CASE REPORT

Uncontrolled hyperglycaemia: a reversible cause of hemichorea–hemiballismus

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SUMMARY

Hyperosmolar hyperglycaemic state (HHS) an acute complication of diabetes mellitus, can be associated with neurological involvement ranging from seizures, involuntary movements to reversible focal neurological deficits without any structural lesions. We report a 71-year-old woman, a known case of type 2 diabetes mellitus who presented with the sudden onset of hemichorea–hemiballismus. On investigations she had hyperglycaemia and urinary tract infection. Achievement of euglycemia with insulin therapy resolved the involuntary movements completely. This highlights the fact that HHS is an uncommon but easily treatable cause of recent onset unilateral hemichorea–hemiballismus.

BACKGROUND

The major causes of unilateral hemichorea–hemiballismus include vascular insult (stroke), traumatic brain injury and neoplasms.[1] The present case reports a patient who presented with unilateral hemichorea–hemiballismus, which was reversed by the treatment of hyperglycaemia. Hyperglycaemia thus represents a reversible cause of unilateral hemichorea–hemiballismus and should be considered in each and every case presenting with hemichorea–hemiballismus as prompt treatment is curative. Usually unilateral manifestations are a sequel of structural/vascular pathology, whereas in this case it was due to a reversible metabolic cause.[1 2]

CASE PRESENTATION

A 71-year-old woman, presented with a history of osmotic symptoms since 10 days and the acute onset involuntary movements of right forearm, arm and fingers, of 2 days duration. The movements were jerky, irregular involving right forearm and arm with abnormal posturing of fingers. Initially the movements were of low amplitude and occurred every 2–3 h but progressed to occur four to five times every hour in last 4 h. They were not suppressed by activity and did not occur during sleep. The patient was conscious but bit confused. She had poorly controlled type 2 diabetes mellitus and hypertension since last 10 years. There was no history of trauma, fever or consumption of medications other than antidiabetics and antihypertensives. There was no past or family history of movement disorders.

On examination she was conscious and oriented. She was dehydrated, had a pulse 110/min and blood pressure 100/70 mm Hg. On neurological examination, the patient was conscious but bit confused (Glasgow Coma Scale 15/15, Mini-Memtal Status Examination 26/30). Asterexis were present. Abnormal involuntary movements suggestive of hemichorea and hemiballismus were present. Cranial nerve examination including the fundus was normal. There were no cerebellar signs. The sensorimotor examination in upper limbs was normal, but in lower limbs all modalities of sensations were diminished below the ankle joints. The deep tendon reflexes including the ankle were depressed and plantars were flexor. Other systemic examination was normal.

INVESTIGATIONS

Investigations on admission revealed a random blood glucose 650 mg/dL. Glycated haemoglobin was 13.5%. Her blood urea nitrogen and creatine were 30 and 2.2 mg/dL, respectively. Plasma osmolality was 336 mOsm/L, sodium 145 mEq/L, potassium 4.8 mEq/L. Arterial blood gas analysis showed pH was 7.41, partial pressure of oxygen=83.6 mm, partial pressure of carbon dioxide=36 and HCO_3=18 mEq/L. Haemoglobin was 12.1 gm/dL and total leucocyte count was 11 400/mm^3 with polymorphonuclear predominance. Serum calcium was 9.2 mg%. The liver function and thyroid function tests were normal. Urine routine showed urine glucose was 4+ and ketones were negative with field full of pus cells. Specific gravity was 1.020.

MRI of the brain performed 3 days later, once the patient was stable, showed no abnormality.

DIFFERENTIAL DIAGNOSIS

The major differential for hemichorea–hemiballismus include structural/vascular lesions such as stroke, neoplasms, granulomatous disorders, neurodegenerative disorders like amyotrophic lateral sclerosis and demyelinating disorders involving basal ganglia, as well as complication of HIV infection. These can be ruled out by neuroimaging, which should be performed irrespective of finding an underlying reversible metabolic cause such as hyperglycaemia, because metabolic cause might be unmasking-underlying structural problem.[1]

TREATMENT

The patient was started on intravenous fluids for rehydration followed by insulin infusion and antibiotics for urosepsis. Her blood sugar normalised over 6–8 h. Involuntary movements completely disappeared in 1 day. She had no involuntary movements during the rest of her hospital stay. Urine culture performed after 14 days of intravenous antibiotics was not showing any growth.

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OUTCOME AND FOLLOW-UP

The patient was shifted on subcutaneous insulin (basal bolus regimen glargine and actrapid) and discharged.

DISCUSSION

Chorea (irregular, unpredictable, brief jerky involuntary movements) and hemballismus (ballistic flinging movements) could result from the number of causes namely, central nervous system infections, neoplasm, intracranial haemorrhage, neurodegenerative disorders, drugs, toxins, systemic disorders and metabolic imbalance.[1]

Hyperglycaemic state (HHS) associated chorea was first described by Bedwell in 1960.[1] In an analysis of 53 cases mean age was 71 years and female-to-male ratio was 1.8 : 1. A majority of patients were Asian women suggestive of a genetic predisposition. Postmenopausal women are predominantly affected and this is related to increased dopamine receptor sensitivity secondary to oestrogen deficiency.[2] Our patient is also an elderly Asian woman with poorly controlled type 2 diabetes mellitus. Chorea can complicate long-standing type 1 or 2 diabetes or can be the presenting symptom of the new onset diabetes.[3]

The classic MRI findings described are hyperintensities of contralateral basal ganglia on T1-weighted images and corresponding hypointensities on T-weighted images, multiple abnormal signals in basal ganglia and invariable involvement of the putamen. CT scan may show hyperdensities in the aforementioned areas.[1 4]

The exact pathogenesis of HHS-associated hemichorea–hemiballismus is not clear. Hyperglycaemia and/or metabolic acidosis produces a decrease in regional cerebral blood flow with maximal reduction in basal ganglia resulting in ischaemia.[5] Hyperglycaemia 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 resulting in depletion of the GABA content in basal ganglia, decreasing inhibitory signals and causing involuntary movements.[5] In ketoacidotic state acetoacetate may be used to synthesise GABA which may explain the rarity of this condition in patients with diabetic ketocidosis and type 1 diabetes. Other proposed mechanisms were hyperviscosity related to HHS state induced GABAergic neuron dysfunction in putamen.[5]

HHS-associated chorea has an excellent prognosis. Only tight blood glucose is sufficient to treat chorea in some cases, as in our case. Other patients may require monotherapy or combination therapy with benzodiazepines, neuroleptics and antiepileptics.[6]

CONCLUSION

HHS is an uncommon but easily-treatable cause of hemichorea and hemiballismus. Checking blood glucose is obligatory whenever an elderly patient presents with the new onset hyperkinesias as hydration and blood glucose control can lead to a rapid and complete recovery.

Learning points

- Whenever a patient presents with the new onset unilateral hemichorea–hemiballismus, apart from considering vascular aetiology one needs to consider metabolic causes of which hyperglycaemia is an important reversible cause.
- Hemichorea–hemiballismus can complicate long-standing type 1 or 2 diabetes or can be the presenting symptom of the new onset diabetes.
- Testing blood sugars as a part of work-up is important as normalisation of glucose may lead to rapid and complete resolution of hemichorea–hemiballismus.

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