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Case Report

Hemichorea-hemiballismus in a patient with hyperglycaemic hyperosmolar state

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ABSTRACT

Hemichorea-hemiballismus (HCHB) is a rare manifestation of hyperglycaemic hyperosmolar state caused by contralateral lesion in basal ganglia. A 74-year-old, known diabetic and hypertensive woman presented with one-week history of high-grade fever and loss of consciousness associated with involuntary movement of the right upper and lower limbs for 10 hours prior to presentation. Physical examination revealed pyrexia, tachycardia and altered sensorium. Blood glucose was 53.8 mmol/l, hemoglobin A1c (Hb A1c) 9.9% and brain computed tomography (CT) scan showed cerebral atrophy with bilateral basal ganglia hyperdensities. Escherichia coli was cultured from the urine. She did well on treatment with soluble insulin, rehydration and intravenous ceftriaxone. HCHB is a rare complication seen in patients with poorly controlled diabetes mellitus. This report highlights the reversibility of the disease with prompt diagnosis and appropriate insulin treatment. HCHB should be distinguished from other intracranial pathologies.

Keywords: Hemichorea-hemiballismus, Hyperglycaemic hyperosmolar, Diabetes mellitus, Poor control, Basal ganglia

INTRODUCTION

Hemichorea-hemiballismus (HCHB) is a clinical spectrum that involves involuntary random and fast jerky motions in the distal parts of the limbs (chorea) and larger amplitude random and violent flinging or kicking involuntary movements mainly in the proximal joints (ballismus).[1,2] This condition is common in postmenopausal women.[3] HCHB can be caused by several disease conditions including central nervous system infections, human immunodeficiency virus infection, intracranial haemorrhage, neurodegenerative diseases, toxins, metabolic disorders and neoplasms.[4] Hyperglycaemic hyperosmolar state is an uncommon cause of HCHB, and predominantly affects elderly, postmenopausal females.[4] The condition may be related to an increased dopaminergic receptor sensitivity secondary to oestrogen deficiency.[5] It may complicate poorly controlled diabetes mellitus but may also be the first presentation of diabetes mellitus.

The aim of this report is to raise awareness and high index of suspicion, highlight the importance of differentiating this condition from intracranial disorders, and enhance prompt diagnosis and treatment.

CASE REPORT

A 74-year-old woman, a known diabetic and hypertensive of 2 years had been on oral antidiabetic drugs (metformin and glimepiride) but with poor compliance, presented with high-grade fever of one week duration and loss of consciousness associated with involuntary, abnormal movements of the right upper and lower limbs for 10 hours prior to presentation. There was no history suggestive of

similar illness in the past and no history of fall or trauma to the head.

Physical examination showed an elderly woman who was dehydrated and febrile (38.5oC). Respiratory rate was 32 cycles per minute, pulse rate 112 per minute, blood pressure 100/60 mm Hg and heart sounds were S1 and S2. She was unconscious with Glasgow coma scale (GCS) of 10/15, there was rapid, jerky and violent flinging movements of the head and right upper and lower limbs. Tendon reflexes were normal and plantar response was flexor. Urine culture yielded Escherichia Coli sensitive to ceftriaxone.

Brain computed tomography (CT) scan showed cerebral atrophy with bilateral basal ganglia hyperdensities (Figure 1). There was no cerebral oedema, mass effect or loss of volume. Echocardiogram was normal.

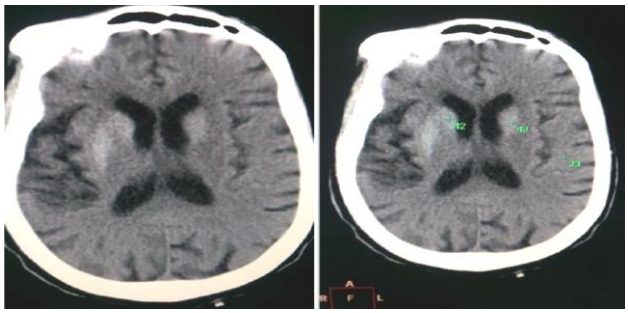

Figure 1: Brain CT scan showing cerebral atrophy and basal ganglia hyperdensities.

Table 1: Laboratory results.

