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Non Ketotic Hyperglycaemic Chorea : A Case Report

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ABSTRACT

Chorea is an infrequent expression of nonketotic hyperglycaemia. This is a distinctive case that shows the need to be aware of nonketotic hyperglycaemia as a cause of chorea in a 72year old Asian female patient with poor glycaemic control. The patient was a known case of type 2 diabetes mellitus and was not on regular medication for diabetes mellitus. She had an acute onset of involuntary movements on both limbs. Laboratory test showed random blood sugar was very high (1013mg/dl), ESR was elevated (72mm/hr), urine sugar (4+) and urine ketone negative. Computerized tomography brain showed age related atropic changes and no evidence of focal space occupying lesion or intracranial haemorrhage. She was started on insulin therapy, movements reduced, however involuntary movement reappeared, so anti chorea medications were added to the treatment. Patient became better, her glycaemic state gradually decreased and the involuntary movements was also improved and got discharged.

INTRODUCTION

Chorea is defined by world federation of neurology as "a state of excessive, spontaneous movements, irregularly timed, non-repetitive, randomly distributed and abrupt in character. These movements may vary in severity from restlessness with mild intermittent exaggeration of gesture and expression, fidgeting movements of hands, unstable dance like gait to a continuous flow of disabling, violent movements." The hereditary cause of chorea is Huntington disease which is one of the tri nucleotide repeat disorders [1]. Numerous illnesses such as cerebrovascular diseases, neurodegenerative diseases, immunological diseases, infectious diseases, and neoplastic diseases are notable as secondary reasons of chorea. Chorea is also an unusual presentation of hyperglycaemia that generally impact on elderly Asian women with miserable glycaemic control [2]. The pathophysiology of chorea associated with hyperglycmia is debatable but these patients show definite neuroimaging characteristics [3]. Hyperglycaemic chorea is characterized by striatial hyperintensity on T1-weighted MRI, but T2-weighted brain MRI observations may vary and short-lasting chorea [3,4]. This short-lasting chorea is presented by strong involuntary movements on one side of the body, these actions mostly include the upper extremity. In the previous three decades, various patients with this ailment correlated to high blood glucose level were reported. Nearly all these patients have a reasonably fast recovery from their symptoms with the rectification of hyperglycaemia. Supplementary anti-chorea medications may be accounted for refractory cases. [4, 5] We detail a case of an adult female who arrived at our hospital with generalized fatigue, decreased food intake, constipation and acute involuntary movements of hand and diagnosed as chorea secondary to nonketotic hyperglycaemia with negative CT changes. We also mark out the role of differential diagnosis based on neuroimaging through this case.

CASE PRESENTATION

A 72 years old female patient presented to our hospital with generalized fatigue, decreased food intake, constipation and acute involuntary movements of hands. She had a history of type 2 diabetes mellitus and was not on regular diabetic medication. She

was also a known case of HTN, old CVA and old NSTEMI. Her random blood sugar was 1013mg/dl, ESR (72mm/hr) and urine sugar (4+) was elevated but urine ketone was negative. Vitals signs were normal upon presentation. The patient's renal function test, liver function tests, thyroid function tests found no abnormal findings. She was started on insulin infusion initially and then changed to injection human actrapid 12units TID. After 3 days, injection human actrapid was changed to injecton human mixtard (30/70) 15-0-16 units. In addition, oral hypoglycaemic agents; tab metformin, vildagliptin combination and tab glimepride were added. MRI brain was advised by neurologist on sixth day but the patient showed abnormal movements even after administered sedatives so MRI was not able to perform, on account of this computerized tomography brain was advised. Computerized tomography of brain showed age related atropic changes and no evidence of focal space occupying lesion or intracranial haemorrhage. ECHO showed grade 1 diastolic dysfunction and aortic valve sclerosis with mild AR and mild MR, while USG abdomen was found to be normal. Arterial blood gas did not show acidosis. She was diagnosed with chorea associated with hyperosmolar hyperglycaemia. Meanwhile there were no further occurrence of abnormal movements of hand, so planned to discharge but then the patient again showed choreiform movements of hand. And so additional anti-chorea medication tab tetrabenazine 25mg half BD and tab amantadine 100mg BD was added to the treatment. Patient became better, her glycaemic state gradually decreased and the involuntary movements was also improved and was discharged with tab tetrabenazine and tab amantadine along with insulin and oral hypoglycaemic agents. After one week of follow up the involuntary movement didn't arise again.

DISCUSSION

Hyperglycaemic chorea is a rare condition that shows in the situation of uncontrolled diabetes mellitus. Nearly all patients presenting with hyperglycaemic chorea are elderly female patients of Asian origin [6]. Various hypotheses have been proposed to describe the mechanism of chorea, hyperglycaemia, ischemia and microheamorrhage. Not all diabetic patients have this complication. The majority patients with hyperglycaemic chorea present sub-acute or acute rise in the involuntary activities of limb, occasionally involuntary movements also develop in the jaw, tongue, facial muscle and negative ketones in urine. Most patients' abnormal movements get better during sleep [2]. In our case the patient was presented with involuntary movements of the extremities with a history of unrestrained diabetes mellitus. The patient's urine ketone was negative, the patient's involuntary movements appeared even during sleep.

Typically in patients with non ketotic chorea Computed tomography manifest hyper attenuation in the striatum contralateral to the altered side.[7] this can sometimes mistake for basal ganglia haemorrhage[5] But sometimes MRI is useful when CT brain is normal. The characteristic MRI feature is T1 and T2 hyperintense deformities and restricted diffusion in DWI [6,8]. There are extremely a small number of patients with a negative presentation in MRI or CT scan [2]. In the present case, computerized tomography brain showed age related atropic changes and no evidence of focal space occupying lesion or intracranial haemorrhage.

The correct mechanism of non ketotic hyperglycaemic chorea remains unclear. The possible mechanism may be, when hyperglycaemia occur, brain cell metabolism gradually transformed into anaerobic metabolism because of glucose metabolism failure. So the main source of energy for brain becomes gamma amino butyric acid. In non ketotic hyperglycaemia patients, the GABA rapidly depleted due to lack of acetoacetate, so activities of basal ganglia damaged and thus it's easy to suffer from chorea [2]. In elderly females the post-menopausal changes of GABA and dopamine receptors may be the reason for higher occurrence in this population [5].

Only few cases have been reported with negative imaging. In a case report [2] they divided the syndrome into two types; patients with history of diabetes, presented with high blood glucose, negative keteone, unilateral or bilateral chorea and typical radiographic changes in MRI or CT of brain, are the most common type and patients with history of diabetes, presented with high blood glucose, negative keteone, unilateral or bilateral chorea and negative imaging are comparatively uncommon type. In the present case patient CT showed no evidence of focal space occupying lesion or intracranial haemorrhage.

The management of blood glucose level is indeed the most important treatment. The involuntary movement in most patients can be alleviated with the reduction in blood glucose level. The recovery of chorea varies from one day to few months. Only few patients can have persistent chorea [5]. Supplementary medications such as D2 receptor antagonist may be accounted for refractory cases, also tetrabenazine has been effectively used in some patients [2,5]. Here in this case in addition with rectification of hyperglycaemia, tab tetrabenazine and tab amantadine was added to the treatment as the patient's abnormal movement reappeared.

CONCLUSION

Non ketotic hyperglycaemic chorea is a treatable neurological condition that deserves attention because it is reversible by treating the hyperglycaemia. Thus early differential diagnosis and providing treatment is necessary to relive the symptoms and avoid further complications.

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