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Diabetic non-ketotic hyperglycemia and the hemichorea-hemiballism syndrome: A report of four cases

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Abstract

Diabetes mellitus is a common affliction. Although the routine diagnosis and treatment of diabetes are quite familiar to medical personnel, it occasionally presents with some unusual manifestations, one such being the hemichorea-hemiballismus syndrome. We report four non-ketotic hyperglycemic diabetic patients who presented with hemichorea-hemiballismus syndrome. Their brain CT scan pictures showed non-enhancing hyperdensities in the basal ganglia and the MRI pictures showed hyperintensities on T1W image and hypo-intensities on T2W image.

INTRODUCTION

Non-ketotic hyperglycemia in patients with diabetes mellitus can be responsible for a rare clinical syndrome characterized by hemichorea-hemiballism associated with unique radiological features.[1] We report four cases of elderly diabetics with this movement disorder along with their CT and MRI appearances. The signal abnormalities were seen to involve the basal ganglia but spared the internal capsule in all cases.

CASE REPORTS

All our four diabetic patients with non-ketotic hyperglycemia presented with more or less similar clinical features. On controlling the blood sugar, the hemichorea-hemiballism movements abated remarkably, but did not disappear fully. All patients were treated with oral haloperidol (mean dose of 10 mg/day for a period of 2 months) for symptomatic relief. The clinical findings, laboratory data and imaging of the four patients are tabulated in Table 1.

DISCUSSION

The above cases illustrate the need for physicians to be aware of non-ketotic hyperglycemia as a cause for a potentially reversible hemichorea-hemiballism syndrome. All patients had poorly controlled diabetes and presented acutely with hemichorea-hemiballism movements. The brain imaging also showed typical radiological features already described, which appear possibly characteristic of the hemichorea-hemiballism syndrome occurring in non-ketotic hyperglycemia. In all our patients, the CT scan was done within 24 hours and the MRI brain was done within a week after the onset of symptoms.

The most common cause of hemichorea-hemiballism in adults is a vascular lesion in the basal ganglia. Rarely, it can also be the first clinical manifestation of non-ketotic hyperglycemia.[2] Hemichorea-hemiballism generally disappears within hours after correction of hyperglycemia.[3] However, it has also been reported that some patients can have persisting involuntary movements for more than 3 months.[4] In all our patients, once they had the diabetes adequately controlled, their involuntary movements abated significantly. Three of the patients who were reviewed two months later continued to have minimal involuntary movements. One patient (Patient 4) had bilateral symptoms persisting during follow-up.

Elderly non-ketotic hyperglycemia diabetic patients presenting with hemichorea-hemiballism and showing hyperdensities in the contralateral basal ganglia on CT scan and high signal intensity in corresponding areas on T1-weighted magnetic resonance scans have been reported already.[1] There is much controversy regarding the cause of these imaging changes. Initially it was thought to be due to calcification.[5] Chang and colleagues had postulated petechial haemorrhage to be the cause.[6] Pathologic findings by stereotactic biopsy from the striatum revealed gliotic brain tissue

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Table 1: Clinical details of the four patients

