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CASE REPORT

The youngest patient with hemi-chorea and diabetic ketoacidosis as presenting manifestation of type 1 diabetes mellitus from India

Abhijeet Vilas Botre[1] · Vishakha Kashyap[1] · Tushar Parikh[1]

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Abstract

Background Type 1 diabetes mellitus (T1DM) is one of the most common endocrine diseases in children. Majority of them present with metabolic crises in the form of diabetic ketoacidosis (DKA) at diagnosis. Various levels of encephalopathy and seizures are the well-known manifestation of DKA.

Case presentation We are reporting an 8-year-old girl who presented with hemi-chorea with DKA as a manifestation of type 1 DM which is extremely rare in literature in Asian subcontinents and has been encountered at such a young age for the first time in India.

Conclusion T1DM with metabolic crises should always be one of the important differentials of hemi-chorea in children.

Keywords Type 1 DM · DKA · Hemi-chorea

Introduction

Lately, T1DM has been on the rise like type 2 diabetes. Three new cases of T1DM/100,000 children 0–14 years of age are seen in India per year [1]. These children usually get diagnosed when they present with DKA, an acute metabolic crisis with varying degrees of encephalopathy. However, it is extremely rare for a child to present with movement disorder at the time of diagnosis. Though some studies have reported adults presenting with variety of movement disorders due to impaired glycemic control (hypo/hyper-glycemia), similar instances in paediatric age have been scarce [2]. We report the youngest girl who presented with acute hemi-chorea as a first manifestation of type 1 DM due to DKA.

Case presentation

An 8-year-old previously healthy female child presented to our emergency department with history of sudden onset of abnormal, slow, involuntary movements involving both right upper and lower limbs, which increased during activity and disappeared in sleep for 5 days. She also had history of behavioural changes in the form of increased irritability, drowsiness and confusion for one week.

She had no history of fever, headache, seizures and neurological illnesses in the past. She was not receiving any medication at the time of illness or before. Her birth had been uneventful, and she had normal development with good academic performance.

On clinical examination, the young girl appeared conscious, well oriented but irritable with blood pressure of 106/78 mm Hg. She had involuntary rhythmic quasi-purposive movements of right limbs involving upper limbs more than lower. She had near continuous right-sided movements resembling hemi-chorea which used to get explosive in character during anxiety and voluntary movements making it difficult for her to sit or walk. Her higher functions and cranial nerve examination were normal. There was hypotonia of both upper and lower limbs with no signs of meningeal irritation or cerebellar involvement. Her other systemic examination was normal.

Initially, clinical diagnosis of acute hemi-chorea secondary to auto-immune disorder like rheumatic chorea, systemic lupus erythematosus (SLE) Vs vascular stroke was suspected. However, upon further enquiry, she was found to have typical history of weight loss, polyphagia, polydipsia and polyuria over the last 15 days. Routine investigations revealed BSL (blood sugar level): 474 mg/dl with metabolic acidosis having anion gap of 9.2 mmol/dl with positive ketone bodies (serum (5.1) and urine (80 mg/dl)) indicating

✉ Abhijeet Vilas Botre
drabhijeetbotre@gmail.com

[1] K.E.M. Hospital and Research Centre, Pune, India

hyper-glycemic ketoacidosis. MRI revealed (Fig. 1) left-sided basal ganglia (caudate and lentiform nucleus) signal which was hyper-intense on T2-weighted and hypo-intense on T1 images suggestive of metabolic aetiology. Other supportive investigations revealed HBa1c – 16.5 and positive anti-GAD antibodies (137.2) with low C peptide (0.40 ng/ml) further confirming the diagnosis of T1DM.

The child was managed efficiently with BSPED 2020 protocol for DKA. Glycemic control was achieved after 48 h with correction of acidosis and dyselectrolemia. She was conscious, alert and active throughout her stay in the hospital. Her chorea became less severe enabling her to walk and stand without support on day 2 of treatment and betterment of blood sugar levels. She was discharged on subcutaneous insulin, oral tetrabenazine with minimal residual chorea but ambulant status. At the last follow-up after 2 months of discharge, there was no focal neurological manifestations with good compliance with T1DM management. MRI was not repeated due to financial constraints.

