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*Case Report*

# The radiological investigation of neurosarcoidosis

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Sarcoidosis is a disseminated disease of uncertain aetiology which causes neurological symptoms in 5% of cases, but more than 10% are found to have neurological involvement at autopsy. The diagnosis of neurosarcoidosis may be difficult if there are no extracerebral manifestations of the disease, particularly in view of the non-specific nature of many of the investigations and Magnetic resonance imaging has made an important advance. We report three recent cases.

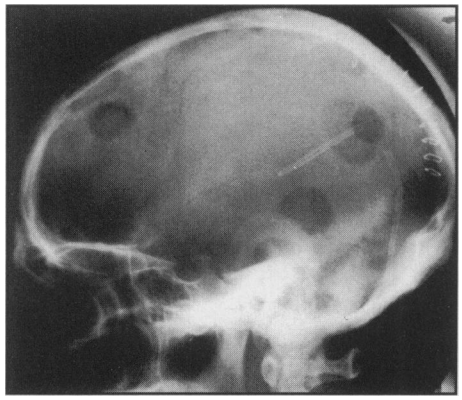

*Fig. 1A – Lateral Skull X-Ray*
Multiple lytic deposits in the skull vault. Note the presence of a ventricuar shunt.

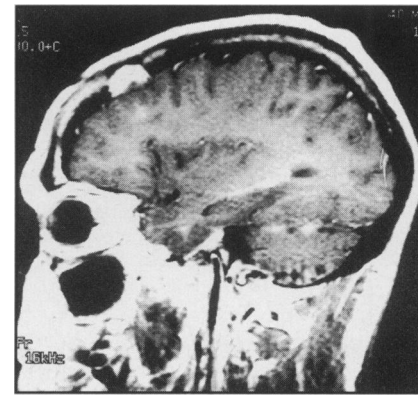

*Figure 1B – Sagittal T1-weighted MR image with gadolinium enhancement*
Enhancing nodule of soft tissue extending from the meninges into the left frontal bone.

## CASE 1

A 40 year old man presented with a short history of headaches and drowsiness. Apart from bilateral papilloedema examination was normal. Routine haematological investigations were normal. Computerised tomography (CT) (not shown) revealed hydrocephalus involving all ventricles, and multiple lytic lesions were noted in the skull vault (Fig lA). A shunt was inserted and CT-guided biopsy of a skull lesion performed. Histological examination showed a

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granulomatous process suggestive of sarcoidosis. Magnetic resonance imaging (MRI) of the brain (Fig 1B) showed multiple focal areas of enhancing soft tissue within the diploic space on the T1-weighted images with gadolinium enhancement. There was meningeal enhancement in the inter-hemispheric fissure anterior to the third ventricle and over the superior aspect of the left temporal lobe. These appearances were consistent with the pathological diagnosis.

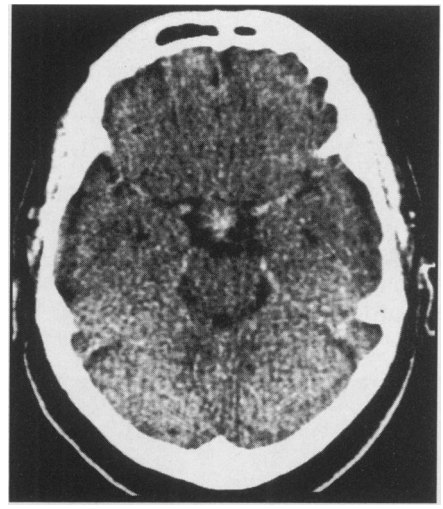

*Figure 2 – Axial CT scan of brain*
Nodule of enhancing soft tissue above the pituitary fossa, lying in the region of the suprasellar cistern and partially effacing the floor of the third ventricle.

## CASE 2

A 53 year old man with a ten year history of sarcoidosis previously involving skin, lungs, nasal space, lacrimal glands and left frontal sinus presented with loss of body hair, pallor, fatigue, myalgia and a thirst. He had required steroid and methotrexate therapy for long periods during the course of his illness. On examination visual fields were full and no muscular weakness or neurological deficit was detected. Serum angiotensin conventing enzyme levels were normal. Hypopituitarism and hypogonadism requiring hormone replacement were found.

A CT scan with intravenous contrast enhancement showed the pituitary fossa to be normal, (Fig 2). There was a nodule of enhancing soft tissue on the pituitary stalk, extending superiorly into the suprasellar cistern and the floor of the third ventricle. No other abnormality was found.

