The rostral interstitial nucleus of the MLF (ri MLF) is thought to be the immediate pre-nuclear structure for generating vertical saccades and receives an ascending input from the PPRF. This nucleus, located in the rostral mesencephalic reticular formation, was almost certainly spared in our case since there was no clinical or radiological evidence of damage to that region; consciousness was never impaired and, although there was initially impairment of convergence, the absence of mydriasis, ptosis and vertical strabismus exclude significant III nerve nuclear or infranuclear involvement. The most likely explanation for the transient vertical gaze palsy in our patient is damage of the PPRF. However, if the PPRF were crucial for the generation of vertical eye movements in man, as it is in the monkey, an explanation is needed for the paucity of cases in the literature. Descending fibres from the frontal eye fields terminate in the mesencephalic and pontine reticular nuclei related to ocular-motor control. Thus, it might be postulated that the ri MLF can be activated from two sources: the first and normal source is the PPRF. Such a source enables integration to occur between the horizontal and vertical vector of any eye movements and is necessary for the development of oblique saccades which require simultaneous discharge from both mesencephalic and pontine reticular gaze centres. If the PPRF is damaged a second source is the direct descending pathway to the mesencephalic gaze centres, which could be re-weighted such that vertical saccades can again be generated. Although this is entirely speculative is is a possible explanation of the variability and transience of vertical gaze defects in pontine lesions. In this or any other explanation, however, the underlying assumption is that there may be differences in the control of vertical saccades between man and monkey.

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**References**


**Late onset posthemiplegic dystonia in systemic lupus erythematosus**

Sir: Delayed development of focal dystonic or athetoid movements following hemiplegia (late onset posthemiplegic dystonia) is rare but well documented. We report a case of late onset posthemiplegic dystonia in a patient with systemic lupus erythematosus (SLE). To our knowledge this association has not been previously reported.

At age 42 years, a female patient developed a left hemiparesis that subsequently resolved completely. A year later she was hospitalised in our Neurology Department because of right hemiparesis and speech disturbances. CT of the head demonstrated the presence of two hypodense nonenhancing lesions, one in the left basal ganglia and internal capsule area and a smaller one in the right lentiform nucleus. Sedimentation rate was 90 mm h and ANF positive with a titre 1:80. The rest of the laboratory tests, including extensive investigation for collagen vascular disease, were negative. Two years later she noticed abnormal movements involving the left sided extremities, which progressed to the point that she lost the ability to use her left hand and had difficulty in walking.

Neurological examination on admission revealed a low output speech with no other aphasic features and a right spastic hemiparesis. The left arm was held in adduction at the shoulder and flexion at the elbow, while the hand was in extension and ulnar deviation at the wrist. The fingers were in extension at the carpometacarpal joints and exhibited prolonged, sustained twisting movements, which increased during attempted use of the arm. The left leg was in extension at the knee, while the foot was in inversion and plantar extension. Deep tendon reflexes were brisk bilaterally but more so on the right. An extensor plantar response was elicited on the right. No sensory abnormalities were found.

A repeat CT scan of the head again demonstrated two hypodense lesions in the same areas of the basal ganglia. Other abnormal laboratory data included: sedimentation rate 135 mm/h, red cell count 2,600,000 mm³, white cell count 3,000 mm³ with a differential count of 62% neutrophils, 28% lymphocytes, 4% eosinophils and 6% monocytes; platelet count 30,000 mm³; BUN was 4.32 mmol urea/l, creatinine 114 2 umol/l, ANF positive (1:2560), anti-DNA positive, anti-ENA positive. Total serum complement was 16.2 kU/ml (normal: 35–45) with C3 9.2 SIU (normal: 100–250) and C4 21 SIU (normal: 20–65). VDRL and FTA were negative. LE cells were present.

The patient was subsequently treated with steroids. The blood picture improved but the neurological manifestations remained unchanged. Two months after her discharge she developed seizures.

In the present case the neurological symptoms preceded other clinical manifestations of SLE by 4 years. In the few previously reported cases choreoathetoid movements in SLE have had a rather acute onset and later subsided. However, in our patient the abnormal movements developed slowly and progressively and involved the left sided extremities only after the weakness had subsided. The movements included sustained athetoid twisting of the fingers and dystonic posturing of the more proximal parts of the involved extremities, which resembled those described in other cases of post-hemiplegic dystonia.

We consider that the low density lesion in the right putamen, apparently an infarct,
was responsible for the abnormal movements, as similar CT findings have been reported in other cases of hemidystonia.2-6
In the necropsied cases partial destruction of the caudate nucleus and the putamen has been described.1 2

Why there is a delay between the occurrence of the hemiplegia and the onset of the abnormal movements, which frequently appear after the weakness has improved, is unknown. It has been proposed that aberrant central nervous system sprouting subsequent to a non progressive cerebral insult is responsible for this delay.2 In the present case as well as in previous ones, the chronological progression with the onset of dystonic symptoms after the improvement of the hemiplegia, and subsequent worsening of the movements, would be in keeping with this hypothesis.

