ORIGINAL RESEARCH PAPER

Medical Science

FACIOBRACHIAL MYOCLONIC SEIZURES AS A RARE PRESENTATION OF DIABETIC KETOACIDOSIS – A CASE REPORT.

KEY WORDS: myoclonus, diabetic ketoacidosis, ballismus, chorea.

Dr. Jaya Lalwani

ABSTRACT

While chorea and ballismus are the most common movement disorders associated with non ketotic hyperglycemia, Facio-brachial myoclonic seizure is a very rare manifestation of diabetic ketoacidosis. Here we report a 53 years old female presenting with facio-brachial myoclonic seizures as an initial manifestation of diabetic ketoacidosis. Here we report a 53 years old female presenting with right sided facio-brachial myoclonic seizures as an initial manifestation of DKA and eventually diagnosed as type 2 diabetes mellitus. Myoclonus disappeared with glycemic control. Apart from expanding the horizon of movement disorders in diabetes mellitus, this case explains about the importance of rapid bedside measurement of capillary blood glucose among patients presenting with movement disorders is highlighted. Prompt diagnosis and proper management remains the corner stone.

INTRODUCTION:

Diabetic ketoacidosis (DKA) is a common acute complication of diabetes mellitus. This causes volume depletion, electrolyte and acid base imbalance. Nausea, vomiting, excessive thirst, polyuria, abdominal pain, shortness of breath are the common presenting symptoms. Non ketotic hyperglycemia is an established cause of chorea and many cases were reported but it is not well documented in DKA [1].

Type 2 diabetes mellitus also develop DKA, due to fast breakdown of fat into ketones in the liver and this causes the blood to become acidic. This is called as ketoacidosis. DKA is less common in type 2 diabetes mellitus, it is usually occurs due to prolonged uncontrolled blood sugar, stress, surgery, infection, and missing doses of medicine.

Myoclonus is characterized by sudden, brief, shock like involuntary movements, associated with bursts of muscular activity (positive myoclonus) and silencing of muscular activity (negative myoclonus) [1]. Metabolic encephalopathies (hypo or hyperglycemia) cause positive or negative myoclonus[2].

The horizon of movement disorders in diabetes mellitus is expanding. While chorea and ballismus are well-recognized acute potentially reversible movement disorders as the presenting manifestation of non-ketotic hyperglycemic states among older type-2 diabetics [3], different types of myoclonus are also being recently reported [4].

We here in report the case of a 53-year-old female presented with acute onset of facio-brachial myoclonus seizures. The patient was found to be suffering from diabetic keto-acidosis (DKA) and eventually diagnosed as type-2 diabetes mellitus (T2DM). This case explains about the important of rapid bedside measurement of capillary blood glucose (CBG) among patients presenting with movement disorders is highlighted. Prompt diagnosis and proper management remains the corner stone.

CASE REPORT

A 53 years old female with no known comorbidities admitted in our hospital with right sided facio brachial myoclonic seizures with intact awareness lasting more than two hours and was not responding to antiepileptics. On physical examination the patient was awake, obeying oral commands and was having abnormal, involuntary, rapid, brief and jerky movements involving face and upper limb on the right side.

She had facio-branchial myoclonic jerks as with asymmetric involvement of right-side upper limb and face than on left side and without any abnormal movement of bilateral lower limbs. These jerks are not associated with any voluntary action or external clues. Deep tendon reflexes and tone of the upper limbs are not assessed due to abnormal involuntary movements. Deep tendon reflex and tone of lower limbs were normal.

Bedside investigations, capillary blood glucose (CBG) level was measured and found to be 572 mg /dl. Arterial Blood Gas (ABG) analysis revealed metabolic acidosis. HBA1C was measured and was found to be 9.2 %.

Urine analysis showed ketone positive. Thus a diagnosis of DKA was confirmed. Other blood investigations were normal. MRI brain revealed cortical hyperintensity on T2W, FLAIR involving right parieto-occipital region with adjacent subcortical white matter hyperintensities. Fundus examination revealed bilateral Non-Proliferative Diabetic Retinopathy (NPDR). Seizure was not responding to antiepileptics but terminated with glycemic control.