Discussion

Type 1 diabetes mellitus (T1DM) is the most common endocrine disorder of children with auto-immune aetiology. Recently, the number of cases of T1DM has increased significantly and we find 3 new cases per 100,000 children of 0–14 years. The peak age at diagnosis is 12 years with girls getting more affected than boys [3]. It may be auto-immune or idiopathic in nature and is present in 9% cases of insulin deficiency. The absence of insulin results in increased concentrations of glucose in the blood (hyper-glycemia) for the elimination of which there is increased water consumption to enable filtration from the kidney leading to a cycle of polydipsia and polyuria until insulin is finally administered. The reduced glucose uptake owing to lack of insulin causes the cells to use fats and proteins for energy which also produces ketones. Diabetic ketoacidosis (DKA) is therefore a common presentation of T1DM. Most children in the US with new onset of T1DM present with the classic signs and symptoms of hyper-glycemia and 30% with (DKA).

Neurologic manifestations, however, are relatively rare and mostly include lethargy, decreased level of consciousness and coma because of DKA. Extreme rarely chorea, cerebellar ataxia, weakness of lower limbs due to sciatic neuropathy leading to atrophy, hemi-paresis like symptoms have also been reported in literature associations of T1DM [4].

Chorea is a continuous spontaneous involuntary movement of distal muscles occurring during rest and activity which tends to disappear in sleep. Hemi-chorea is chorea involving only one side of the body. They are usually seen in lesions of the basal ganglia, tumours of the brain and metabolic, vascular, or degenerative disease. In rare instances, hemi-chorea has also been reported in type 2 DM in adults over 50 years [5]. However, it is extremely rare in the paediatric age group and less than 10 cases have been reported so far over the world but few from India [4, 6].

The exact pathogenesis of this manifestation is obscure, but there has been hypothesis that during hyper-glycemia there is a shift of cerebral metabolism to the anaerobic pathway with the inactivation of the tricarboxylic acid cycle [7]. The brain then uses GABA as an alternative source of energy which eventually leads to metabolic acidosis. In ketotic hyper-glycemia, GABA can be re-synthesized using ketone bodies, whereas GABA and acetate are depleted rapidly in non-ketotic hyper-glycemia-reducing acetylcholine synthesis

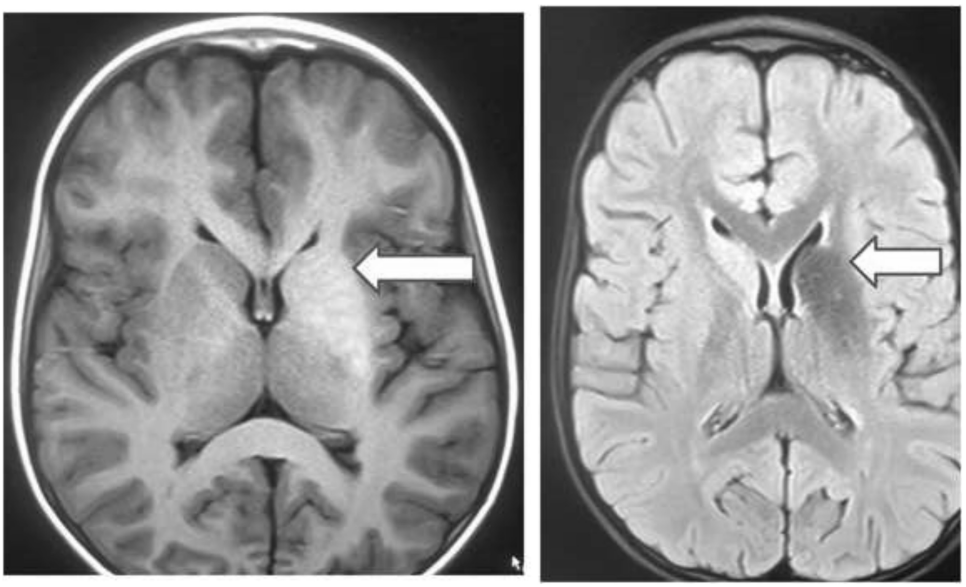

Fig. 1 MRI brain showing hypo-intensities on T1-weighted images and hyper-intensities on T2-weighted images in the left caudate and putamen

