

Small Fibre Neuropathy

Small fibre neuropathy is common; patients complain of burning feet and occasionally also in the hands. In most patients the condition is irritating but not deteriorating but 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 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. Thankfully nowadays neurosarcoidosis is rarely a fatal disease.

Who We Are

SarcoidosisUK provide support and information to anyone affected by sarcoidosis. We also raise awareness and fund research. The vast majority of our funds go towards researching a cure. Contact us for information on our Support Groups network and free Nurse Helpline.

How You Can Help:

Donate to fund research:
www.sarcoidosisuk.org/donate

Get involved in medical research:
www.sarcoidosisuk.org/research/get-involved-sarcoidosis-research/

Thanks to:

Dr D. Kidd, Consultant Neurologist, Royal Free Hospital, London.

Contact Us

SarcoidosisUK, 49 Greek Street,
London, W1D 4EG

020 3389 7221

info@sarcoidosisuk.org

www.sarcoidosisuk.org

Charity number: 1063986

Facebook: facebook.com/groups/sarcoidosisuk

Twitter: @sarcoidosisuk

SarcoidosisUK

SARCOIDOSIS and the Nervous System



Sarcoidosis and the Nervous System

Sarcoidosis can occur in almost any organ. Sarcoidosis affects the nervous system in 5% of all patients (neurosarcoidosis). Neurosarcoidosis is therefore uncommon (only 20 cases per million people) but can be severe. Nonetheless, provided with expert care, the disease is usually straightforward to treat. Only a minority of patients suffer lasting neurological impairments.

The disease can affect any part of the nervous system. It does so through the development of granulomatous inflammation (in the same way it affects other organs such as the lungs, skin and liver). Symptoms, diagnosis and treatment options all depend on which part of the nervous system is inflamed. The main types of neurosarcoidosis are outlined in this leaflet. 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.** In this process the inner lining of the brain becomes inflamed and the inflammation speeds quickly in to the brain which itself swells up. This can cause a multitude of symptoms, depending mostly on the site of entry and the severity of the inflammation. Most patients develop headache, drowsiness, slowness of thinking and then other features such as weakness or numbness, balance, visual and hearing problems. The MRI scan is always abnormal, and the spinal fluid shows inflammatory cells.
- **Pachymeningitis.** The outer lining of the brain or spinal cord becomes inflamed. This causes headaches and focal neurological signs such as weakness or numbness down one side, and occasionally seizures. Patients have abnormal brain scans and occasionally can be diagnosed with brain tumours incorrectly because of what is shown on the scan.
- **Vasculitis.** This is the least common form of neurosarcoidosis and 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. Sometimes how vasculitis appears on a scan can be misinterpreted as strokes or multiple sclerosis. Occasionally the blood vessels can become narrowed and misshapen.

Diagnosis 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 is with a high dose of steroids, suppression of the immune system with chemotherapy, and immunotherapy drugs such as an infliximab. Treatment is needed for at least 5 years. Most patients respond well to this medication, but they need to be monitored carefully and most importantly by a neurologist with experience of the disease. This should be within a multidisciplinary team consisting of respiratory physicians, rheumatologists, endocrinologists, immunologists and specialist nurses.

Peripheral Neuropathies

When the peripheral nervous system (bodily nerves) is involved, a condition known as 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.

