SARCOIDOSIS AND THE NERVOUS SYSTEM

Dr D. Kidd, Consultant Neurologist

Sarcoidosis affects the nervous system in 5% of all patients (neurosarcoidosis). The disease can affect any part of the nervous system.

Symptoms, diagnosis and treatment options all depend on which part of the nervous system is inflamed. Patients are sometimes affected by multiple types.

Cranial neuropathy

Half of all patients with neurosarcoidosis have a simple cranial neuropathy such as a weakness of one half of the face. Sometimes other nerves are affected, causing problems with hearing, numbness of the face, weakness of the tongue, difficulty swallowing or double vision. Patients with cranial neuropathies respond well and quickly to steroids and the condition usually fully resolves itself.

Other Forms of Neurosarcoidosis

Of the remaining half of patients, two thirds have leptomeningitis, one quarter pachymeningitis and the remainder the vasculitic form. These cases are much more serious and require urgent evaluation and treatment

Leptomeningitis: The inner lining of the brain becomes inflamed and the inflammation speeds quickly into the brain which itself swells up. Symptoms can include headache, drowsiness, slowness of thinking, weakness or numbness, balance, visual and hearing problems.

Pachymeningitis: The outer lining of the brain or spinal cord becomes inflamed causing headaches, focal neurological signs such as weakness or numbness down one side, and occasionally seizures.

Vasculitis: This is caused by inflammation within the blood vessels of the brain. The blood vessels can become blocked, and small areas of inflammation can be seen on the surface of the brain on MRI scans.

Peripheral Neuropathies

When the peripheral nervous system (the nerves in the body rather than the brain and spinal cord) is involved, peripheral neuropathy may develop. This occurs in around 10% of patients and causes numbness and occasionally weakness of the hands and feet. It tends to be painless and mild and does not worsen.

Some patients can have a severe problem known as acute demyelinating polyradiculoneuropathy which develops at the onset of the systemic disease, but this improves with treatment. Some develop mononeuropathies when only one nerve is affected, for example in the hand.

Vasculitic neuropathy is a rarer and more severe condition which always deteriorates quickly and so requires urgent treatment.

Diagnosis

Diagnosis of neurosarcoidosis can usually be made after blood and spinal fluid tests and an MRI scan, but occasionally a biopsy of the brain or spinal cord is necessary.

Treatment

Treatment is with a high dose of steroids, suppression of the immune system with chemotherapy, and immunotherapy drugs such as Infliximab. Treatment is needed for at least 5 years.

Most patients respond well to medication but need to be monitored carefully by a neurologist, within a multidisciplinary team of respiratory physicians, rheumatologists, endocrinologists, immunologists and specialist nurses.

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Small Fibre Neuropathy

Small fibre neuropathy is common; patients complain of burning feet and occasionally in the hands. In most patients the condition is irritating but not deteriorating. In some it can be very severe and painful. Treatment is always helpful; anti-neuralgia medications (such as Gabapentin and Duloxetine) work well and those with very severe pain respond to Infliximab.

Polymyositis and Muscle Pain

When muscle is affected, it may be painful and cause weakness – this is known as polymyositis. It may also be a slowly deteriorating problem with muscle wasting and cause no pain. The polymyositis form responds to treatment, but the rare wasting form does not. Muscle involvement occurs in only 5% of neurosarcoidosis cases.

Early Diagnosis

The effectiveness of treatment in severe forms of neurosarcoidosis can be improved considerably by early and aggressive treatments given after investigation in specialist centres. Patients need to ensure that their treating doctors understand neurosarcoidosis sufficiently. SarcoidosisUK can help them find the units providing expert care.

Outlook

The earlier the treatment is given and the stronger the treatment used for the circumstance, the more likely it is that patients will respond well and recover reasonable neurological function. Some patients suffer damage to the nervous system which cannot be corrected, but many others do improve. Only a minority of patients suffer lasting neurological impairments. Thankfully nowadays neurosarcoidosis is rarely a fatal disease.