due to acetate depletion. Reduction of GABA and acetylcholine (inhibitory neurotransmitters) in the basal ganglia with associated metabolic acidosis and the lack of energy production is thought to cause a basal ganglia dysfunction and subsequent chorea [5]. Another proposed theory attributes occurrence of chorea to hyper-glycemia-induced hyper-viscosity leading to reduced regional cerebral blood flow [8]. Analyses of biopsy specimen of basal ganglia have revealed changes similar to infarction in the form of neuronal loss, gliosis and reactive astrocytosis [9]. These histological changes are also responsible for characteristic MRI signal intensities. Since all the proposed mechanisms are due to hyper-glycemia, the correction of blood sugar levels has shown amelioration of neurological symptoms and sometimes complete resolution of chorea [10]. Nevertheless, there are instances where additional antipsychotics have been required. In a meta-analysis of 53 adults with chorea and non-ketotic hyper-glycemia, 16 had complete resolution only with glycemic control, 31 needed treatment with haloperidol ($n = 18$) and others in combination ($n = 13$), though 7 patients had fluctuating course and were resistant to therapy. No such research has been done in paediatric population as of now.

Imaging studies may/not reveal any changes. Common involvement although has been seen in the basal ganglia especially the putamen and caudate nucleus in the form of hyper-intensities which interestingly resolve after correction of hyper-glycemia [11].

Conclusion

T1DM is a common disorder in the paediatric age group. Majority of them are diagnosed at DKA at emergency unit needing intensive care. Hemi-chorea has been reported extremely rarely with T1DM. This study emphasizes the need for considering it as one of the differentials of acute hemi-chorea which needs immediate attention and prompt treatment.

Abbreviations T1DM: Type one diabetes mellitus; DKA: Diabetic ketoacidosis; Hg: Mercury; GAD: Glutamic acid decarboxylase autoantibodies; SLE: Systemic lupus erythematosus; GABA: Gamma aminobutyric acid; BSPED: Britsh Society for Paediatric Endocrinology and Diabetes

Author contribution AVB helped conceptualized the idea, literature search and summarising concept, discussion and summarising.
VK helped in case summary and discussion of case report.
TP helped in conceptualization and discussion.
All contributed (AVB, VK, TP) in editing and approving the article.

Data Availability Not applicable.

Declarations

Ethics approval and consent to participate Not applicable.

Consent for publication Taken.

Competing interests The authors declare no competing interests.

References

1. Kumar KMP. Incidence trends for childhood type 1 diabetes in India. Indian J Endocrinol Metab. 2015;19(7):S34–5.
2. Mulder L, Onur O, Kleis L, Borders H, Cemeroglu AP. Atypical neurologic presentations of new onset type 1 diabetes mellitus in pediatric age group: a report of five unusual cases and review of the literature. J Pediatr Endocrinol Metab. 2014;27(7–8):749–56.
3. Das AK. Type 1 diabetes in India: overall insights. Indian J Endocrinol Metab. 2015;19(7):S31–3.
4. Alves C, Sampaio S, Barbosa V, Machado M. Acute chorea and type 1 diabetes mellitus: clinical and neuroimaging findings. Pediatr Diabetes. 2012;13(6):1–5.
5. Hashimoto KI, Ito Y, Tanahashi H, Hayashi M, Yamakita N, Yasuda K. Hyperglycemic chorea-ballism or acute exacerbation of Huntington's chorea? Huntington's disease unmasked by diabetic ketoacidosis in type 1 diabetes mellitus. J Clin Endocrinol Metab. 2012;97(9):3016–20.
6. Rai S, Kaul V, Singh S, Kaur S, Thenmurugan P. Diabetic striatopathy: a new challenge in type 1 pediatric diabetic patients. Oman Med J. 2022;37(1):e332. https://doi.org/10.5001/omj.2021.47.
7. Satish PV, Pujitha K, Agrawal N, Mathew T, Vidyasagar S. Hemichorea in a patient with ketotic hyperglycemia: an unusual presentation. J Clin Diagnostic Res. 2017;11(5):OD24-5.
8. Duckrow RB, Beard DC, Brennan RW. Regional cerebral blood flow decreases during chronic and acute hyperglycemia. Stroke. 1987;18(1):52–8.
9. Nath J, Jambhekar K, Rao C, Armitano E. Radiological and pathological changes in hemiballism-hemichorea with striatal hyperintensity. J Magn Reson Imaging. 2006;23(4):564–8.
10. Oh SH, Lee KY, Im JH, Lee MS. Chorea associated with nonketotic hyperglycemia and hyperintensity basal ganglia lesion on T1-weighted brain MRI studya meta-analysis of 53 cases including four present cases. J Neurol Sci. 2002;200(1–2):57–62.
11. Mihai CM, Catrinoiu D, Stoicescu RM. Atypical onset of diabetes in a teenage girl: a case report. Cases J. 2008;1(1):1–5.