SHORT REPORT

Acute disseminated encephalomyelitis presenting as a solitary brainstem mass

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Received 21 February 1992
and in final revised form
22 October 1992
Accepted 6 November 1992

Abstract

A 36 year old woman presented with a subacute brainstem syndrome. MRI showed a solitary, gadolinium enhancing brainstem mass, which on biopsy showed perivenous inflammation and demyelination compatible with acute disseminated encephalomyelitis.

(*J Neurol Neurosurg Psychiatry* 1993;56:920-922)

Acute disseminated encephalomyelitis (ADEM) is a monophasic, frequently post-infectious, CNS disorder, in which there is usually clinical evidence of multiple lesions involving CNS white matter.^{1,2} The characteristic pathological appearances are of widespread, multiple small foci of perivenous inflammation and demyelination³; large lesions with mass effect are uncommon. We report a case of probable ADEM in which clinical and imaging features demonstrated a solitary mass lesion in the brainstem.

Case report

A 36 year old woman was admitted to hospital with a two week history of mild headache and nausea, accompanied by dysphagia and nasal regurgitation. For five days, she had noticed unsteadiness on walking. Examination revealed a left Horner's syndrome, gaze evoked bilateral horizontal nystagmus, reduced right facial sensation with bilaterally impaired corneal reflexes, and a diminished left gag reflex. There was mild gait ataxia. CSF revealed 1 white cell $\times 10^6/l$, protein 0.34 g/l, glucose 3.4 mmol/l, and no oligoclonal bands. Visual evoked potentials were normal. CT head scan was normal.

Over the next week, her condition worsened, with increasing gait ataxia, along with ataxia of left sided limbs. She developed an aspiration pneumonia, and was intubated and ventilated. At this time, MRI revealed a mass lesion in the left side of the medulla and pons, situated dorsally and adjacent to the fourth ventricle; it displayed uniform gadolinium enhancement (fig 1). No other abnor-

Figure 1 (a) Sagittal and (b) axial gadolinium enhanced T1 weighted brain MRI reveal an enhancing brainstem mass in the medulla.

Figure 2 (a) Photomicrograph of a small blood vessel in brainstem white matter surrounded by small mononuclear cells. Haematoxylin and eosin $\times 300$. (b) Photomicrograph showing the patchy distribution of the myelin loss. In the demyelinated areas the cell density is not increased. Luxol fast blue/cresyl violet. $\times 120$.

malities were present.

She then underwent stereotactic MRI guided biopsy of the brainstem lesion, which revealed multiple small foci of perivascular inflammation and demyelination (fig 2). The appearances were considered to be compatible with a perivenous encephalomyelitis seen following viral infections. Multiple sclerosis (MS) was thought to be less likely, because of the uneven distribution of the areas of demyelination.

She was treated with pulsed high dose intravenous methylprednisolone, but in spite of this her condition continued to deteriorate over the next two weeks. She developed a complete bulbar palsy, with loss of all tongue and palate movements, left hemiparesis with loss of posterior column sensation in the left arm, a conjugate gaze palsy to the left, left facial weakness, and defective upgaze. Repeat MRI showed an increase in the size of the pontomedullary mass, particularly on the left, and confirmed that the biopsy site was from within the lesion. She was treated with a further course of high dose intravenous methylprednisolone, in combination with antibiotics and acyclovir. Subsequent viral serology, which included herpes simplex and zoster, was negative.

Over the next year there was a gradual improvement in her clinical state. After two months, she was successfully weaned from daytime positive pressure ventilation, but has continued to require nocturnal ventilation. After 18 months, she still has severe disequilibrium and limb ataxia, and had required epiglottic plication and cricopharyngeal myotomy to prevent recurrent aspiration pneumonia to severe bulbar weakness. Repeat MRI after three months showed a marked reduction in size of the original lesion with loss of mass effect.

Discussion

This patient presented with a subacute brain-

stem syndrome, and MRI revealed a solitary, enhancing, mass lesion. The differential diagnosis included glioma, lymphoma, or granuloma. A biopsy was performed which showed perivenous inflammation and demyelination suggestive of ADEM.

For several reasons, ADEM was considered unlikely in the initial differential diagnosis. Firstly, there was no history of a preceding infection. A prior clinical infection occurs in most but not all ADEM cases. Secondly, the MRI features were unusual for ADEM in that there was mass effect, and only one lesion.

Mass lesions do occasionally occur in ADEM, both in the cerebral hemispheres and spinal cord,⁴ although this is the first reported case we know of with a brainstem mass. Also, while there are usually multiple cerebral white matter lesions on MRI in ADEM,² this is not invariably the case.⁵

The evolution of the MR lesion in our patient is certainly typical of an inflammatory/demyelinating CNS lesion. Enhanced CT or MRI characteristically show blood-brain barrier impairment in acutely symptomatic lesions both in MS⁶ and ADEM.⁷ In MS, gadolinium enhancement has been correlated with perivascular inflammation,⁸ and it is likely that the same applies to ADEM. The subsequent reduction in size is also characteristic of inflammatory/demyelinating CNS lesions,⁹ and is probably due mainly to resolution of oedema, although a degree of remyelination is not excluded.

Although acyclovir was given, the pathological features and negative viral serology make it unlikely that this was brainstem encephalitis due to herpes simplex. We consider that it is best regarded as a localised form of ADEM, akin to the well recognised syndromes of post-infectious transverse myelitis and optic neuritis, in which immunopathogenic mechanisms are probably important. Transverse myelitis may also occur without evidence of disseminated MR

white matter lesions.^{2 10 11} Another possibility is that this was the first episode of MS. However, this seems less likely since there were no oligoclonal bands in the CSF, there was no MR evidence for disseminated lesions, and the pathological features were more typical of ADEM than MS.

This case emphasises that ADEM should be considered in the differential diagnosis of subacute MR enhancing brainstem masses.

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