Delayed-onset posthemiplegic dystonia and imitation synkinesia

The late development of hemidystonia following hemiplegia is an uncommon sequela of stroke. Its pathogenesis is still controversial but may involve lesions in the contralateral caudate nucleus, lentiform nucleus, and thalamus, or combinations of these. Imitation synkinesia was defined by Marie and Foix as “involuntary symmetrical movements tending to reproduce on one side of the body, movements executed by the other.” It is usually associated with a thalamic or parietal lesion. We report a patient who developed a previously undescribed association of hemidystonia and imitation synkinesia as a late complication of a thalamic haemorrhage.

A 74-year-old man with a three-year history of hypertension developed acute headache, right hemiplegia, and hemianesthesia in October 1982. The right limbs were hypreflexic and the right plantar response extensor. Computed tomography (CT) showed a left thalamic haematoma with involvement of the internal capsule. Two months later, the patient was able to walk with crutches although his hemianesthesia remained unchanged. Six months after the stroke, he developed voluntary pulling and twisting movements in his right arm and shoulder. Progressively, the involuntary movements spread to the neck, mouth, tongue, and the right side of his face. As a result, he had slight difficulty with speech and swallowing. His eyelids intermittently screwed up forcefully, especially on the right side. His right leg was deformed, with a flexed knee, extended hip, and clawed foot. Because of the leg dystonia, he found it difficult to walk and was almost confined to a wheelchair from November 1984. Burning paraesthesia gradually developed in his arm and leg, and his body. In August 1985, the patient’s dystonic right hand started to imitate involuntarily simple movements that were performed by the left hand, such as waving fingers.

In December 1986, he was admitted because of increasing swallowing disturbance, hemidystonia, and paraesthesia of the right arm and leg. On examination, he was alert and intellect was not impaired. His cranial nerves were intact and eye movements were full and the optic fundi were normal. Mild hemiparesis and hyperreflexia without muscle atrophy were noted in the right limbs but plantar responses were flexor. All sensory modalities were diminished in the right side of the body and in the right extremities. His right shoulder was adducted and fixed, and the elbow was slightly flexed. The forearm was deformed with a flexed wrist and hyperextended fingers, particularly seen in an outstretched position. There were orolingual dystonia, bilateral blepharospasm, and grimacing of the right side of his face. Waving his left hand voluntarily, the patient simultaneously waved his right hand unintentionally. These imitative movements of the right hand could also be induced by moving either foot but not by passive movement of the left hand or either foot.

Routine haematological, electrolyte, and liver and renal function tests were normal. Serology for syphilis was negative. A chest radiograph was normal. CT showed hypodensity in the left thalamus. Surface EMG recordings showed simultaneous muscle activities on both right and left abductor pollicis brevis, when the patient was asked to abduct his left thumb (fig 1a). Similarly, there were muscle activities on the right wrist extensor muscles simultaneously with left anterior tibialis during dorsiflexion of the left foot (fig 1b). An EOG showed diffuse slow waves. Somatosensory evoked potentials (SEP) following stimulation of the left median and tibial nerves were normal. No cortical SEP was obtained following the right median or tibial nerve stimulation. Daily regimens of 6 mg diazepam, 300 mg levodopa, 6 mg haloperidol, 15 mg baclofen, 75 mg dantrolene, 300 mg phenytoin and 600 mg carbamazepine had been tried without effect.

A left stereotactic thalamotomy was performed in January 1987. Post-operatively, there was marked improvement of both involuntary movements and posthemiplegic dystonia, blepharospasm, hemidystonia, imitation synkinesia and in the burning paraesthesia in the right limbs. The mild hemiparesis and paraesthesia were unchanged. Two weeks after operation the patient was able to take food without difficulty and to walk with support. He remained in the same condition, without medication, two years after the operation.

The points of particular interest in this patient were first, the unique association of the late development hemidystonia with imitation synkinesia in the hemiparetic limbs and, second, the marked improvement of both involuntary movements and posthemiplegic dystonia. These observations may indicate that hemidystonia and imitation synkinesia share a common pathogenesis.

Disorganisation of thelemniscal system at any level may be responsible for imitation synkinesia.1 This might be a plausible explanation for the appearance of imitation synkinesia on the right hand with ipsilateral hemianesthesia in our patient. Further studies are needed to understand the basis of thalamic haemorrhage. However, it is difficult to explain the distinct improvement of imitation synkinesia after stereotactic thalamotomy.

Burke et al have suggested that delayed onset dystonia is the result of slowly evolving aberrant neuronal sprouting, stimulated by the original lesion. In our patient, the effect of thalamotomy may have been the result of removal of such aberrant neuronal sprouting, leading to the relief of hemidystonia.

CHENG-YANG CHHANG CHIN-SONG LU
Department of Neurology,
Chang Gung Medical College Hospital,
Taipei, Taiwan

Correspondence to: Chin-Song Lu, Department of Neurology, Chang Gung Medical College Hospital, 199 Tung Hwa North Road, Taipei, Taiwan.


Magnetic resonance imaging in advanced leukodystrophy presenting as spino cerebellar degeneration

Magnetic resonance imaging of a patient with advanced leukodystrophy presenting as spino cerebellar degeneration, showed symmetrical lesions in the corticospinal tract and cerebellar white matter adjacent to the dentate nuclei. These findings differentiate