Letters

to report the patient originally referred by him, and Dr RCG Rowe for making available histological material.

References


Accepted 26 October 1987

A compulsive movement disorder with cavitation of caudate nucleus

Sir: The movement disorders include a number of conditions in which the patient is conscious of a compulsion to move. In some, such as akinesia, the movement appears to be a response to an unpleasant sensation, but in others like Gilles de la Tourette syndrome the compulsion less convincingly has such a provocation.

Laplane et al. reported three patients (two after an episode of carbon monoxide poisoning and one after coma following a wapping) who had CT scans showing bilateral cavitation in the basal ganglia who had psychic akiinesia and two of whom had a movement disorder. In their first case the patient showed stereotyped activities such as mental counting and this was sometimes accompanied by gesturing although it was difficult to tell if this was voluntary or involuntary. The patient also had chorea, tics and some Parkinsonism. Their third case had Parkinsonian signs. We wish to report a patient with cavitation in the region of the basal ganglia from an unknown cause who had a compulsion to move her hands simulating a typing exercise and who had no other form of movement disorder. She also displayed psychic akiniesia.

The patient is a 48-year-old lady who was employed as a manageress of a card shop but was forced to resign owing to the consequences of her current illness. Her present complaints began 2 years ago and coincided with her mother’s death; the patient thought that depression was the explanation for this illness. She had become apathetic, not rising till 1 p.m., and then watching television for the remainder of the day. She stopped doing any housework, shopping or social activities. She relinquished her previous interests, conversation became limited and so did her sense of humour. Her personal hygiene became poor and she had not had a bath for 18 months. One year previously she had begun to move her hands purposelessly. This was more obvious on the right than on the left. She could stop the movements happening for long periods if repeatedly asked to but if left to herself they would recur within a minute and persist. She said she found it comforting to move her fingers and described it as though she was practising typing exercises. The movements did correspond to the beginner’s exercise a s d f space bar, 1 k j space bar. In other words the movements consisted of flexion of her fingers towards the palm of her hand in repeated sequences running from the little to the first finger and then the thumb. Despite finding this activity comforting she thought it was “dreadful” that she did it. The movements would stop if she used her hands so they were not at all disabling. Apart from these movements and the impression of moderately impaired intellectual function neurological examination was normal. Psychometry showed on the Wechsler a verbal IQ of 96 and a performance IQ of 76.

The patient and her family denied any possible episode of carbon monoxide poisoning.

Her CT scan (fig) demonstrates cavitation in the region of both caudate nuclei also involving the putamen at least on the right. Haematological and biochemical screen was normal as were thyroid function tests. Treponemal serology and antinuclear factor were negative. Copper, caeruloplasmin and slit lamp examination of her cornea were all normal. CSF examination showed a normal protein with no cells. Serum and CSF pyruvate and lactate levels were within the normal range.

Her diagnosis is unclear. Of the known causes of cavitation in the region of the caudate nucleus carbon monoxide poisoning and Wilson’s disease have been excluded. Bilateral ischaemic lesions are possible. Leigh’s syndrome was considered but her CSF, pyruvate and lactate were normal.

The pathology is not germane to the more interesting phenomenon of an abnormal caudate and putamen in a patient with a compulsive movement disorder. We cannot prove that the lesions seen on CT scan are the cause of the movement disorder but as this part of the brain has so much to do with abnormal movements it seems likely that it is involved.

AC WILLIAMS
C OWEN
DA HEATH

Departments of Neurology and Medicine,
Queen Elizabeth Hospital,
Edgbaston,
Birmingham B15 2TH, UK
References


Accepted 22 September 1987

New onset choreiform disorder in an adult with recent Group A beta haemolytic streptococcal pharyngitis

Sir: Sydenham’s Chorea (acute chorea, St Vitus’ dance, chorea minor, rheumatic chorea) is a late manifestation of acute rheumatic fever occurring generally 1 to 6 months following an acute group A beta haemolytic streptococcal throat infection. About one third of patients with rheumatic fever develop chorea. The acute chorea is a disease of childhood; over 80% of the cases occur between the ages 5 and 15 years. Onset before age 5 year is rare and the occurrence of the first attack after 15 years is uncommon, except during pregnancy or related to the use of oral contraceptives in late teens or early twenties.\(^1\) Attacks generally subside within 2 to 6 months. Sydenham’s Chorea persisting late into adulthood is exceedingly uncommon. Recently, Gibbs et al\(^2\) reported on an elderly woman with a history of rheumatic chorea (“St Vitus’ Dance”) that persisted into and intensified late in adult life. She had a history of chronic rheumatic heart disease, but had no history of neuroleptic intake or family history of chorea. Recurrences many years after the initial attack are uncommon and suggest that late chorea may be due to reactivation by another mechanism, such as pregnancy (chorea gravidarum, especially in the prima gravidae) and drugs, such as phenytoin, levodopa, amphetamines, thyroid hormones, and oral contraceptives.\(^3,5\)

I report the case of a 71 year old man, with no prior history of rheumatic fever or chorea, who had the subacute onset of a bilateral choreiform disorder following a recent Group A beta haemolytic streptococcal pharyngitis.

