

LESSON OF THE MONTH

Sarcoid tumour: continuing diagnostic problems in the MRI era

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The effects of sarcoidosis on the nervous system are protean.¹⁻³ They include involvement of the meninges (cranial nerve palsies, pituitary, and hypothalamic effects), peripheral nerves (symmetric polyneuropathy, multifocal neuropathy), and brain parenchyma (lesions of brain and spinal cord without obvious meningeal involvement)¹⁻³; and also secondary effects of systemic disease, such as intracerebral haemorrhage related to sarcoid induced thrombocytopenia.⁴

Solitary sarcoid mass lesions of the CNS, mimicking tumours, have been described but are rare, with less than 100 cases in the world literature.⁵⁻⁸ They may either arise from the dura or be located entirely within the parenchyma, with radiological appearances which resemble meningioma and glioma respectively. Computerised tomography was generally not helpful in distinguishing these lesions from other intracerebral masses⁵⁻⁷ and it was therefore hoped that the advent of MRI would be useful in the differentiation of sarcoid tumours.² We present two patients with solitary intracerebral mass lesions which were thought to be tumours, in whom the diagnosis of neurosarcoidosis was entirely unexpected until histological specimens were available, despite appropriate MRI.

Case reports

CASE 1

A 28 year old AfroCaribbean man presented with episodes of cramping and numbness in the right calf, followed by loss of consciousness for about 2 minutes, during which time he was seen to convulse and froth at the mouth but without urinary incontinence or tongue biting. No abnormal neurological signs were found on examination. The episodes were diagnosed as focal seizures with secondary generalisation and he was started on 100 mg carbamazepine twice daily. Subsequently he had an episode of jerking of the right leg not associated with loss of consciousness. Initial investigations included CT of the brain which was reported to be normal, and an EEG which showed prominent background theta activity but no epileptiform activity. Brain MRI showed a focal lesion over the precentral gyrus (a location which may account for the normality of the CT) attached to dura. This solitary lesion showed uniform enhancement with intravenous gadolinium-DTPA (fig 1) but there was no meningeal enhancement. These features were thought to be indicative of a meningioma, despite the patient's young age. At surgery, the lesion was resected; its gross appearances were thought to be in keeping with the diagnosis of meningioma.

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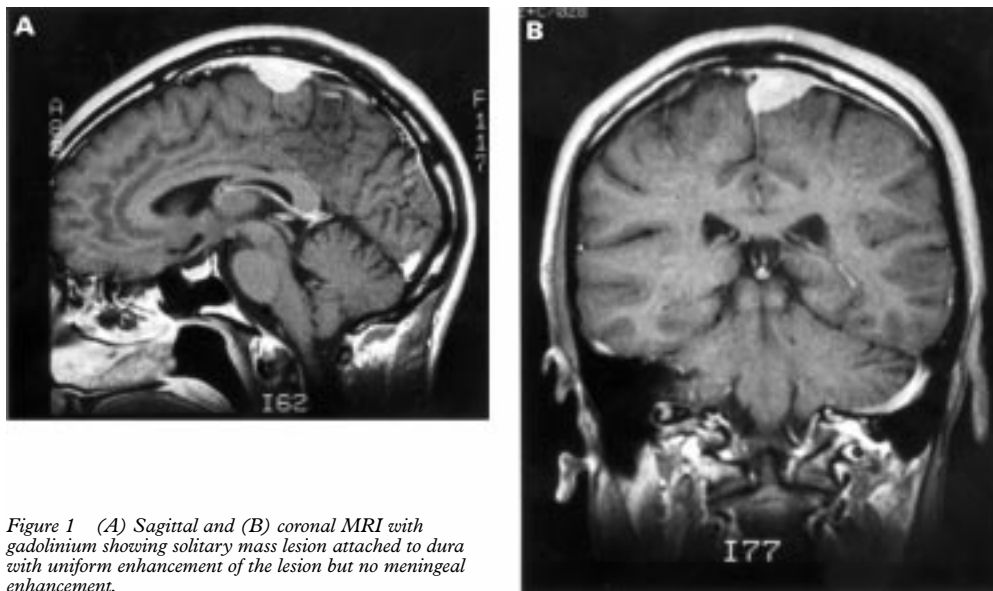


Figure 1 (A) Sagittal and (B) coronal MRI with gadolinium showing solitary mass lesion attached to dura with uniform enhancement of the lesion but no meningeal enhancement.

