Sarcoidosis is an inflammatory disease, first identified over 100 years ago, which can affect almost any body organ, although it usually starts in the lungs or lymph nodes.

The cause remains unknown, and the disease can appear and disappear suddenly, or develop gradually with fluctuating symptoms that can persist throughout life.

As the disease progresses, small lumps, termed granulomas, appear in the affected tissues.

In the majority of cases, these granulomas clear up, either with or without treatment.

However, in some cases, they do not remit and there is ongoing tissue inflammation that leads to scarring (fibrosis).

The USA National Institute of Health (NIH) notes:

"Sarcoidosis is currently thought to be associated with an abnormal immune response. Whether a foreign substance is the trigger; whether that trigger is a chemical, drug, virus, or some other substance; and how exactly the immune disturbance is caused are not known." ( [1] )

CNS sarcoidosis has been reported in approximately 5% of patients with sarcoidosis ( [2] ; [3] )
Chronic leptomeningitis in the basilar cisterns and hypothalamic regions are typical manifestations of CNS sarcoidosis ([4]). Bahr et al. ([5]) looked at 6 cases of intracranial sarcoidosis and found that communicating hydrocephalus with sarcoid arachnoiditis is the most common finding, although arteritis and masses have also been reported.

Kendall and Tatler ([6]) found that the spinal cord may be involved by intra-medullary granulomas or meningeal infiltration causing arachnoiditis.

Leptomeningeal granulomatous infiltration manifests as an intense meningeal enhancement on gadolinium-enhanced T1-weighted images (enhancement of the basal meninges still suggests an active inflammatory process, whereas its absence suggests fibrosis ([7]) and may go unnoticed with unenhanced MR ([8]).

The suprasellar and frontal basal meninges and the depths of the sulci are most frequently affected.

Occasionally, granulomas coalesce to form mass-like lesions, particularly in the region of chiasm, floor of the third ventricle, and pituitary stalk([9]).

Hydrocephalus may be a complication of chronic meningitis.

Hosseini and Tourbah ([10]) described a case of sarcoid related optochiasmic arachnoiditis, presenting as bilateral visual loss, pain on eye movement and headache, developing rapidly within a few days.

The patient had longstanding pulmonary sarcoidosis. CSF showed evidence of aseptic meningitis.
Cranial MRI showed hypertrophy of the chiasma and of the cisternal portion of both optic nerves, a hypersignal on T2 weighted images and post-gadolinium enhancement on T1 weighted images.

The patient improved on steroid treatment. Follow-up MRI 2 months later showed decrease in size of the affected nerves and loss of enhancement on T1 images.

Russian authors Makarov et al. ([11]) looked at neurosarcoidosis; they found granulomatous lesions in the central nervous system in 14% of patients with respiratory tract sarcoidosis.

Clinical classification of neurosarcoidosis included specific granulomatous forms: CNS arachnoiditis and perivasculitis, sarcoid myositis, as well as non-specific forms (vasoautonomic dystonic syndrome, angiotrophoneurosis).

Willigers and Koehler in The Netherlands ([12]), described a case in a 20 year old woman who presented with subacute amnesia. She was found to have neurosarcoidosis.

There were several features: isolated bilateral temporal hydrocephalus, caused by ventriculitis/arachnoiditis as well spinal arachnoiditis as demonstrated by myelography, which, incidentally, was not noticed on MR scan.

Cooper et al. ([13]) reported a case of neurosarcoidosis with optic nerve involvement as well as two intracranial parenchymal lesions and granulomatous arachnoiditis.

The authors noted that MRI offered no advantages over CT in the orbit but was significantly more accurate intracranially.


[The diagnosis and treatment of neurosarcoidosis] 
