Diagnosing multiple sclerosis: expect the unexpected

Introduction

Isolated cranial nerve palsies are rarely encountered as part of the phenotype of multiple sclerosis. This article reports a patient presenting on two occasions 5 years apart with isolated third and sixth cranial nerve palsies whose neuroimaging and CSF findings were diagnostic of multiple sclerosis.

Discussion

This patient had a number of unusual features for a diagnosis of multiple sclerosis. Whereas optic neuritis, internuclear ophthalmoplegia and nystagmus are frequent features of multiple sclerosis, and diplopia a common symptom, ocular motor nerve palsies are rare. Although third cranial nerve fascicular lesions are said to be most often caused by infarction, haemorrhage or demyelination (Brazis et al, 2001), multiple sclerosis presenting with an isolated third nerve palsy is very uncommon (Desai and MacFadyen, 1987; Newman and Lessell, 1990; Thömke et al, 1997). Likewise, isolated sixth nerve palsy reflecting pontine tegmental multiple sclerosis plaques is rare, estimated to be the presenting sign of multiple sclerosis in 0.5% of patients (Barr et al, 2000).

Furthermore, the prevalence of multiple sclerosis in Hong Kong Chinese is very low (Yu et al, 1989; Lau et al, 2002), and since this patient emigrated after adolescence his risk should remain that of his place of birth. It has also been reported that the presence of CSF oligoclonal banding in Hong Kong Chinese with multiple sclerosis is low (40%; Yu et al, 1989).

Hence this patient showed the conjunction of three very unusual features for multiple sclerosis: consecutive presentations with isolated cranial nerve palsies and his racial origin. All of these features might have served to lower clinical suspicion of the diagnosis, had it not been for the typical appearances of demyelinative supratentorial plaques on magnetic resonance brain imaging. Although imaging did not identify causative brainstem or cranial nerve lesions, no other explanation for the clinical picture was forthcoming. The lesson we have learned is that in multiple sclerosis one must always be ready to expect the unexpected. BJHM

Case Report

A previously healthy 41-year-old Hong Kong Chinese man, who emigrated to the UK at the age of 20 years, presented with a complaint of visual blurring and of painless double vision but without directional selectivity. Ophthalmological assessment found normal visual acuity, colour vision, visual fields, and pupillary reactions; examining eye movements there was a left hypertropia, with weak left inferior rectus on Hess chart. A diagnosis of acute partial pupil-sparing left oculomotor (third) nerve palsy was made.

Symptoms resolved spontaneously within 2 weeks, but magnetic resonance brain imaging arranged at the initial consultation showed multiple high signal intensity lesions in the white matter of the cerebral hemispheres periventricularly and in the corpus callosum, suggestive of demyelination, although no brainstem lesions were noted. When subsequently seen in the neurology clinic, the patient was asymptomatic and there were no neurological or neuro-ophthalmological signs. Serum angiotensin-converting enzyme level and chest X-ray were both normal. Interval magnetic resonance brain imaging 7 months later showed no new changes, and since the patient was asymptomatic he declined further investigation or follow up.

Five years later, the patient presented again with a complaint of double vision without directional selectivity. Signs were now of a right abducens (sixth) nerve palsy, without any other neurological or neuro-ophthalmological signs. Symptoms resolved spontaneously after 4 months. Magnetic resonance brain imaging again showed multiple high signal lesions in the periventricular white matter and corpus callosum, but no brainstem or cerebellar lesions were seen. Pattern shift visual evoked potentials were within normal limits, providing no evidence for a demyelinating optic neuropathy. The patient agreed to lumbar puncture, and CSF analysis showed normal protein, glucose, cell count, angiotensin-converting enzyme level, and cytology, but oligoclonal bands were present in a type 2 pattern consistent with a diagnosis of multiple sclerosis.

Barr D, Kupersmith MJ, Turbin R, Bose S, Roth R (2000) Isolated sixth nerve palsy: an uncommon presenting sign of multiple sclerosis. *J Neurol* **247:** 701–4

Brazis PW, Masdeu JC, Biller J (2001) *Localization in Clinical Neurology.* 4th edn. Lippincott Williams & Wilkins, Philadelphia: 182

Desai HB, MacFadyen DJ (1987) Multiple sclerosis presenting as third nerve palsy. *Can J Neurol Sci* **14:** 178–9

Lau KK, Wong LKS, Li LSW, Chan YW, Li HL, Wong V (2002) Epidemiological study of multiple sclerosis in Hong Kong Chinese: questionnaire survey. *Hong Kong Med J* **8:** 77–80

Newman NJ, Lessell S (1990) Isolated pupil-sparing third-nerve palsy as the presenting sign of multiple sclerosis. *Arch Neurol* **47:** 817–18

Thömke F, Lensch E, Ringel K, Hopf HC (1997) Isolated cranial nerve palsies in multiple sclerosis. *J Neurol Neurosurg Psychiatry* **63:** 682–5

Yu YL, Woo E, Hawkins BR, Ho HC, Huang CY (1989) Multiple sclerosis amongst Chinese in Hong Kong. *Brain* **112:** 1445–67

Dr J Ramtahal *is Clinical Lecturer in Neurology and* **Dr AJ Larner** *is Consultant Neurologist in the Walton Centre for Neurology and Neurosurgery, Fazakerley, Liverpool L9 7LJ*

Correspondence to: Dr AJ Larner