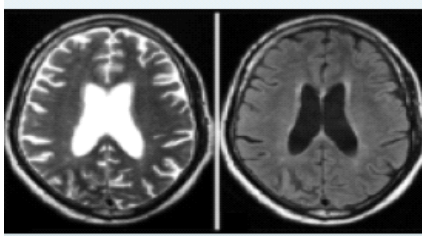

Fig 1: MRI Brain

Patient was newly diagnosed of type 2 diabetes mellitus in DKA with a neurological manifestation. Usually in DKA, ketones have an anti-convulsant effect owing to intracellular acidosis and increased GABA, while HHS lowers the seizures threshold.

DISCUSSION:

Myoclonus is not necessarily representing a pathological phenomenon. Myoclonus is a symptom of neurodegenerative or epileptic disorder. Data relevant to its incidence and prevalence are not well documented [1]. Symptomatic myoclonus was the common type. Neurodegenerative and dementing diseases are the most common cause of symptomatic myoclonus, about 70% [5]. The most important aspect of this case was facio-brachial myoclonus seizures as the first clinical manifestation in DKA.

Epilepsia partialis continua with secondary generalization [6,7], facio-brachial dystonic seizures [8,9,10], Wernicke's encephalopathy with myoclonus and Anti-GAD antibody associated limb myoclonus with encephalopathy [11] are the differential diagnosis.

Movement disorder (chorea and ballismus) are noted in different states of diabetes mellitus. In most of the cases, chorea is being reported in non ketotic hyperglycemia. In our

case, we found out DKA is a rare trigger for movement disorders. With our report we confirmed there is a relationship between DKA and abnormal movements.

The pathophysiology of movement disorders in DKA is unclear. This is multifactorial and hyperglycemia is the important factor. Sheng Feng Sung et al reported that hyperintensity on MRI brain revealed selective neuronal death and gliosis. These lesions may produce abnormal movements sometimes in hyperglycemic state. The focal neurological deficit is more common in cases of non ketotic hyperglycemia[12]. Similarly in our case, patient with type 2 diabetes in DKA, MRI brain showed cortical hyperintensity on T2W, these may result in movement disorders.

Joana Siva Marques et al[13] reported a case of a female patient with uncontrolled type 2 diabetes presented with hyperglycemic hemichorea on the right upper limb. In our study, we found right sided facio-brachial myoclonus in DKA.

Myoclonus among diabetic patients may generally occur in the setting of uremia, resulting from diabetic nephropathy[14], drug-toxicity[15], or in relation with some rare syndromes[16]. Dubey et al[17] reported, diaphragmatic and action myoclonus a presenting sign in non-ketotic hyperglycemic state in a previously undiagnosed diabetic woman. Myoclonus as the presenting feature of diabetic amyotrophy has also been described [18,19]. Myoclonus had been reported in hyperosmolar hyperglycemic state [20]. Ghia et al [21] explained, a 17-month-old child presented with opsoclonus-myoclonus-ataxia syndrome and investigations revealed a thoracic neuroblastoma and eleven days later, she re-presented with DKA.

Chatterjee at al reported, a case with previously healthy patient presented with acute onset facio-brachial myoclonus for 10 days. She was suffering from DKA and eventually diagnosed as type1 diabetes mellitus [22].

Our case is unique from all other previously published cases, a 53 years old female with no known comorbidities with facio-brachial myoclonic seizures is an initial manifestation of DKA in type 2 diabetes mellitus.

CONCLUSION:

Acute movement disorders are usually the presenting features of non ketotic hyperglycemic state, but acute movement disorder as the presenting feature of DKA is rare in type2 diabetes. The importance of CBG measurement among patients presenting with movement disorders is highlighted. Prompt diagnosis and proper management remains the cornerstone.

