Gadolinium ring enhancement and mass effect in acute disseminated encephalomyelitis

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Abstract. A 9-year-old boy presented with a subacute history of optic neuritis followed by brainstem involvement, with fever and a lymphocytic pleocytosis in the cerebrospinal fluid. Gadolinium-enhancing ring lesions were demonstrated in the white matter of the cerebrum, brainstem and cerebellum on day 17 of the illness, all appearing simultaneously as part of a monophasic illness. A parietal lesion exerted mass effect. Needling and biopsy yielded no evidence of a pyogenic lesion, tumour or tuberculosis and showed vasculitis. There was insufficient material for myelin staining. Dexamethasone therapy lead to rapid improvement of the radiological lesions: MRI and CT on day 34 of the illness showed complete clearing of the lesions except for residual abnormality at the biopsy site.

Key words: Acute disseminated encephalomyelitis – CT – MRI – Ring lesions – Mass effect

Acute disseminated encephalomyelitis (ADEM) includes postexanthematous, postvaccinial and postinfective widespread inflammation of the central nervous system, representing a host immune reaction. We present the gadolinium enhancement findings on MRI in this condition. This is the second report of enhancement with gadolinium [1] and the first to describe ring enhancement on MRI and mass effect of demyelination in ADEM.

Case report

A 9-year-old boy presented with a four day history of bilateral blindness, papillitis and unreactive pupils. No history of antecedent illness was obtained. CT of the brain on day 4 of the illness was normal (Fig. 1a) although a polyp was noted in the left maxillary antrum. His level of consciousness deteriorated to a Glasgow Coma Scale (GCS) score of 6/15 between day 4 and day 8 of the illness. Brain stem involvement was noted, with skew deviation, internuclear ophthalmoplegia and bilateral upper motor neuron signs in the limbs, more pronounced on the right. CT on day 8 showed areas of decreased density in both parietal regions, with contrast enhancement, but without mass effect (Fig. 1b). Analysis of the cerebrospinal fluid revealed a lymphocytic pleocytosis (25 per mm³), glucose 3.8 mmol/l (serum glucose 6.3 mmol/l) and protein 0.17 g/l. A test for oligoclonal bands was not performed. The patient received penicillin and chloromycetin for possible sinusitis and septic thrombophlebitis, although an otorhinolaryngologist did not consider the sinus abnormalities significant. A course of acyclovir was given, although antibody tests for Herpes simplex, measles and Epstein Bar viruses were negative. Rheumatoid and antinuclear factors were negative and protein electrophoresis showed an acute phase reaction. The patient's level of consciousness gradually improved to normal by day 29 of the illness.

Magnetic resonance imaging (MRI) of the brain on day 17 showed areas of low signal intensity on T1 weighting in the white matter of the cerebrum, brainstem and cerebellum (Fig. 2a) which appeared bright on T2 weighting (Fig. 2b). These enhanced in a ring-like fashion with gadolinium (Fig. 2c). CT (day 18) showed several ring-enhancing lesions (Fig. 3). The left parietal lesion exerted mass effect, and was initially thought to represent an abscess, but attempted drainage the same day failed to yield pus or fluid. A brain biopsy the next day excluded an abscess, tuberculosis or tumour but did show neutrophilic arteritis and a perivascular polymorphonuclear leukocytic infiltrate in the meninges. Perivascular haemorrhage was present (Fig. 4). Unfortunately the biopsy did not yield enough tissue for myelin staining.

Dexamethasone 5 mg thrice daily was given orally on day 18, and CT on day 22 showed remarkable improvement (Fig. 5). The neurological condition continued to improve, so that the patient could talk, walk and see fingers. By day 34 the MRI had returned to normal (Fig. 6) with the exception of some enhancement at the biopsy site.

Discussion

The clinical and radiological course, anatomical location of the lesions, favourable response to steroids and brain biopsy findings would be consistent with the diagnosis of ADEM.

CT in ADEM has demonstrated multiple areas of low attenuation [2-4] including involvement of the centrum semiovale [5] and grey matter [4]. Spotty, nodular gyral enhancement [3, 6] or ring enhancement [4, 7] has been observed. Other demyelinating conditions showing ring enhancement [7-9] and mass effect [7, 9] on CT have been
reported. Normal CT studies with abnormal MRI in ADEM are well described [10–13]. Our patient had normal CT in the early phase of the illness, but subsequently showed low attenuation lesions, gyral and multiple ring enhancement, which improved rapidly on high dose steroid therapy.

There have been several reports of high signal lesions in the cerebral white matter on T2-weighted images in ADEM [5, 6, 14]. Their multiplicity [10–12, 15], with involvement of the cerebrum, cerebellum and brain stem [1, 12, 13, 16] in a way indistinguishable at times from multiple sclerosis [10, 15] has been noted. Rapid improvement with high dose steroid therapy has been reported [13]. Decreased signal intensity of the lesions on T1 weighting has been observed [14, 16]. Diffuse and nodular gadolinium enhancement of the demyelinating lesions has been reported [1], as part of a monophasic illness. Our patient demonstrated multiple gadolinium ring-enhancing lesions which appeared to be part of a monophasic illness involving the white matter of the cerebrum, cerebellum and brainstem. Gadolinium ring enhancement has been reported with other demyelinating conditions such as multiple sclerosis [17] and central pontine myelinolysis [18].

Demyelination causing mass effect has been reported in multiple sclerosis [7, 9] and ADEM [7]. Our patient showed multiple lesions with relatively little mass effect.