

15. Headache in Behçet's Disease

Headache is one of the most continual and troublesome symptoms in Behçet's disease. Some patients are lucky and hardly ever get headaches; others have daily chronic severe throbbing headaches that are very disabling.

What causes headache?

There are two kinds of headaches? primary and secondary. In the secondary type, the headache is adjudged to be due solely to a process of disease within the brain. Headache often accompanies the neurological complications that may occur in Behçet's disease, of which there are three main types: inflammation within the brain (meningoencephalitis), inflammation of the lining of the brain (meningitis) and a disturbance of blood drainage from the brain leading to an increased pressure in the brain (intracranial venous sinus thrombosis).

Inflammation within the brain leading to meningoencephalitis is rare, affecting only 5% of patients, but can be very severe. Characteristically, an escalating, often throbbing headache develops and worsens over several days. Photophobia (an intolerance of light) and neck stiffness arise, and signs of neurological impairment slowly evolve. This usually involves the part of the brain known as the brainstem, so that the neurological signs involve loss of balance or ataxia, problems with eye movement leading to double vision, and difficulty speaking. Many other neurological problems can develop, however. As the syndrome resolves over 2-10 weeks, the headache disappears.

Isolated meningitis is even more rare and would have very similar symptoms without the accompanying neurological signs.

People with Behçet's disease are prone to the development of blood clots in veins and, less commonly, arteries. Some patients have blood clotting disorders, but in others the reason is not understood and is thought to reflect all the inflammation going on in the body. When a blood clot affects the veins that remove blood from the brain there is a back-pressure effect, leading to an increase in brain pressure. This causes pain and may lead to visual problems if not treated quickly.

These problems constitute neurological emergencies in Behçet's disease, and will lead to hospital admission and treatment with steroids and immune suppressants or blood thinning drugs. However, these problems, thankfully, are rare.

Occasionally people can develop raised pressure within the brain very similar to that which occurs when the venous sinuses are blocked, called intracranial hypertension. In this case the venous sinus is not blocked, but the symptoms, although less severe, are similar. Treatment with drugs to lower the pressure, or occasionally a shunt to remove fluid, will settle the condition down easily.

Many people find that headache is a frequent and troublesome symptom yet no evidence of neurological involvement is found. The headache syndrome can be just as severe as those caused by meningitis or venous sinus thrombosis. These people have primary headache, and do not need the same kinds of treatment as those with secondary headache. Primary headache is the most common form of headache in Behçet's disease, and we do not currently understand why it is so common.

How common is it?

The first study to identify the prevalence of headache in Behçet's disease was carried out by me and the members of the Behçet's Syndrome Society. Members were asked to fill out a questionnaire in which they were asked whether they had ever had neurological symptoms, including headache, loss of balance, double vision, numbness or tingling, vestibular symptoms and others. A further questionnaire was sent to those who had responded, asking them to note the characteristics of any headache they had experienced, its frequency and duration, the presence of additional symptoms, and whether or not there was an aura. For those with recurrent headaches, the treatment they chose to use, and its usual effect, was also noted, and a severity score, validated in migraine, was used to assess headache-related disability.

Of the 327 members who returned the first questionnaire, 270 (83%) had noted headache as a recurrent symptom. Of the 223 who returned the second questionnaire, 201 (90%) had noted headache. Headache was recurrent and often lasted for days. The presence of throbbing, a unilateral onset, photophobia and phonophobia were taken to indicate a migraine-type headache; the absence of these symptoms, with a description of a dull constant ache around the head, was taken to indicate a tension-type headache. The severity scores suggested that 50 people (25%) had little or no disability, 28 (14%) had mild disability, 31 (15%) had moderate disability, and 95 (46%) had severe disability.

Most patients were inadequately treated; 84% used over-the-counter medicines, and only 5% were using treatments specifically for migraine headaches. Even fewer patients were using a migraine preventative such as pizotifen or a beta-blocker.

This study showed that people with Behçet's disease have headaches much more often than others, that these are more likely to be typical of migraine, and that they are more severe and frequent than in people with migraine who do not also have Behçet's disease. In London, we are about to start a series of further studies to find out more about these headaches, their relationship to other aspects of the disease and its treatment, and how we can best treat them. We are also about to start a project using special MRI scans that will help us to find out what happens in the brain when these headaches develop.

How is it treated?

I treat patients with primary headache disorders who have migraine-type headaches in the same way as I treat other patients with migraine. This involves using drugs such as anti-inflammatories (ibuprofen, diclofenac and the like) or triptans for the acute headache when it arises, and daily preventive medications such as pizotifen and beta-blockers and sometimes drugs for epilepsy and depression. All of these work very well in my experience, and people should under no circumstance continue to suffer these severe and disabling headaches.