REFERENCES:

1. Caviness J.N. Myoclonus. Mayo Clin. Proc. 1996;71:679–688. doi: 10.1016/S0025-6196(11)63006-1.
2. Angel M.J., Chen R., Bryan Y.G. Metabolic encephalopathies. In: Young G.B., Bates W.E., editors. Handbook of Clinical Neurology. Elsevier; Amsterdam, The Netherlands: 2008. pp. 115–165.
3. Chua CB, Sun CK, Hsu CW, Tai YC, Liang CY, Tsai IT. "Diabetic striatopathy": clinical presentations, controversy, pathogenesis, treatments, and outcomes. Sci Rep. 2020; 10: 1594. DOI: https://doi.org/10.1038/s41598-020- 58555-w
4. Dubey S, Chatterjee S, Mukherjee D, Ghosh R, Sengupta S, Lahiri D, et al. "Dancing belly" in an old diabetic lady. J Family Med Prim Care. 2020; 9: 2580–2.
5. Caviness J.N., Brown P. Myoclonus: Current concepts and recent advances. Lancet Neurol. 2004;3:598–607. doi: 10.1016/S1474-4422(04)00880-4.
6. 3.Lozsadi D. Myoclonus: a pragmatic approach. Pract Neurol. 2012; 12: 215–24. DOI: https://doi.org/10.1136/ practneurol-2011-000107
7. Khan Z, Arya K, Bollu PC. EpilepsiaPartialis Continua. 2020 Sep 4. In: StatPearls [Internet]. Treasure Island, FL: StatPearls Publishing; 2020 Jan–. PMID: 30335308.
8. Iyer RS, Ramakrishnan TCR, Karunakaran, Shinto A, Kamaleshwaran KK. Faciobrachial dystonic seizures result from fronto-temporo-basalganglial network involvement. Epilepsy Behav Case Rep. 2017; 8: 47–50.
9. Morano A, Fanella M, Giallonardo AT, Di Bonaventura C. Faciobrachial Dystonic Seizures: The Borderland Between Epilepsy and Movement Disorders. Mov Disord Clin Pract. 2020; 7: 228–9. DOI: https://doi.org/10.1002/mdc3.12884
10. Simabukuro MM, Nóbrega PR, Pitombeira M, Cavalcante WCP, Grativvol RS, Pinto LF, et al. The importance of recognizing faciobrachial dystonic seizures in rapidly progressive dementias. Dement Neuropsychol. 2016; 10: 351–7.
11. McKeon A, Tracy JA. GAD65 neurological autoimmunity. Muscle Nerve. 2017; 56: 15–27.
12. S. Sung et al. Focal Neurological Symptoms as the Presenting Manifestations of Nonketotic Hyperglycemia: Report of Two Cases. Medicine. 2007; 18: 206-211
13. Joana Siva Marques et al. Hyperglycaemic hemichorea. Eur J Case Intern Med. 2018; 5(4): 000807.
14. Jagota P, Bhidayasiri R, Lang AE. Movement disorders in patients with diabetes mellitus. J Neurol Sci. 2012; 314: 5–11.
15. Guddati AK, Zafar Z, Cheng JT, Mohan S. Treatment of gabapentin-induced myoclonus with continuous renal replacement therapy. Indian J Nephrol. 2012; 22: 59–61.
16. Serino D, Davico C, Specchio N, Marras CE, Fioretto F. Berardinelli-Seip syndrome and progressive myoclonus epilepsy. Epileptic Disord. 2019; 21: 117–21.
17. Dubey S, Chatterjee S, Mukherjee D, Ghosh R, Sengupta S, Lahiri D, et al. "Dancing belly" in an old diabetic lady. J Family Med Prim Care. 2020; 9: 2580–2. 18.Pegg E, Tanveer R, Lekuwwa G, Mills R, Emsley H. Diabetic polyradiculoneuropathy presenting with myoclonus. J NeurolNeurosurg Psychiatry. 2016; 87: e1.
19. Zaidi SA, Chhetri SK, Lekwuwa G, Majeed T. An unusual presentation of diabetic amyotrophy: myoclonus. BMJ Case Rep. 2013; 2013: bcr2012008245.
20. Morres CA, Dire DJ. Movement disorders as a manifestation of nonketotic hyperglycemia. J Emerg Med. 1989; 7: 359–64.
21. Ghia T, Kanhangad M, Alessandri AJ, Price G, Gera P, Nagarajan L. Opsoclonus-Myoclonus Syndrome, Neuroblastoma, and Insulin-Dependent Diabetes Mellitus in a Child: A Unique Patient. Pediatr Neurol. 2016; 55: 68–70.
22. Subhankar Chatterjee et al. Faciobrachial Myoclonus as the Presenting Manifestation of Diabetic Keto-Acidosis. Tremor and Other Hyperkinetic Movements. 2021; 11(1): 9, pp. 1–5.