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. Attacks generally subside within 2 to 6 months. Sydenham’s Chorea persisting late into adulthood is exceedingly uncommon. Recently, Gibbs et al. 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.

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 orolingual dyskiniesias. 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, polyarthria, chronic alcoholic, Wilson’s disease (hepatolenticular degeneration), chorea-achanthocytosis, haemorrhagic and nonhaemorrhagic infarcts of either the caudate or the putamen, or both, and with epidural, extradural, and subdural haematoma. Paroxysmal chorea may also be associated with hypoglycaemia, acute carbon monoxide exposure, and possibly with migraine headaches. Tardive dyskinesia may appear after months to years of chronic neuroleptic (antipsychotic) drug therapy and is characterised by rapid, repetitive stereotypic movements, as well as orobuccolingual dyskinesia, akathisia (motor restlessness), body rocking movements, and marching in place. Hemiballismus 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.
movements. In considering the possible aetiologies of this patient's chorea, there is no evidence to support any of the typically considered causes of chorea in an adult, such as Huntington's disease, systemic lupus erythematosus, polycythaemia, cerebral infarcts or haematomas, Wilson's disease, recent neuroleptic ingestion or other iatrogenic, metabolic, or toxic causes. Hemiballismus, with its typically abrupt onset and unilateral features, is unlikely, especially in view of the absence of focal findings on the MRI scan. There was also no history of a previous choreiform disorder or of rheumatic heart disease. The temporal association of the transient choreiform disorder and the recent Group A beta haemolytic streptococcal pharyngitis reported in this patient suggests that this may be the first reported case of Sydenham's chorea beginning in late adulthood.

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References

Rapid development of basal ganglia calcification caused by anoxia

Sir: Basal ganglia calcification occurs in a number of conditions. It is not known as to how long it takes for the calcification to occur. We recently encountered a patient in whom basal ganglia calcification was demonstrated on sequential computed tomography (CT) scan after only 9 days.

A 58-year-old woman had a history of diabetes mellitus for 23 years for which she was treated with insulin. She had an operation for cataract under local anaesthesia. Two days after the operation, she developed sudden onset of dyspnoea and orthopnoea, and was referred to our hospital. On physical examination her temperature was 36.4°C, blood pressure was 118/64 mmHg, and respiratory rate was 40 breath/min. She was intubated, and ventilatory support was begun. Chest examination revealed dullness to percussion and diminished breath sounds at the base of the left chest. There were bronchovesicular breath sounds in the bilateral lower chest. Abdomen was normal. On neurological examination, she was stuporous, and with intermittent spontaneous movements in all her extremities. The pupils were 3 mm in diameter, equal, and reactive. Full doll's eye responses were obtained both horizontally and vertically. She had intact gag and ciliospinal reflexes. Deep tendon reflexes were brisk bilaterally with absence of ankle jerks and extensor plantar responses. Arterial blood gas levels (room air) revealed a PaO2 of 49 mmHg, PaCO2 38 mmHg; pH 7.16 a repeat blood gas determination while the patient breathed 6l/min nasal oxygen revealed a PaO2 54 mmHg. The white blood cell count was 6400/mm3. Blood sugar was 86 mg/dl. Serum Ca, P, Mg, Al-p, and parathyroid hormone levels were normal.

A chest radiograph revealed massive enlargement of the cardiac silhouette and pulmonary oedema.

Next day, the arterial blood gas levels continued to show remarkable hypoaxemia and metabolic acidosis. She was comatose and quadriparetic, but remained responsive to noxious stimuli and could say a few words. The pupils were motic with light responses. Intermittent spontaneous movements in the extremities persisted and plantar responses were extensor bilaterally. It was noted for the first time that if her neck was laterally rotated she developed a sudden jerking flexion of both shoulders and extension of the arms at the elbows. The movement lasted less than 1 second. The EEG was isoelectric. Later that day, she developed a decerebrate posture; other neurological signs being unchanged, one could still produce movement of the arm with neck rotation. There was occasional movement of the neck towards extension when it was passively and suddenly flexed. The movement varied from moment to moment with neck motion. During that time, she sustained two cardiopulmonary arrests with successful resuscitation. Intermittent spontaneous movement and decerebrate posture with an occasional few words of spontaneous speech persisted unchanged until 11 days after the onset, when the blood pressure progressively failed and the heart stopped. Necropsy was not performed. An initial CT scan at admission was normal. A CT scan 9 days later showed extensive basal ganglia calcification (fig).

Microscopic basal ganglia calcification was independently reported by Virchow and Bamberger in 1855. Fritzche first described the radiographic appearance in 1935. In 1939, Eaton et al noted the association of calcification of the basal ganglia with hypoparathyroidism, and in 1944, Sprague et al reported its occurrence with pseudohypoparathyroidism. It has since been described in association with many pathological conditions. In our case, it is likely that the underlying cause of basal ganglia calcification was anoxia.

The vulnerability of the basal ganglia, particularly of the globus pallidum, to anoxic injury has long been recognised. It is rare for the corpus striatum to suffer this type of anoxic injury without concomitant pallidal injury.

Our case suggests that basal ganglia are not only highly susceptible to anoxic injury and consequent dystrophic calcification, but also that basal ganglia calcification can...
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