The patient, a right handed white man, was well until he developed a Group A beta haemolytic streptococcal pharyngitis in August 1986, followed weeks later by the gradual onset of a bilateral choreiform disorder, involving his left side more than his right side. Head CT scan and EEG were unremarkable. He was treated with haloperidol (Haldol) 2 mg po qid and developed agitation. Haloperidol was discontinued and chlorpromazine (Thorazine) 50 mg po tid was started with some sedative effect and decrease in the choreiform movements.

The patient subsequently presented to the University Hospital in Stony Brook for further evaluation of his choreiform movements. There was no history of a prior choreiform disorder or rheumatic heart disease, nor was there a family history of a choreiform disorder. He had a history of hypertension treated with Dyazide since 1982, malaria, Dengue fever, and Bell’s palsy in 1980. Medications were Dyazide and chlorpromazine. He had smoked tobacco previously, but denied current tobacco or ethanol use. He denied allergies.

General examination was normal, without cardiac murmurs, carotid bruits, or peripheral stigmata of emboli. He was alert, oriented, and cooperative though somewhat agitated and garrulous, with intact speech and cognition. Cranial nerves, including fundoscopy, were normal. There were no otorhinolaryngologic symptoms. Strength was normal without pronator drift. There were severe left arm and left leg choreiform movements which abated with sleep. Deep tendon reflexes were slightly brisker on the left but were difficult to elicit reliably due to the choreiform movements. Plantar responses were flexor. There was no dysmetria or gait ataxia. Sensation was normal to all modalities except for mildly decreased vibratory sensation in the legs. Grasp was absent.

Studies included normal CBC, differential, electrolytes, creatinine, calcium, magnesium, ESR, liver function tests, PT/PTT, thyroid function tests, vitamin B12, Lyme titre, immune complex assay and C3 and C4 complement levels. There was mild proteinuria on urine analysis. Electrocardiogram showed normal sinus rhythm with incomplete right bundle branch block and left anterior fascicular block, axis about negative 45°. Throat culture was negative for beta streptococcus. Antistreptolysin O (ASLO) titre was positive at 800 (normal less than 100). C-reactive protein was elevated at 1-2 (0-08-0.8). Antinuclear antibody (ANA) and rheumatoid factor (RF) assays were negative. Serum copper, ceruloplasmin, and urine copper were normal. Cerebrospinal fluid was normal including oligoclonal banding, IgG index, myelin basic proteins, and cytology. Creatinine clearance was 52 ml/min (70-157) and urine protein concentration was 436 mg/day (0-150). Renal ultrasound showed no hydronephrosis but renal cortices were irregular, consistent with chronic renal disease. Echocardiogram was normal.

On admission, the chlorpromazine was increased to 75 mg po bid and 200 mg po qhs. During the first few days, he had a few transient episodes lasting less than 1 minute each, while sitting or standing, of a sudden loss of body tone, unresponsiveness to auditory and tactile stimulation, occasional drooling, without any tonic-clonic activity or incontinence. There was no consistent change in vital signs or EKG associated with these transient episodes. He was not orthostatic. Electrolytes including glucose were normal. There were no neurologic sequelae. Chlorpromazine was discontinued and a 48 hour electroencephalogram was normal. However there were no reported similar episodes during this period. Head CT with and without intravenous contrast and head MRI revealed only mild cortical atrophy without any focal lesions.

He was maintained off chlorpromazine without any further agitation. This elderly man had the gradual onset of transient, initially bilateral choreiform disorder manifesting weeks after a known Group A beta haemolytic streptococcal pharyngitis, supported by a positive antistreptolysin O titre and elevated C-reactive protein. Chorea is an uncommon movement disorder and is seen in relatively few conditions, including Huntington’s disease, systemic lupus erythematosus,\(^6\) polyarteritis,\(^7\) chronic alcoholism, Wilson’s disease (hepatolenticular degeneration), chorea-achanthocytosis, haemorrhagic and nonhaemorrhagic infarcts of either the caudate or the putamen, or both,\(^8,9\) and with epidural, extradural, and subdural haematomas.\(^10,11\) Paroxysmal chorea may also be associated with hypoglycaemia,\(^12\) acute carbon monoxide exposure,\(^14\) and possibly with migraine headaches.\(^15\) Tardive dyskinesia may appear after months to years of chronic neuroleptic (antipsychotic) drug therapy and is characterised by rapid, repetitive stereotype movements, as well as orobuccolingual dyskinesia, akathisia (motor restlessness), body rocking movements, and marching in place.\(^16\) Hemibalismus is a rare disorder generally caused by haemorrhagic or ischaemic lesions in the subthalamic nucleus and is characterised by the acute onset of violent involuntary movements of one half of the body, but it may present with hemichorea rather than with full blown ballistic
A compulsive movement disorder with cavitation of caudate nucleus.
A C Williams, C Owen and D A Heath

*J Neurol Neurosurg Psychiatry* 1988 51: 447-448

doi: 10.1136/jnnp.51.3.447

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