CASE REPORT

Haemorrhagic stroke or hyperglycaemia?

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SUMMARY

A 56-year-old woman with no medical history was admitted with acute onset hemiballistic-choreiform movements of right extremity. On admission, her serum glucose and osmolality were elevated. Her glycosylated haemoglobin (HbA1c) was 17.9 with average blood sugars of 467 mg/dL. A CT scan of the brain revealed unilateral hyperintensities in the basal ganglia. A complete stroke work-up, including MRI and MR angiography of brain was otherwise unrevealing. Subcutaneous insulin was instituted, which led to complete resolution of her symptoms within 48 h of hospital admission. She was readmitted 4 weeks after her initial discharge for similar, but less severe, symptoms. This time her HbA1c was 13.9 with an average blood sugar of 352 mg/dL. A repeat MRI demonstrated a persistent abnormal signal within the left basal ganglia without infarct. She was started on subcutaneous insulin. Her symptoms improved but did not resolve. Haloperidol and gabapentin were initiated and she was again discharged after stabilisation to a rehabilitation centre as per physiotherapy recommendations.

BACKGROUND

Hemichorea-hemiballismus consists of a neurological syndrome characterised by violent proximal involuntary movements on one side of the body.^{1 2} A focal vascular lesion in the contralateral basal ganglia is the most common cause of this dyskinesia. This can be a rare presenting sign of long-standing poorly controlled diabetes. Recognition of this unique presentation of uncontrolled diabetes is important because correction of the underlying hyperglycaemia may lead to rapid and improvement in many cases.

CASE PRESENTATION

A 56-year-old Haitian woman with no medical history presented to the hospital with acute onset of choreiform movements of her right upper and right lower extremities for 3 days that were associated with both right-sided weakness and numbness along with 2 weeks of severe headache. The patient noted weight loss, polyuria, polydipsia, blurry vision and altered sensations in her feet for upto 2 years prior to admission. She denied previous motor disorders or symptoms. Her weight had remained stable despite an increased appetite. There was no history of environmental or occupational toxin exposures. She was not on any herbal or prescription medications. Family history was negative for movement disorders. On neurological examination, she had a Glasgow Coma Score of 15. She had uncontrolled choreatic-type

movements of the right arm and leg. She was found to have limited vision, a positive pronator drift on the right side. Initial stroke workup revealed a CT scan of the brain consistent with left caudate nucleus haemorrhage. Initial fingerstick reading was undetectably high (>600 mg/dL).

INVESTIGATIONS

Further studies revealed non-ketotic hyperglycaemia with a serum glucose of 515 mg/dL, with a correlative glycosylated haemoglobin (HbA1c) of 17.9 (4% and 5.6% units, normal). Serum osmolality was high. Sodium was 125 mEq/L, potassium 4.1 mEq/L, chloride 90 mEq/L, CO₂ 26.9 mEq/L and anion gap of 8 mEq/L. Her thyroid stimulating hormone was 1.060 µIU/mL and free T4 levels were 0.90 mg/dL. aspartate aminotransferase was 23 IU/L, alanine transaminase of 18 IU/L and alkaline phosphatase was 74 IU/L. CT of the head showed a unilateral diffuse hyperdensity in the left caudate and lentiform nuclei. Also noted was a focal decreased density in the anterior limb of the left internal capsule that was initially felt to be an internal capsule haemorrhagic stroke. However, MRI showed a mildly bright signal within the left basal ganglia with normal diffusion weighted sequence, which is non-suggestive of acute stroke (figures 1 and 2).

Figure 1 Initial CT of head: (black arrow) hyperdensity is seen centred within the left caudate head and left lentiform nucleus. Small focal hypodensities are seen within the left caudate head and within the anterior limb of the left internal capsule. There is no evidence of an intra-axial mass lesion. There is preservation of the grey-white matter differentiation without findings of acute territorial infarction. There are no areas of acute haemorrhage.

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Figures 2 (A and B) Initial MRI of head—T1 and T2 fluid attenuation inversion recovery, respectively. Diffusion weighted sequence demonstrates mildly bright signal within the left basal ganglia (black arrow) without definitive decreased signal on the antibody drug conjugate trees sequence suggesting of T2 shine through. Additionally, this area demonstrates abnormal bright T2 foci. Otherwise, the diffusion weighted sequence is normal without evidence to suggest acute stroke. There is spontaneously bright T1 signal within the caudate head and basal ganglia on the left (black arrow).

DIFFERENTIAL DIAGNOSIS

Stroke was initially high on the differential diagnosis for this patient, considering her focal neurological changes and cerebrovascular risks. Other diagnosis to consider includes thyroid disorder, either hypothyroidism or hyperthyroidism. In addition, senile chorea needs to be considered, but can be differentiated from hemiballismus by its insidious onset and bilateral presentation. Liver failure can also produce similar motor findings but involvement is usually bilateral with gross derangement in liver function tests. Infectious aetiologies need to be considered in the presence of fever and positive inflammatory markers, including AIDS in which involuntary movements have been increasingly recognised. Postinfective autoimmune central nervous system disorders including poststreptococcal autoimmune disorder may have similar presentations. Drugs, toxins and malignancy can lead to similar abnormal movements and their respective screenings should be obtained to rule them out. Finally, genetic diseases may be considered and can include Wilson's disease, Huntington's chorea, Friedreich's ataxia and systemic lupus erythematosus (box 1).