MRI scan of the brain was then performed. T1-weighted images with gadolinium enhancement confirmed the presence of a focal, nodular soft tissue mass of high signal adjacent to the pituitary stalk and optic chiasm extending to the floor of the third ventricle. There was no leptomeningeal enhancement. Although the appearances are not entirely specific, in view of the history a diagnosis of neurosarcoidosis was made.

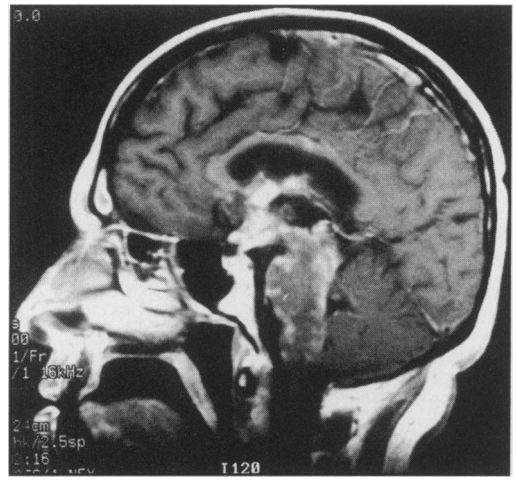


*Figure 3 – Sagittal T1-weighted MR image with gadolinium enhancement*
Extensive nodular meningeal enhancement from the upper cervical cord to the suprasellar cistern.

## CASE 3

A 47 year old woman presented with a history of increasing weakness of her left leg, urinary frequency and incontinence. Pulmonary sarcoidosis had been diagnosed twenty years previously. On examination she had a spastic paraparesis. No other neurological abnormality was detected and haematological tests were unhelpful.

Myelography was normal. MRI scan of the brain demonstrated extensive nodular meningeal enhancement from the upper cervical cord to the suprasellar cistern on the T1-weighted images with gadolinium (Fig 3). There were multiple subependymal nodules throughout the ventricular system, including the fourth ventricle and the septum pellucidum. Soft tissue enhancement was seen in the suprasellar cistern extending around the anterior aspect of the third ventricle and into the inferior part of the right frontal lobe. On the T2-weighted images there was patchy high signal in the pons, lower mid-brain and in the periventricular white matter.

## DISCUSSION

The typical presentation of neurosarcoidosis is with cranial nerve palsies, particularly facial, but others include hypopituitarism and paraparesis. Less common presentations include amnesic syndrome,[1] intractable hiccoughs,[2] seizures,[3] psychosis,[4] and acute stroke.[5] This wide range of neurological manifestations is reflected in the many neuroradiological signs.

The most commonly reported radiological finding in neurosarcoidosis is hydrocephalus, as in case I. This is usually the result of fourth ventricular outflow obstruction or of reduced resorption of cerebro-spinal fluid. Meningeal involvement is usually in the form of plaques or nodules, showing as focal or diffuse thickening with contrast enhancement. This finding is rather nonspecific

and is also seen with meningeal metastases and infection. Parenchymal disease is usually isodense or hypodense on CT, and may demonstrate contrast enhancement.[6] Magnetic resonance imaging is superior to computerised tomography, however, especially for imaging the hypothalamic region, the periventricular white matter [7] and for its increased sensitivity to meningeal disease.

Magnetic resonance imaging with gadolinium enhancement is now recognised as the most sensitive imaging investigation for neurosarcoidosis. In a series of 20 patients with neurosarcoidosis, 17 had an abnormal MRI, but only three of these were detected on unenhanced T1-weighted scans confirming the importance of using intravenous gadolinium.[7] The MRI appearances reflect the pathological process of granulomatous dural invasion with plaques, focal masses and a leptomeningitis with parenchymal infiltration along the perivascular spaces of Virchow-Robin.[8] Inflammation of the Virchow-Robin spaces can develop into granulomatous masses associated with oedema (seen optimally on T2-weighted images) due to disruption of the blood/brain barrier, this process occurs most commonly in the basal areas of the brain. The most common site of involvement is the leptomeninges, but others include hypothalamus, periventricular white matter, optic chiasm and the pituitary gland. Gadolinium alters local magnetic environments to change signal intensity in tissues where it accumulates. It does not cross the intact blood-brain barrier to any great extent and therefore a normal scan can result if insufficient disease is present or if there is primarily extracranial involvement of the cranial nerves.

The appearance of leptomeningeal enhancement on MRI can be mimicked by bacterial and fungal infections, metastatic infiltration and leukaemia but has become the imaging technique of first choice where neurosarcoidosis is clinically suspected.

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