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References


Recovery from bilateral wrist-drop in Whipple’s disease

Sir. The neurological manifestations of Whipple’s disease are varied,1 and have been described in conjunction with characteristic histological features occurring in the central nervous system.2 The central element of the CNS involvement comprises dementia, ophthalmoplegia and myoclonus.3 Hypothalamic disturbance, meningitis and seizures have also been observed.4 5 These neurological complications have emerged despite successful treatment of the intestinal component of Whipple’s disease with tetracycline or penicillin.4 6 This report details the unusual occurrence of peripheral nervous system involvement in conjunction with the more common systemic and CNS features of Whipple’s disease.

A 59 year old male electrical engineer presented in June 1985 with lethargy, confusion, weight loss and ophthalmoplegia. He had a 9 year history of arthritis affecting the right knee, and for 12 months he had difficulty remembering details at work. Over this time he had become sexually impotent, and his family described personality change with anxiety, lethargy and frequent aggression. From April 1985 he suffered with intermittent diplopia which progressed to loss of all voluntary eye movements. He became intermittently confused and drowsy during the daytime with indistinct rambling speech. In the 3 months before admission he had also developed steatorrhea, and had lost 7 kg in weight.

On admission he complained of tiredness, and soreness of his eyes. His alcohol intake was minimal, and his only medication was slow-release indomethacin. Examination revealed pallor, mild generalised wasting, finger clubbing, axial lymphadenopathy and areas of hyperpigmentation on the lower legs and feet. His abdomen was distended and rectal examination revealed palpable greasy stools. There was a soft pansystolic murmur at the cardiac apex, and some basal crepitations in the lung-fields. A tense, tender effusion of the right knee joint caused limitation of knee movement. He was orientated in person, but not in time or place, with a poor short-term memory. His intellectual state fluctuated from mild disorientation to confusion, and he had episodes of daytime hypersomnolence and absence. He suffered bilateral conjunctivitis and filamentous keratitis, but there was no iritis. His pupils reacted sluggishly to light, and his visual fields were full to confrontation. The eyes were fixed in forward gaze; no accommodation response to convergence was possible, and pursuit and voluntary eye movements were absent. There were no doll’s eye movements or optokinetic nystagmus. He had an intermittent convergent spasm affecting the left eye, bilateral ptosis and prominent left facial myoclonus. Mild dysarthria was noted with limitation of tongue and palatal movement. Peripheral tone, power and sensation were initially normal.

Abnormal laboratory findings included: microcytosis, hypochromic anaemia (Hb 7 g/dl), with low serum iron and TIBC. Serum albumin was also low (30 g/l), and serum electrophoresis showed diffuse elevation of immunoglobulins. ESR (Westergren) was raised to 110 mm/h. Normal findings included syphilis serology, serum thyroxine, TSH and testosterone, vitamins E, B12, folic acid, and red cell lead and transferrin.

Malabsorption was confirmed with duodenal fat estimation of 23 g (normal below 5 g) and an undetectable urinary excretion of xylose in 5 hours after a 5 g dose. Gastroscopy revealed severe erosive oesophagitis, and barium studies of the small intestine demonstrated thickened mucosal folds in the distal ileum. Histology of an axillary node showed reactive changes only, and there was no evidence of amyloid on rectal biopsy. Liver biopsy, performed at laparoscopy, showed non-specific hepatitis with a lymphocytic infiltrate of periodic acid Schiff (PAS) positive Kupffer-like cells in the portal chyma. Jejunal Watson capsule biopsy was performed and confirmed Whipple’s disease with numerous PAS-positive macrophages in the lamina propria. Electron microscopy identified rod-shaped organisms within these macrophages. The CSF was acellular with a protein of 500 mg/l (normal 250-450) and glucose 3.8 mmol/l, with negative VDRL assay. An EEG showed a generally low voltage recording with little alpha activity of low amplitude and low-voltage slow waves in the temporal regions. CT head scan revealed generalised atrophy with dilatation of the lateral ventricles and widening of sulci. Assessment with the Weschler Adult Intelligence Scale (WAIS) gave a verbal IQ of 123, but a poor non-verbal memory and digit span.

The ophthalmoplegia was not responsive to IV edrophonium, nor to group B vitamins and vitamin E taken during the course of investigations. Before the institution of specific therapy, physical examination revealed progression of muscle wasting with proximal muscle weakness. Daytime hypersomnolence and confusion persisted with intermittent polydipsia and hyperphagia.

Six weeks after presentation, bilateral wrist-drop became apparent with quick abrupt onset of severe weakness occurring in the right and then left forearms. He neither...