Clinical case review

Tonic spasms and demyelinating lesions

Commentary by MICHAEL P. PENDER MB BS(Hons I), MD, PhD, FRACP

After an episode of dysarthria, severe left-sided spasms, weakness and incoordination, this patient's MRI brain scan reveals three brain lesions that are consistent with a demyelinating process.

Case scenario

A 39-year-old mother of two children who had been working very hard and studying presented with exhaustion, recurrent headaches and some features of depression. She also mentioned that she had experienced frequently recurring 'shudders' down the left side of her body, which she had attributed to her chronic tiredness. Over the previous few months, she also felt that her speech had been slurred at times, but again she attributed this to fatigue. A full neurological examination at the time was normal.

Shortly after this presentation, the patient experienced an acute episode of dysarthria and severe left-sided spasms, weakness and incoordination. An urgent MRI brain scan revealed three lesions: one deep in her left frontal area, a second in the left parietal region and a third in the pons. The lesions were reported as being consistent with a demyelinating process. The patient had no previous neurological problems and in particular no history of visual or bladder disturbance.

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Commentary

This patient's episodes of 'shudders' and spasms involving the left side of the body are suggestive of the phenomenon of tonic spasms. These are characterised clinically by brief episodes of dystonic posturing, usually involving the upper and/or lower limb and sometimes the face on the same side of the body. The episodes are usually brief (lasting up to 30 seconds) but can occasionally last for up to two minutes and occur as frequently as several times in an hour. There is no associated alteration in consciousness. During the spasms the patient may experience pain and/or other discomfort in the muscles involved.

Tonic spasms are most commonly due to multiple sclerosis (MS) but can also occur in other neurological disorders such as cerebral infarction. In MS, the lesions responsible for tonic spasms involve the corticospinal tracts anywhere below the cerebral cortex, with the internal capsule and cerebral peduncle being the most common sites.1

Underlying mechanism

The probable mechanism underlying tonic spasms is ectopic impulse generation in demyelinated corticospinal axons spreading radially to adjacent, closely packed, demyelinated fibres by ephaptic ('false synaptic') transmission. Sometimes hyperventilation, emotion, movement or tactile stimulation trigger the spasms, which are often preceded by a transient sensory disturbance in the region affected. Within each patient the spasms are stereotyped. Depending on the region involved, they consist of:

- flexion of the wrist and metacarpophalangeal joints
- extension and abduction of the fingers
- flexion or extension of the elbow
- flexion or extension of the lower limb
- facial distortion
- head turning to the side opposite the involved limbs.

Distinguishing tonic spasms from epileptic seizures

Tonic spasms can be distinguished from epileptic seizures by the preservation of consciousness, the brief duration and high frequency of episodes, the often painful nature of the attacks and the fact that during and between attacks the electroencephalogram is normal.

Diagnosis and prognosis

In this patient, the most likely explanation for her left-sided spasms is a lesion involving the corticospinal fibres in the posterior limb of the right internal capsule or in the right side of the brainstem. The MRI report does not mention any lesions in the right cerebral hemisphere but does describe a pontine lesion, which could be responsible for her spasms. The pontine

continued

lesion could also account for her dysarthria and left-sided weakness and inco-ordination.

The patient's pontine and left cerebral MRI lesions are most likely due to MS, which would also explain her fatigue and depression. Fatigue is a common manifestation of MS and major depression occurs at some stage in 50% of cases.² At present, this patient does not meet the criteria for a diagnosis of MS because there is insufficient evidence of dissemination of CNS lesions in space and time. It is probable that in the future she will experience further episodes of neurological dysfunction involving other regions of the CNS. Subsequent MRIs of the brain and spinal cord may also show evidence of new lesions.

Management

Tonic spasms respond dramatically to treatment with carbamazepine (Tegretol, Teril) 100 to 200 mg twice daily. Because tonic spasms in MS generally remit spontaneously after several weeks, carbamazepine can usually be withdrawn after about two months. If the patient's symptoms of dysarthria, weakness and incoordination are interfering with her activities of daily living, an intravenous infusion of methylprednisolone (Solu-Medrol, Methylprednisolone Sodium Succinate for

Injection) 1 g daily for three days should also be considered because it accelerates recovery from MS attacks.

References

- 1. Spissu A, Cannas A, Ferrigno P, Pelaghi AE, Spissu M. Anatomic correlates of painful tonic spasms in multiple sclerosis. Movement Disorders 1999; 14: 331-335.
- 2. Siegert RJ, Abernethy DA. Depression in multiple sclerosis: a review. J Neurol Neurosurg Psychiatry 2005; 76: 469-475.

DECLARATION OF INTERESTS: None.

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