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 assoication 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 hyperreflexic and the right plantar response extensor. Computed tomography (CT) showed a left thalamic haemorrhage 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 ankle, and clawed toes. 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 1986, the patient's dystonic right hand started to imitate involuntarily simple movements that were performed by the left hand, such as waving his 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.

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 bactofen, 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 his hemidystonia, blepharospasm, hemidystonia, imitation synkinesia and in the burning paraesthesia in the right limbs. The mild hemiparesis and hemianesthesia persisted. 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 in the stereotactic thalamotomy. These two observations may indicate that hemidystonia and imitation synkinesia share a common pathogenesis.

Disorganisation of the lemniscal system at any level may be responsible for imitation synkinesia. This might be a plausible explanation for the appearance of imitation synkinesia on the right hand with ipsilateral hemianesthesia in our patient post-thalamic haemorrhage. However, it is difficult to explain the distinct improvement of imitation synkinesia after stereotactic thalamotomy.

Burke et al has 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.

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Magnetic resonance imaging in advanced leukoencephalopathy presenting as spino-cerebellar degeneration

Magnetic resonance imaging of a patient with advanced leukencephalopathy presenting as spino-cerebellar degeneration, showed symmetrical lesions in the corticospinal tract and cerebellar white matter adjacent to the dentate nuclei. These findings differentiate...
Adrenoleukodystrophy (ALD) is a sex-linked recessive, peroxisomal disorder that causes diffuse demyelination of the central nervous system. ALD presenting as spinocerebellar degeneration (SCD), a variant form of ALD, has been recognized. The brain computed tomography (CT) finding most commonly seen in this form of ALD is atrophy of the brain stem and the cerebellum. However, MRI has not previously been reported.

A 29 year old man was admitted to our hospital because of gait disturbance. His parents were not consanguineous. His mother was asymptomatic though neurological examination revealed mild spasticity of both legs. At age 26, the patient complained of dysarthria and gait disturbance, and these symptoms were slowly progressive. Impotency, incontinence, and weight loss developed.

He was euphoric and mildly pigmented. Neurological examination revealed slow slurred speech, considerably increased deep tendon reflexes, extensor plantar responses and, ataxia of all limbs. His gait was markedly spastic and ataxic and he could not walk without support. His total IQ on Wechsler adult intelligence scale was 76.

The following laboratory data were normal or negative: full blood picture, electrolytes, liver function, kidney function, plasma glucose, creatine kinase, serological tests for syphilis, tests for SLE, anti-HIV antibody, anti-HTLV-I antibody, lymphocytes lysosomal enzyme activities including beta-galactosidase, beta-hexosaminidase and arylsulfatase A. Fasting plasma ACTH was 81 pg/ml (normal less than 65 pg/ml) while cortisol was normal. Serum cortisol response to ACTH (rapid ACTH test) showed a low response pattern. Cerebrospinal fluid was normal except for a raised protein (61 mg/dl).

Very long-chain fatty acids in plasma sphingomyelin fraction were considerably increased, that is C24:0/C22:0 was 1.171 [normal (SD), 0.735 (0.131)], C25:0/C22:0, 0.0380 [0.0143 (0.0039)], C26:0/C22:0, 0.0199 [0.0057 (0.0017)]. Brain CT showed atrophy of the cerebellum and the brain stem. MRI revealed abnormal intensity areas in both corona radiata, internal capsules, cerebral peduncles and white matter around the dentate nuclei of the cerebellum (fig 1) in addition to atrophy of the cerebellum and the brain stem (fig 2).

Pathological findings for ALD presenting as SCD include diffuse demyelination of the cerebellar white matter and the brain stem involving the internal capsule, corpus callosum and optic nerves. Marked loss of Purkinje cells and neurons of the dentate nuclei, and lesions in superior cerebellar peduncles have also been reported. In this patient, MRI clearly showed lesions in the pyramidal tracts and cerebellar white matter adjacent to the dentate nuclei, consistent with the clinical findings. The cerebellar lesions seen in this patient also shows one of the common pathological findings in the classic type of ALD, because a discrete lesion involving the cerebellar white matter adjacent to the dentate nucleus was reported in five of 17 necropsy cases. MRI in the classic type ALD has shown diffuse lesions in the cerebral white matter including the pyramidal tract, and the auditory and visual pathways. Areas of abnormal intensity in the internal capsule have been reported in adrenomyeloneuropathy. Thus, ALD presenting as SCD and classic type ALD differ in extent of lesions though the basic abnormality in lipid metabolism is common to both.

The lesions in the cerebellar white matter and the pyramidal tracts may be responsible for the symptoms in this patient which CT failed to detect. A common CT finding in ALD presenting as SCD is atrophy of the brain stem and the cerebellum, which is indistinguishable from that in SCD. In SCD, however, MRI abnormality suggesting the existence of demyelination as seen in this patient, has not been reported. Thus patients with cerebellar ataxia should be examined with MRI for the possibility of ALD.

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