ORIGINAL RESEARCH PAPER

Volume-7 | Issue-5 | May-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

DIABETIC STRIATOPATHY - AN UNCOMMON COMPLICATION OF TYPE I DIABETES MELLITUS : A CASE REPORT

Paediatrics

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ABSTRACT

Diabetic striatopathy (DS) is a rare complication of diabetes, which appears as a result from a vascular and metabolic insult to the basal ganglia due to chronic hyperglycemia in the context of poorly controlled type 2 or rarely type 1 diabetes. Very few cases have been found in literature, as it's a rare complication of type 1 diabetes. We describe one such case of diabetic striatopathy found in 16 year old boy with type 1 diabetes. DS was presented with symptom of involuntary movements involving left side of the body with poor glycemic control. Glycemia control allowed rapid clinical recovery despite established striatal lesions documented on neuroimaging. Hence early identification and prompt treatment of DS results in good clinical recovery.

KEYWORDS

Diabetic striatopathy, Type 1 Diabetes, Hemichorea and hemiballismus.

INTRODUCTION:

Diabetic Striatopathy denotes a clinic-radiologic syndrome of chorea-ballism and hyperintensities on neuroimaging1.Clinically, it may be presented by abnormal movements of 1 body side,hemichorea–hemiballism (HH) contralateral to the brain lesions. Brain MRI shows signal abnormalities on contralateral basal ganglia to the symptoms. Treatment includes the correction of hyperglycemia and in some patients the addition of specific medications may be required to inhibit abnormal movements, such as neuroleptics, dopamine-release inhibitors, or anticonvulsants2. The clinical outcome is favourable.DS is a well-known condition in adults, very few cases are found as a complication with type 1 diabetes mellitus. We here present the case of a 16year old boy who presented with subacute HH revealing underlying contralateral striatopathy as a complication of poorly controlled insulin-dependent diabetes.

CASE REPORT:

A 16-year-old boy, diagnosed to have type 1 diabetes at the age of 5 years presented with the acute complaints of intermittent abnormal involuntary movements in his left side of the body before his first visit to our centre. The movements were exacerbated by physical activity and disappeared during sleep. There was no history of autoimmune diseases, neurologic disorders of any kind, or abnormal movements in the family. On admission, there was presence of continuous irregular movements of low amplitude and high frequency involving the left upper limb and lower limb sparing face, mixed with higher-amplitude, brisk jerks involving the left arm and the left leg, consistent with HH. There was no additional extrapyramidal sign like rigidity or bradykinesia.

Child was diagnosed to have type 1 diabetes at the age of 5 years and is on regular insulin therapy(4 Units of Mixtard Insulin twice daily, inadequate and inappropriate dosage for a 30kg weighing boy). On admission child was investigated for glucose levels, which revealed a value of 31mmol/L (N for random measurement <11.1mmol/L). The glycosylated hemoglobin A1c (HbA1c) level was 20.0%(N < 7%), urine ketones were absent and the pH was 7.39. These results showed poorly controlled type 1 diabetes mellitus without ketoacidosis.The results of all other laboratory values on admission are summarized in (Table 1).

Table 1: Laboratory results.

Tests	Results	Normal range
pH	7.38	7.35-7.45
Potassium	4.3	3.0- 5.0 mEq/L
Sodium	136	135-145 mEq/L
Glycated HB	20.2%	<7%
RBS	526 mg/dl	90-130 mg/dl
Urine ketones	Nil	-
Serum ketones	Nil	-

RBS: Random Blood Sugar.

CT Brain Plain done on admission showed hyper dense area seen involving right Basal Ganglia.In the view of acute clinical history, hyper dense area involving right Basal Ganglia could suggestive of Diabetic Striatopathy(Fig 1). Child was started on intravenous insulin at initial dosages of 1.1 IU/kg per day allowed a rapid normalization of blood glucose levels and a clearance of ketones, later switched to subcutaneous insulin. Even after the correction of hyperglycemia, theabnormalmovementswere still persisted. Therefore, sodium valproate treatment was started. After 2 weeks, abnormal movements decreased and sodium valproate was tapered and stopped without recurrence of movements. On the last follow-up visit, at 8 months after onset, child was asymptomatic with no recurrence of abnormal movements.

Figure 1

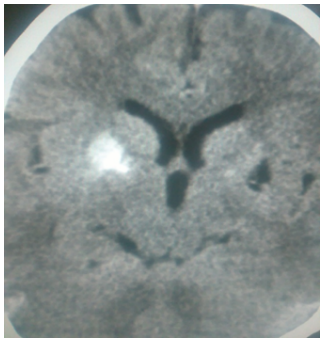

Fig 1: CT Brain Plain showing hyper dense area involving right Basal Ganglia. An abnormal signal of the right striatum, in the caudate nucleus and putamen, characterized by hyperintensity on T1 sequences, suggestive of diabetic striatopathy.