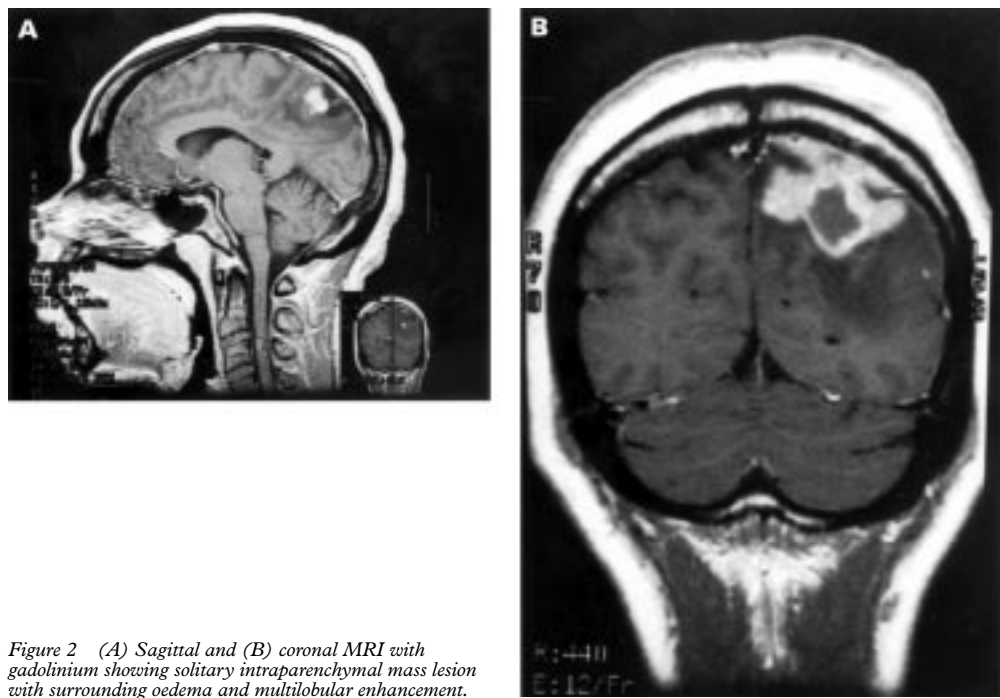


Figure 2 (A) Sagittal and (B) coronal MRI with gadolinium showing solitary intraparenchymal mass lesion with surrounding oedema and multilobular enhancement.

Histological examination of the mass disclosed multiple small granulomata of epithelioid macrophages with occasional giant cells and lymphocytes, with no foci of necrosis. Stains were negative for acid fast bacilli and fungi. A pathological diagnosis of granulomatous pachymeningitis was made. Further investigations showed normal blood levels of ionised calcium and serum angiotensin converting enzyme (ACE), but a mild lymphopenia ($0.7 \times 10^9/l$); chest radiography showed minimal hilar lymphadenopathy but spirometry and gas transfer were normal. Gallium scan showed marked lacrimal gland uptake with some mediastinal foci. A Kveim test showed an 8 mm papule at 4 weeks, biopsy of which showed granulomatous inflammation with several multinucleate giant cells, consistent with the diagnosis of sarcoidosis.

CASE 2

An AfroCaribbean woman aged 33 presented with a cluster of four right sided sensorimotor seizures with secondary generalisation. On examination she was drowsy and had a mild right hemiparesis. Brain MRI showed a left posterior parietal intraparenchymal mass lesion with surrounding oedema. After intravenous gadolinium-DTPA, the lesion showed multilobular enhancement with a non-enhancing core; there was neither meningeal enhancement nor any other lesion seen (fig 2). A malignant glioma was strongly suspected, so much so that it was debated whether biopsy was necessary to confirm the diagnosis or whether the patient should be referred directly for radiotherapy. The ensuing stereotactic biopsy showed granulomata of epithelioid histiocytes without necrosis, suggestive of sarcoidosis. A percutaneous liver biopsy showed multiple non-caseating granulomata.

Discussion

Sarcoidosis affects the nervous system in only about 5% of cases. Most of these patients are known to have systemic sarcoidosis and then develop neurological signs; a small group have systemic disease presenting with neurological signs (as is the case for the two patients reported here); very rarely, sarcoidosis may be confined to the nervous system.¹⁻³ According to a recently developed classification, both our patients had clinically definite CNS sarcoidosis on account of their positive histology.⁹

The neuroradiological features of neuro-sarcoidosis include meningeal enhancement with contrast agents, involvement of the hypothalamic and hypophyseal regions, and high signal intensity in the white matter on T2 weighted MRI. Periventricular and multifocal white matter lesions were the most common pattern seen in one MRI series, which concluded that MRI was better than CT for the detection of parenchymal changes in CNS sarcoidosis.¹⁰ The two cases reported here exemplify sarcoid mass lesions which resemble cerebral neoplasms radiologically. Despite the use of MRI protocols appropriate to diagnose the commoner patterns of neurological involvement in sarcoidosis, solitary lesions without meningeal enhancement may elude specific diagnosis until histology is available. The hope that MRI might be a more specific investigation to establish the diagnosis of sarcoid tumour may not therefore be the case.

A very high index of clinical suspicion remains necessary to consider the diagnosis of sarcoid tumour preoperatively, clues being the occurrence of mass lesions in young patients of AfroCaribbean origin. In the future, magnetic resonance spectroscopy may be able to differentiate sarcoid tumour from

other cerebral mass lesions; this technique is reported to be helpful in the differential diagnosis of cerebral tumours non-invasively.¹¹

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