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Familial writer's cramp

Sir: The aetiology of writer's cramp is still controversial.¹⁻⁴ In spite of the facts that favour an organic origin, genetic influence on this anomaly has not been demonstrated. We report a patient with writer's cramp, whose father had presented the same disorder 35 years previously.

The propositus was a 41-year-old male right-handed lawyer. He had a previous history of hyperuricaemia and gout treated with allopurinol, 100 mg/day. He had not received any other specific treatment before, was not exposed to contact with toxic substances, did not consume illegal drugs and had not suffered any psychiatric or neurological illness. There was no consanguinity between his parents who did not belong to a particular ethnic group. The patient had had postural tremor since youth. At the age of 33 years he suffered epicondylitis in the right elbow which remitted with local therapy. Since then, he had experienced difficulty in writing with the right hand, that adopted an abnormal posture such that it attracted the attention of other persons. The disturbance was

accompanied by a feeling of stiffness and pain in the forearm and wrist. Any other manual activity was carried out normally. The difficulty in writing had remained stable since the onset of the symptoms but its intensity varies at times and is influenced by the emotional state of the patient. On examination, moderate obesity and very light postural tremor in upper limbs were noted; in addition, there was a minimal tremor in the finger-nose-finger test. His writing was legible, but he wrote with moderate difficulty, with the wrist tending to extension from the start, while the fingers bent together clutching the pen. The pressure on the paper was slightly increased. There was no micrographia. These signs lasted as long as writing continued. The remainder of the examination was normal. Routine haematological and biochemical studies, uric acid, LE phenomenon, rheumatoid factor, radiographs of the chest and skull, ECG, brain high-resolution CT with contrast, ceruloplasmin and cornea were normal.

The father of the propositus is a 70-year-old retired lawyer, right-handed. Since the age of 35 he had experienced a non-progressive severe difficulty in writing, consisting of abnormal posture of the hand, inability to maintain the pen in the correct position, involuntary extension of the index finger ("it escapes"), and tremor. All these symptoms were present only during writing or detailed drawing. The resulting script was almost illegible, and the difficulty in writing so marked that he has used a typewriter from the onset of trouble. He has practised artistic painting without problems except for delicate drawings, when aid with the left hand becomes necessary. For 2-3 years before consultation, he has had cephalic tremor and postural tremor in upper limbs. He had no other symptoms. Examination revealed slight head tremor, mild postural tremor in upper limbs and very slight tremor on finger-nose testing. A moderate increase in tone in wrists, a little more marked on the right, with contralateral activity was noted. Writing and drawing were very difficult: from the start, the right wrist was placed in extension and ulnar deviation, with its ventral aspect on the table and fingers forced in flexion. While writing, the posture worsened, a remarkable tremor was superimposed and irregular jerks separated the index finger; the pen became sustained between thumb, middle and ring fingers and, eventually, escaped. When the attempt to write ceased all these signs disappeared. The remainder of the examination was normal, as was

routine analytical and radiological tests. The patient refused specific studies.

These patients were considered to suffer from simple writer's cramp.^{1,5} According to Marsden,⁵ even simple writer's cramp is a "minor" form of dystonia. Occasionally, some patients with writer's cramp provide data suggestive of familial involvement,^{1,6} but there is no clear evidence of genetic factors in most patients with writer's cramp.^{1,7} Writer's cramp was obviously familial in our cases, as in other variants of focal dystonia.^{8,9} This observation favours an organic origin of the disorder.

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Asterixis due to pontine haemorrhage

Sir: Asterixis is a common sign of metabolic encephalopathy and arises from various causes.^{1,2} This involuntary movement has also been observed in a focal brain lesion.³⁻⁷ In this letter, we describe a patient with asterixis due to pontine haemorrhage.

A 65-year-old woman was admitted because of left motor weakness and dip-

lopia on lateral gaze. There had been no history of hepatic, renal or pulmonary disease. The patient denied habitual use of any kind of drugs. The initial examination revealed a blood pressure of 200/100 mm Hg and slightly confused mentation. Neurological examination showed a mild left motor weakness of the upper and lower extremities and left hemisensory loss of all modalities. Pupils were isocoric and the light reaction was prompt and complete. Optic disks were normal. Eye movements were full on up and downward gaze. However, there were a mild left hemifacial sensory deficit deviation of the jaw to the right when the mouth opened, bilateral abduction palsy, mild left facial motor weakness and horizontal rotatory nystagmus on lateral gaze to left and right. Deep tendon reflexes were hyperreactive on both sides. Planter response was extensor bilaterally. Sensory testing revealed severe left hemisensory disturbance of all modalities. There was a marked swing of outstretched left arm and severe ataxia on finger-to-nose-testing of the left arm. A CT scan revealed a circumscribed high density area in the midpons (fig). Electroencephalo-

grams showed symmetrical 8-9 Hz alpha activity over the occipital region with occasional diffuse dysrhythmic bursts of 0.5-1.0 ms duration.

