Cerebellar ataxia in coeliac disease – no evidence of a humoral aetiology

Sir,

Many patients with dermatitis herpetiformis also have a small bowel enteropathy which responds to withdrawal of gluten from the diet. However, in this setting, coeliac disease is usually mild and rarely results in clinical malabsorption. Both disorders have a high association with HLA B8 and DR3 antigens and are suspected to have, at least in part, an immune aetiology. Some 

10 of patients with coeliac disease develop neurological complications (box). Cook1 compiled a series of 16 patients and drew attention to the fact that these were the same neurological disorders that occur in paraneoplastic disease.

Paraneoplastic cerebellar ataxia is most frequently associated with small cell lung cancer or carcinoma of the breast or ovary. Patients with breast or ovarian carcinoma have high titres of an IgG directed against a cytoplasmic antigen in cerebellar Purkinje cells.2 Whether this antibody is involved in the pathogenesis of the disorder or simply a phenomenon is not yet clear. The aetiology of cerebellar ataxia found with coeliac disease is equally obscure although vitamin deficiency, particularly of vitamin E, does not appear to be relevant.1 We were interested to look for the presence of an anti-Purkinje cell antibody in a patient with coeliac disease and anti-Purkinje cell antibody in a patient with coeliac disease and cerebellar ataxia.

A patient was a 65-year-old caucasian woman who presented with a 12-month history of progressive unsteadiness of gait and slurring of speech. She had a past history of reversible airways disease, dermatitis herpetiformis and, two years prior to her neurological presentation, had been found to have coeliac disease. Despite having no symptoms of malabsorption, investigations had revealed an abnormal xylene tolerance test (1h plasma xylene 0.69 mmol/l; normal range 0.65–1.35 mmol/l), positive antigliadin and endomysial antibody and severe villous atrophy of the small bowel. She had been unable to tolerate a gluten-free diet. She smoked 10 cigarettes per day and she did not drink alcohol. At the time of the investigation she was taking Becotide and salbutamol inhalers. There was no family history of note.

General medical examination was normal. She had a cerebellar dysarthria, normal external ocular movements and an ataxia of limb and gait. Deep tendon reflexes were preserved, plantar responses flexor, and there were no sensory signs.

Investigations revealed normal haematological and biochemical indices, thyroid function, B12, folate and chest X-ray. Wasserman reaction and autoantibodies were negative. Antigliadin IgG and antietomy IgA antibody were positive. CT scan and the cerebrospinal fluid (CSF) xylene were normal in all respects; no oligoclonal bands were detected. Serum vitamin E and vitamin E/lipid ratio were normal. Using an indirect immunofluorescence technique serum and CSF were screened for activity against human and rat cerebellar Purkinje cells and dorsal root ganglia neurones. No activity was found. Normal controls, sera from patients with other neurological diseases, and positive controls were also examined.

In Cook’s original series1 of 16 patients, three were found to have ataxia. Most also had a polyneuropathy so it is difficult to be certain whether the gait disturbance was entirely cerebellar. However, the majority of post-mortem examinations in this and other series2 indicate a demyelinating cell loss and a variable depopulation of the neurones in the granular layer and dentate nuclei. Subsequent reports3–4 whilst confirming the clinical association, have failed to cast any light on the aetiology. A dysimmune hypothesis is attractive – in both coeliac and paraneoplastic disease there is an increased incidence of B8 and DR3 antigens, and several syndromes may co-exist in the same patient. Some disorders occur in both autoimmune disease and in association with malignancy, eg, Lambert–Eaton myasthenic syndrome (LEMS). In LEMS the antibody has been shown to be responsible for the clinical syndrome.

At least in this patient we have failed to demonstrate any anti-Purkinje cell activity and a humoral aetiology remains unproven.

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We thank Dr JB Pilling for his permission to report this case.


5 Kinney HC, Burger PC, Hurwitz BJ, Hijmans JC, Grant JP. Degeneration of the central ner-


Delayed severe rhombomylolysis after taking 'ecstasy'

Sir,

A previously fit 36-year-old man was admitted in the early hours of New Year’s Day following a convulsion at a night club. According to friends he had taken "kush" Ecstasy tablet, two bottles of beer and a large quantity of water during the night. On admission his axial temperature was 36.2°C and his Glasgow coma score was 3/15. His pupils were equal and reactive to light and he had no focal neurological abnormalities.

Shortly after admission, he had two further convulsions, was incontinent of urine, vomited several times and became restless. Chest X-ray showed abnormality in the left lung field. Blood tests revealed a serum sodium concentration (Na+) of 115 mmol/l (normal 137–144 mmol/l) and a creatine kinase (CK) of 1572 IU/l (<250 IU/l); potassium, urea and creatinine were normal. His arterial pH was 7.5 with an oxygen saturation of 97%, on air. He also had a raised white cell count (14.6 x 10⁹/l) with a metaphilic leucocytosis. Clotting studies and liver function tests were normal. Further tests revealed serum 3,4-methyleneidioxymethylamphetamine (MDMA) levels of 0.013 mg/l with both MDMA and its metabolite 3,4- methylenedioxymethylamphetamine in his urine. Treatment was with diazepam and chlorpromazine for the fits and agitation and co-amoxycilav and hydrocortisone for a presumed aspiration pneumonia. Intensive ravenous hydration was commenced with 11 l of normal saline over 12 h, and a nasogastric tube passed.

Twelve hours after admission the patient remained comatose. His respiratory rate had risen to 32/min, his oxygen saturation had dropped to 90%, on air and he had developed an axillary pyrexia of 39°C. He had also become polyuric, passing 5 l of urine since admission. Repeat biochemistry showed a serum Na+ of 120 mmol/l with a CK of 2461 IU/l. His plasma osmolality was 259 mmol/kg (275–285 mmol/kg) with a urine osmolality of 153 mmol/kg (50–1400 mmol/kg) and a urine Na+ of 26 mmol/l. CT brain scan was normal. He received oxygen, and paracetamol for his pyrexia which peaked at 39.7°C (axilla).

Eighteen hours after admission his temperature started to fall and his level of consciousness began to improve. By 30 h after admission his serum Na+ was 132 mmol/l with a plasma osmolality of 278 mmol/kg. However his CK had risen to 81900 IU/l with an alanine aminotransferase (ALT) concentration of 132 IU/l (<34 IU/l). He had also become oliguric – passing 250 ml of urine in the previous hours. His urine tested positive for myoglobin.

He was treated aggressively with a forced alkaline diuresis induced by intravenous

Neurological complications of coeliac disease

- peripheral neuropathy
- myelopathy
- brain stem encephalitis
- cerebellar ataxia
- Lambert–Eaton myasthenic syndrome

References

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