Laboratory parameter	Intial result	2nd result	3rd result	4th result
Fasting blood glucose (4-7 mmol/l)	-	-	5.6	6.5
Random blood glucose (4-8 mmol/l)	53.8	20.8	6.6	-
Glycated haemoglobin (4-6%)	9.9	-	-	-
Sodium (235.0-145.0 mmol/l)	123	135	138	140
Potassium (3.5-5.0 mmol/l)	4.6	4.0	4.0	4.3
Chloride (97.0-107.0 mmol/l)	84	101	109	102
Bicarbonate (20.0-31.0 mmol/l)	18	20	24	23
Urea (1.7-8.3 mmol/l)	18.9	19.1	4.9	4.5
Creatinine (male: 80.0-115.0 umol/l, female 53.0-97.0 umol/l)	281	186	69	69
White blood cell (4.8-10.8×10^9/l)	19.4	-	10.2	6.7
Neutrophils (40-75%)	84	-	73	-
Lymphyocytes (20-45%)	16		21	
Monocytes (2-10%)	-		6	-
Eosinophils (1-6%)	-	-	-	-
Basophills	-	-	-	-
Red blood cell (4.5-5.5×10^{12}/l)	3.8	-	2.6	-
Haemoglobin (11.2-16.5 g/dl)	11.1	-	7.8	8.1
Packed cell volume (40-54 g/dl)	32	-	26	27
Mean corpuscular volume (75-100 fl)	85	96	-	-
Mean corpuscular haemoglobin (25-35 pg)	29	29	-	-
Platelets (140-400×10^9/l)	325	-	169	-
Total cholesterol (<200 mg/100 ml)	87	-	-	-
HDL cholesterol (>35 mg/100 ml)	21	-	-	-
LDL cholesterol(<130 mg/100 ml)	42	-	-	-
Triglycerides (<160 mg/100 ml)	119	-	-	-

Diagnosis of HCHB secondary to hyperglycaemic hyperosmolar state precipitated by urinary tract infection (UTI) was made. She was rehydrated with normal saline and successfully managed on insulin, antibiotics and anticoagulant. She regained consciousness on the 7th day of admission and tremors reduced, she was then placed on a low dose haloperidol. She had 3 sessions of physiotherapy per week with passive movement of limbs, effleurage massage, walking education and sitting and standing exercises. Random blood glucose was 6.5 mmol/l and tremor resolved completely by 47th day of admission. She was counselled on diabetes mellitus (DM) education, discharged home on subcutaneous insulin glargine and scheduled for follow up at the clinic.

DISCUSSION

The exact pathogenesis of hyperglycemic hyperosmolar-associated HCHB is not clear. Background metabolic conditions, including hyperglycemia, can have various neurological presentations. HCHB is a rare manifestation reported to occur with severe hyperglycaemia in hyperosmolar state and predominately affects elderly, postmenopausal female as seen in our 74 year old case.[4]

Hyperglycemia and/or metabolic acidosis produces a decrease in regional cerebral blood flow with maximal reduction in the basal ganglia resulting in ischaemia.[6] Hyperglycemia also shifts cellular energy demands towards anaerobic metabolism which causes increased metabolism of gamma-amino butyric acid (GABA) in the brain as an alternate energy source: this results in depletion of the GABA content in basal ganglia, decreasing inhibitory signals and causing involuntary movements.[6] In ketoacidotic state, acetoacetate may be used to synthesize GABA which may explain the rarity of this condition in patients with diabetic ketoacidosis and type 1 diabetes. Other proposed mechanisms are hyperviscosity related to hyperglycaemic hyperosmolar state induced GABAergic neuron dysfunction in the putamen.[6]

Hyperglycaemic hyperosmolar HCHB is diagnosed by its typical clinical presentation in the setting of hyperglycemia and neuro-imaging findings. Brain computed tomography (CT) scan shows areas of hyperdensity in the basal ganglia, as described in our patient. This hyperdensity is different from a haemorrhagic lesion due to the absence of mass effect, oedema or loss of brain volume as well as sparing of the internal capsule. Magnetic resonance imaging (MRI) findings are also characteristic for high signal intensity on T1 weighted images in the contralateral putamen.[7] These changes may persist despite resolution of clinical symptoms but would usually resolve.

The mainstay of treatment of HCHB associated with hyperglycaemic hyperosmolar state is prompt and aggressive glycaemic control. Clinical symptoms would usually resolve on normalization of blood glucose level. Some patients may, in addition, require monotherapy or combination therapy with benzodiazepines, neuroleptics, anti-epileptics and most recently reports have shown improvement with the use of topiramate.[8,9] Given the rarity of HCHB as a complication of hyperglycaemic hyperosmolar state, a high index of suspicion is necessary to ensure early diagnosis, prompt treatment and good outcome.

CONCLUSION

Hyperglycaemic hyperosmolar state is a rare but important differential diagnosis in patients with HCHB as prompt diagnosis and treatment of hyperglycaemia has an excellent prognosis.

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