Choreoathetosis: a result of uncontrolled diabetes

We describe a case of a 90-year-old woman who presented with a one-week history of acute onset involuntary movements of the right arm and leg. She had a background of type 2 diabetes for 30 years, a cerebrovascular accident (CVA) in 1980 with no residual deficit, hypertension and renal impairment (GFR 21) Stage 4 CKD. Her drug history included atenolol, candesartan, felodipine, atorvastatin, aspirin and glidazide 160mg bd, but she had poor concordance.

On direct questioning, the patient revealed a few weeks’ history of osmotic symptoms, polyuria, polydipsia, and weight loss. On presentation she was orientated and alert, and had a Mini Mental State score of 10/10. She had a blood pressure of 120/60mmHg, pulse of 80bpm, and was in atrial fibrillation. Neurological examination showed involuntary choreathetoid writhing movements of the right arm and to lesser extent the right leg, along with tongue involvement. Tone was normal throughout, power 5/5 in left upper and lower limbs, with normal sensation and down-going planters. There was no cogwheeling or rigidity. Eye movements were normal, and there were no nystagmus or cerebellar signs.

In a woman of this age group the differential diagnosis would be a CVA, other possibilities being medication related, CNS infections, metabolic derangements and Huntington’s chorea. Our patient was not on any causative medication, there was no suggestion of infection or family history of Huntington’s and it would be a very late presentation.

She was hyperglycaemic (blood glucose [BG] 23mmol/L) and had renal impairment (urea 26.6mmol/L, creatinine 238μmol/L) but was not in diabetic ketoacidosis. We therefore started her on subcutaneous insulin and not on a sliding scale. As her BG reduced, her movements slowed down and also decreased in magnitude. Over the next 48 hours, her BG levels improved to 6–10mmol/L and her involuntary movements resolved. CT scan of the brain two days post admission showed no evidence of a cerebrovascular event.

A neurologist examined her and found no evidence of neurodegenerative disorders, structural lesion, vascular event, paraneoplastic syndrome, infection, or medication responsible for her chorea. In the absence of other metabolic abnormalities, it was concluded that this patient’s chorea was likely the result of her hyperglycaemia.

She continued to show good progress with physiotherapy and was discharged three days post admission. She needed long-term diabetes treatment and follow up as her HbA1c was 16.2% (154mmol/mol) previously having been 6.1% (43mmol/mol). She had regained control of movements and was seen again a month later; there were no further episodes.

Discussion

‘Chorea’ is a borrowed Latin word that derives from the Greek khoreia, a choral dance, with athetosis being derived from athetos (not fixed). Hemiballism-hemichorea designates a clinical spectrum of continuous, writhing, non-patterned, involuntary movements involving one side of the body.

Acute hyperglycaemia on a background of poor diabetic control can cause choreoathetoid movement, which is reversible. An association between hyperglycaemia and hemichorea was first described in 1960 and, by 1982, authors confirmed that the spectrum of focal neurologic deficits in hyperglycaemia may include choreoathetosis and ballism.1

Hyperglycaemia associated movement disorders and striatal hyperdensity usually occur in elderly patients with poorly controlled diabetes and are related to acute peaks in BG levels.

CT brain scan usually shows hyperdensity in the contralateral putamen and/or caudate. An MRI scan reveals abnormal hyperintensity on T1-weighted images in the acute stage.2

Proposed hypotheses for hyperglycaemia as a cause of choreoathetosis are: (a) hyperglycaemia and hyperosmolality induce mild ischaemia in the putamen via hypoperfusion, due to osmotic shifts; and (b) hyperglycaemia induces anaerobic metabolism which leads on to GABA depletion.3

An impairment of GABAergic or cholinergic neurons, which would normally inhibit dopaminergic activity in the nigrostriatal system, may explain this hyperactive state. One postulated mechanism leading to dysfunction of GABAergic neurons is hyperviscosity.1,4 Furthermore, cerebral hypoperfusion and a direct effect of hyperglycaemia on cerebral metabolism result in depletion of gamma-aminobutyric acid (GABA) in the corpus striatum.5 With reduced levels of striatal GABA, increased pallidal activity results in dyskinesia.6 The cause of hyperdensity is due to protein hydration inside the cytoplasm of swollen gemistocytes.1,3

One series reports that 74% of patients had complete resolution of chorea after a period ranging from one day to 10 months, with the large majority reaching a full recovery within six months. An alternative case series of 10 patients reports dramatic resolution of dyskinesia within days after control of hyperglycaemia in all but one patient. In 86% of those patients who had a follow-up MRI after improvement in their chorea, the high signal intensity basal ganglia lesion on T1-weighted MRI images had resolved.3

Hyperglycaemia is one easily treatable and completely reversible cause of choreoathetosis. Knowledge of this cause can prevent anxiety related to a cerebrovascular event and give patients and families a prognosis of improvement, as treatment of hyperglycaemia will revert the involuntary movement.

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References are available online at www.practicaldiabetes.com.
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