Disease and other vasculitis diseases.