DISCUSSION:

Diabetic striatopathy secondary to nonketotichyperglycemiais a well-known complication of diabetes in adults[1]. These patients usually lack the presence of ketone bodies in their urine or blood (nonketotichyperglycemia), the cause is not known. Their brain MRI shows lesions in the striatum, either contralaterally to the clinical manifestations2 or more rarely bilaterally[2]. The prognosis of abnormal movements in that situation is usually good, and generally, most symptoms disappear completely after a few days, weeks or months on normalisation of blood glucose levels[1].Very few similar cases have been reported so far in pediatric age group with type 1 diabetes.

Rarely patients may require treatment with drugs to control abnormal movements as in our case. Other options to use are neuroleptics, dopamine release inhibitors, benzodiazepines, or anticonvulsants[2].

Resolution of radiological features takes a longer course, few months to years.

The first pediatric case was described by Mihaiet al[3]. They reported the case of a previously healthy 15-year-old girl with a history of sudden-onset repeated involuntary movements involving her right side of the body including face. She had initially presented with paraesthesia of the right leg, face, and all the right side of her body, with symptoms increasing during exercise and disappearing during sleep. A diagnosis of Sydenham chorea was first considered, but the search for cause was negative. Blood tests showed hyperglycemia at 20.3 mmol/L and HbA1c levels of 13.9% (128.4 mmol/mol), and urine tests showed glycosuria and ketonuria; all of these results confirmed the diagnosis of type 1 diabetes. Insulin treatment was started, and after 24 hours of treatment the blood glucose levels were corrected and the abnormal movements had disappeared.

In 2012, Alveset al[4] published the second case of a boy known for type 1 diabetes mellitus since the age of 2 months. His follow-up was irregular, and the metabolic control of his disease was poor. He had been taking human insulin on a regular basis for several years and was admitted at 8 years because of subacute onset of uncontrolled upper limb movements. HH was noted on examination, with movements involving mainly the patient's right side. At the time of admission, blood samples showed slightly elevated blood glucose levels of 8.7 mmol/L, the patient's HbA1c levels were 13.5% (124 mmol/mol), indicating poor metabolic control over the previous 3 months. The brain MRI showed abnormal signals in the striatum, bilaterally and symmetrically. The patient was initially treated with valproic acid (16 mg/kg per day). Two days later the abnormal movements had disappeared.

Faundezet al[5] reported a 13-year-old, previously healthy teenager of Uzbek origin with intermittent abnormal movements in his left hand. The movements were present and continuous when the patient was at rest; they were exacerbated by physical activity and disappeared during sleep. His personal history revealed a significant weight loss of 10 kg in the past 6 months, and his appetite was intact. Given the context of polydipsia, polyuria, and weight loss, when checked the patient's glucose levels, which revealed hyperglycemia, these results permitted the diagnosis of diabetes mellitus without ketoacidosis. MRI brain done showed an abnormal signal of the right striatum, suggestive of subacute ischemia with hemorrhagic alterations. A week after the correction of glycemia, the movements were still present and disabling requiring treatment with tetrabenazine.

The most frequently accepted pathophysiological hypothesis for diabetic striatopathy involves cerebral local hypoperfusion due to hyperviscosity that follows chronic hyperglycemia[6]. This hypoperfusion leads to anaerobic metabolism, which in turns reduces the levels of γ-aminobutyric acid. Its decreased concentration leads to an increase in thalamocortical activity, causing abnormal movements like HH[3,7]. In most cases, hyperglycemiaassociated with diabetic striatopathy is nonketotic and is therefore not complicated by acidosis at diagnosis. This fact may explain the patient's tolerance to chronic hyperglycemia, likely to be necessary to cause the aforementioned pathologic changes in the basal ganglia. The details of the mechanisms that underlie the basal ganglia lesions remains unknown. In typical cases, cerebral MRI shows hyperintense signal of the putamen, in T1-weighted images, probably reflecting the accumulation of manganese in gemistocytes (ie, astrocytes with large protein content found during acute brain injury)[7,8,9]. The rare histopathological studies of autopsied adults revealed the presence of gliosis, the accumulation of gemistocytes and a localized loss of neurons, without evidence of hemorrhage or infarct.[10]Similar studies in the pediatric population are wanting, and specific pathologic mechanisms in that age group remain to be understood.

CONCLUSION:

Diabetic striatopathy is a rare entity in childhood with Type 1 Diabetes mellitus secondary to hyperglycemia. Child presenting with abnormal movements on unilateral side of the body should be evaluated and treated appropriately and to avoid appearance of diabetic-related other complications.

ABBREVIATIONS:DS: Diabetic Striatopathy HbA1c: HemoglobinA1c HH: Hemichorea–Hemiballism

Consent: Written informed consent was obtained from the patient's mother for publication of this case report.

Competing interests: No

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