Subsequent examination over the next several weeks disclosed improvement in both mental state and ocular movement, which was full in all directions. Diplopia and nystagmus disappeared. However, the mild left motor weakness, the severe left hemisensory loss and the severe ataxia on the left arm remained. Deep tendon reflexes were exaggerated on both sides and plantar response was equivocal bilaterally. There was prominent asterixis of hyperextended hands and fingers on both sides.

Electromyographic recordings were carried out and other electrophysiological studies including somatosensory evoked potentials (SEPs) were undertaken. Surface EMG recordings showed intermittent electrical cessations of ongoing EMG discharges of both the agonist and antagonist muscles simultaneously (asterixis). This asterixis was observed bilaterally. The SEP component following left median nerve stimulation was completely abolished sug-

gesting that the haemorrhagic lesion had interrupted the lemniscal sensory volleys in the right half of the pons. The components of the SEP following right median nerve stimulation were detectable with normal latencies and amplitudes suggesting that the lemniscal sensory pathway remained intact.⁸ These findings were consistent with the CT scan finding and clinical sensory testing.

Several reports have appeared of asterixis due to focal brain lesions. Plum and Posner⁹ described a patient with right parietal empyema who showed asterixis. However, as their patient was stuporous, it could not be determined whether the disturbance of consciousness was due to mass effect of the empyema on the reticular formation of the brainstem caused asterixis, or to the focal lesion itself. On the other hand, the disturbance of consciousness of the patient which Tarsy *et al*³ reported was considered as a focal sign of disturbance of reticular formation of the midbrain. Other reported cases of asterixis due to a focal lesion suggest that asterixis could result from a dysfunction of sensorimotor integration occurring in the parietal lobe and the midbrain.⁴⁻⁷ Unilateral asterixis without disturbance of consciousness also has been reported. Young *et al*¹⁰ showed that asterixis could be produced by stereotactic thalamotomy. These reports and descriptions indicate that asterixis can be due to a focal lesion in the parietal lobe, thalamus, or midbrain. It is emphasised here that the circumscribed pontine lesion in our case did not extend to the midbrain or the thalamus. Asterixis has never been observed in a spinal lesion.

We thank Dr Naoyasu Motomura who kindly allowed us to examine this patient.

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Fig CT scan without contrast enhancement. A circumscribed high density area was seen at the right dorsal part of the pontine tegmentum.

Letters

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Correction

"Mitochondrial malic enzyme in Friedreich's ataxia: failure to demonstrate reduced activity in cultured fibroblasts" *J Neurol Neurosurg Psychiatry* Vol 48 Page 70-74.

Page 71—Methods

Column 2, Line 8 should read "The cells from a 175cm² flask were harvested, washed, frozen and thawed once in 20 µl of distilled water and sonicated on ice with two 20 second bursts"

Matters arising

Cerebral glucose utilisation in Parkinson's disease

Sir: We were interested in the paper by Rougemont *et al*¹ in which no alteration of local cerebral glucose utilisation was found between treated and non-treated Parkinsonian patients. However, the same parameter was found to be moderately increased in the basal ganglia of these patients compared to controls. In a recent study² one of us demonstrated that low concentrations of dopamine combined with insulin *in vitro* increased glucose transport in the isolated rat adipocytes. However high concentrations of dopamine combined with high insulin concentrations inhibited glucose transport. If this occurred *in vivo*, then alterations in dopaminergic function (for example decreased dopaminergic activity) could result in impaired glucose transport in neuronal cells. This would be in agreement with the findings by Lenzi *et al*³ who demonstrated decreased glucose metabolism in the parietal lobe of patients with hemi-Parkinsonism. Moreover Rougemont *et al*¹ demonstrated slightly increased glucose metabolism in the basal ganglia of Parkinsonian patients. This, we postulate, could result from reduction of dopamine content in these areas with resultant compensatory enhancement of insulin activity in these areas. It is thus possible that increased glucose utilisation in the basal-ganglia of Parkinsonian subjects could reflect impaired dopaminergic activity. The degree of the regional glucose utilisation could thus serve as a marker for loss of dopaminergic activity in these areas.

Dementia is a common associated symptom of Parkinson's disease.⁴ It is possible that by normalising glucose transport into the cortical cells which have been shown to have decreased utilisation in Alzheimer's type dementia, that the condition can be improved. This could possibly be achieved by administration of insulin, glucose and levodopa.

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Sympathetic skin response

Sir: Techniques for evoking the psychogalvanic response and determining conduction velocity along autonomic nerve fibres have long been available¹ but have met with limited interest in electroneurography. The simplicity of Shahani *et al*² technique of eliciting the sympathetic skin response makes it particularly suitable in the study of the autonomic nervous system during routine EMG sessions. In effect, psychogalvanic responses can be easily induced by any internal or external stimulus of sufficient "novelty": comparable sympathetic skin response in one hand can be obtained by electrical stimulation of the ipsi- or contralateral wrist, of the glabella and by a sudden auditory burst applied by earphones (fig). Thus, exploring several eliciting modalities of sympathetic skin response may have a localising value. The technique has however some drawbacks, which, if unrecognised, could result

Fig Sympathetic skin response evoked in the left hand by (a) stimulation of left median nerve; (b) stimulation of right median nerve; (c) stimulation at glabella; (d) auditory burst.