Box 1

Differential diagnosis
Cerebrovascular accident
Drugs/toxins
Genetic diseases such as Huntington's disease, Friedreich's ataxia etc
Haematological abnormalities such as neuroacanthocytosis
Infectious processes such as AIDS, toxoplasmosis, poststreptococcal autoimmune disorder etc.
Liver failure
Malignancy
Senile chorea
Thyroid disorders—hypothyroidism or hyperthyroidism
Vascular disorders such as systemic lupus erythematosus
This list is not fully encompassing.

TREATMENT

She was started on an aggressive insulin regimen and had complete resolution of involuntary movements within 48 h.

OUTCOME AND FOLLOW-UP

Twenty-five days after the first admission, the patient returned with right-sided chorea-like movements despite fairly tight glucose control. We believe that the patient was not compliant with insulin at home and may have led to worsening of symptoms. During her second admission for similar symptoms, her HbA1c was 13.9 in 23 days of insulin initiation. A repeat MRI was performed demonstrating a persistent abnormal signal within the left basal ganglia without evidence of acute vasculature for infarct. Insulin was continued but symptoms did not resolve completely (figure 3). This time neurology initiated haloperidol and neurontin for her abnormal movements. Physical medicine and rehabilitation also provided a treatment plan to return the patient to her baseline functional capacity. She had a significant improvement of symptoms with a decrease in choreiform movements. She was again discharged after stabilisation to a rehabilitation centre as per physiotherapy recommendations.

DISCUSSION

Our patient presented with unilateral involuntary movements of the entire right extremity. She did not have a history of diabetes known to her but she did have symptoms suggestive of long-standing undiagnosed diabetes. Abnormal movements remitted completely after rapid glycaemic control with institution of aggressive intravenous insulin therapy but returned despite fairly tight glucose control in about 4 weeks. Haloperidol and gabapentin helped control the symptoms but did not completely resolve suggesting permanent neuronal damage secondary to persistent hyperglycaemia.

Hemiballismus–hemichorea is a neurological syndrome that constitutes a clinical spectrum of continuous, involuntary movements involving one side of the body, isolated mainly to the upper extremity.

The triad of chorea, hyperosmolar non-ketotic hyperglycaemia and a high-signal basal ganglia lesion on the T1-weighted brain MRI is considered a unique syndrome. The

Figures 3 (A and B) repeat MRI of head—T2 and T2 fluid attenuation inversion recovery. Although degraded by motion artifact, there is no evidence of acute vasculature for infarct. Again noted is persistent abnormal T2 hyperintensity with T1 shortening involving the left basal ganglia (black arrows).

MRI will usually appear as small infarcts or haemorrhages in the vicinity of the subthalamic nucleus contralateral to the hemiballismus. It is typically characterised by unilateral involuntary movements, contralateral to the striatal abnormality and rapid remission of symptoms after correction of hyperglycaemia. Generally, patients with this symptom have long-standing, poorly controlled diabetes.^{3–4} Our patient did not have a known history of diabetes. Our presentation is consistent with previous literature in that abnormal movements were first manifestation of her undiagnosed diabetes. Despite significant improvement in glycaemic control, she presented with involuntary movements a month later. While many literature cases demonstrate permanent reversal of symptoms,⁵ some demonstrate incomplete recovery. It is proposed that in long-standing uncontrolled hyperglycaemia, patients may develop permanent neuronal changes and damage.⁶

The diagnosis is made mainly by a high clinical suspicion through the presence of the triad consisting of chorea/ballismus, hyperosmolar non-ketotic hyperglycaemia and basal ganglia findings on MRI as well as the absence of another likely diagnosis. As discussed, the differential diagnosis is lengthy but an extensive workup maybe unnecessary if the symptoms abate quickly with glycaemic control.

Recognition of the syndrome is important because correction of the underlying hyperglycaemia, with temporary use of haloperidol and the addition of diazepam if necessary, will lead to rapid improvement. In our patient, hyperkinesia resolved dramatically after control of the hyperglycaemia within 48 h during the initial admission but reappeared later necessitating the use of other agents for symptom control. Abnormal lesions in the basal ganglia display gradual resolution following treatment on follow-up CT or MR images; however, resolution of the lesions on the imaging studies is slower than the clinical progress. The sequential MR findings in our patient also correlated well with the clinical course.⁷

Clinical outcome is generally good. Resolution of symptoms is seen in most cases.⁸ Rarely chorea does not resolve with glycaemic control as the result of irreversible neuronal damage as in our patient. However, few patients develop permanent hemichorea. Treatment should be directed at managing blood sugars optimally, with symptomatic management in those with residual symptoms. Physicians caring for patients presenting with acute

hemiballistic movements should consider this syndrome and address glucose metabolism issues promptly. Management of long-term comorbidities associated with this syndrome is also essential, as is physical therapy and rehabilitation.

Learning points

- ▶ Consider hemichorea hemiballismus syndrome in patients presenting with acute hemiballistic movements.
- ▶ The typical triad includes hyperglycaemia which may be new onset accompanied by chorea, and characteristic MRI findings.
- ▶ Once diagnosis is suspected, early and aggressive glycaemic intervention can lead to complete resolution of the condition.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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