Parameters	Patient 1	Patient 2	Patient 3	Patient 4
Age / Sex	57 / Male	56 / Female	76 / Female	84 / Male
Duration of Diabetes	4 years	10 years	7 years	5 years
Blood Glucose (mmol/L) on admission	19.1	12.8	16.9	17.5
Serum Osmolality (mOsm/L)	306	304	290	292
HbA1c (%)	12.5	13.6	10.5	12.0
Ketones in urine/ plasma	Negative	Negative	Negative	Negative
Description of the hemichorea-hemiballism and other involuntary movements(if any)	Uncontrollable, jerky movements involving the left upper and lower limbs which had evolved over six days.	Continuous, violent, flinging movements of the right upper and lower limbs which had commenced rather abruptly.	Continuous jerky movements of the right upper and lower limbs associated with abnormal chewing movements of the mouth which had commenced all of a sudden.	Generalized body weakness lasting 3 days followed by non-rhythmic, purposeless movements involving all 4 limbs, which had commenced abruptly.
Treatment given: Oral hypoglycemics / insulin	Actrapid 10 units thrice daily subcutaneously.	Gliclazide 80 mg twice daily orally.	Actrapid 10 units thrice daily subcutaneously.	Gliclazide 80 mg and metformin 850 mg twice daily orally
CT brain	Hyper-densities in the right putamen and caudate nucleus. (Figure 1)	Hyper-densities in the left putamen and caudate nucleus.	Normal	Hyper-densities in the basal ganglia.nuclei bilaterally (Figure 3)
MRI brain	Hyper-intensities on T1WI and hypo-intensities on T2WI in the right putamen and caudate nucleus	Hyper-intensities on T1WI over the left putamen and caudate nucleus and hypo-intensities over the same area on T2WI.(Figure 2)	Hyper-intensities on T1WI and hypo-intensities on T2WI in the left basal ganglia.	Not done (Patient was not willing for MRI)
Response after correction of the hyperglycemia	Involuntary movements improved markedly in 4 days.	Involuntary movements abated markedly within 24 hours, though not completely.	Abnormal movements of the mouth disappeared within 24 hours, though the hemichorea-hemiballism movements persisted in the limbs.	Improvement in the involuntary movements occurred gradually in the next 2 weeks.
Status during 2 month review	Mild persistent movements.	Very occasional mild bouts of hemichorea-hemiballism movements.	Mild improvement in hemichorea-hemiballism.	Movements persisted

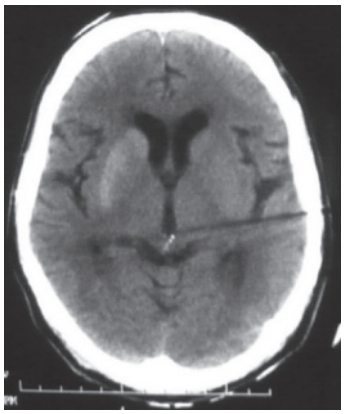

Figure 1. CT Brain showing hyper-densities in the right putamen and caudate nucleus (Patient 1)

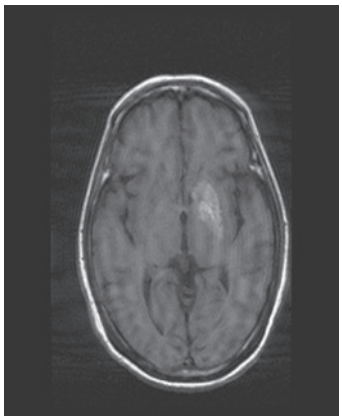

Figure 2. T1WI showing hyper-intensities in the left putamen and caudate nucleus (Patient 2)

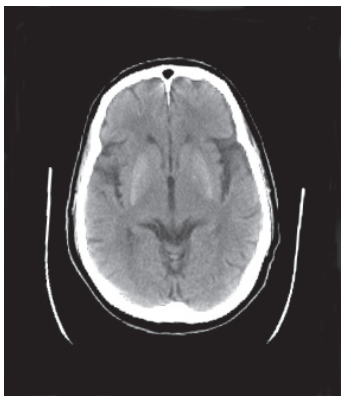

Figure 3. CT Brain showing bilateral basal ganglia hyper-densities (Patient 4)

with abundant gemistocytes suggesting that the hyperintensities in T1 could be due to the protein hydration layer inside the cytoplasm of the swollen gemistocytes, as is observed in cases of gemistocytic astrocytoma.[7]

The exact cause for hemichorea-hemiballism occurring in non-ketotic hyperglycemia is poorly understood. People of Asian origin seem to be particularly susceptible to hemichorea-hemiballism, which suggests a possible genetic predisposition to the disorder.[8] Shan and colleagues have postulated that the gemistocytes abundantly present in the basal ganglia cause excessive neuronal activity especially in the GABA-ergic projections and thus may be responsible for generating hemichorea-hemiballism.[9] The basal ganglia hyperintensity generally resolves within a few months, although there has been a case report of persisting striatal hyperintensity on T1-weighted MRI for up to 6 years.[9]

It may be concluded that hemichorea-hemiballism occurring in diabetes mellitus owing to non-ketotic hyperglycemia is a rather benign condition with a good prognostic outcome once the hyperglycemia is recognized early and corrected.

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