International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Chorea Secondary to Hyperglycemia and Chronic Kidney Disease: A Tale of Two Cases of a Rare Dance

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Abstract: ***Chorea Hyperglycemia Basal Ganglia Syndrome (CHBG) is an uncommon manifestation associated with poorly controlled nonketotic diabetes mellitus. This case reports describes a unique presentation of chorea in a patient with concomitant chronic kidney disease (CKD) and diabetes mellitus. Case - 1 involves a 49 - year - old male, while Case - 2 features a 63 - year - old male, both known cases of diabetes mellitus and CKD presenting with uncontrolled choreiform movements. While MRI findings supported the diagnosis in Case - 1, Case - 2 lacked such evidence. Both patients exhibited significant improvement after gradual control of hyperglycemia, underlining the relevance of recognizing and addressing CHBG promptly. This report contributes to the understanding of CHBG, highlights the challenges in managing chorea in the context of complex comorbidities, emphasizing early intervention and the importance of an interdisciplinary approach for effective patient care.***

Keywords: CHBG, Diabetes Mellitus, CKD, Insulin, Neuroimaging

1. Background and Aims

Chorea is defined as rapid, semi purposeful, graceful, dancelike non patterned involuntary movements involving distal or proximal muscle groups. Chorea is a rare manifestation in individuals with chronic kidney disease and diabetes mellitus but critical manifestation that requires prompt recognition and intervention. This case aims to shed light on the intricate management of chorea in the context of these concurrent chronic conditions.

These cases illustrate the need to be aware of hyperglycemia as a cause of hemiballism/ hemichorea, which is now referred to in the medical literature as C - H - BG (chorea, hyperglycemia, basal ganglia) syndrome. The precise cause of C - H - BG syndrome has not been determined.

The pathogenesis of C - H - BG syndrome is not fully understood but several theories have been suggested. Hyperglycemia impairs cerebral autoregulation, causing hypoperfusion and the activation of anaerobic metabolism and depletion of gammaaminobutyric acid (GABA) in basal ganglia neurons. GABA is the main inhibitory neurotransmitter in the basal ganglia. GABA and acetate are depleted rapidly in non - ketotic hyperglycemia, causing a reduction in acetylcholine synthesis. The hyperviscosity induced by hyperglycemia causes a disruption of the blood brain barrier and transient ischemia of vulnerable striatal neurons. The synergistic effects of uncontrolled hyperglycemia and vascular insufficiency causes an incomplete transient dysfunction of the striatum, which eventually leads to hemichorea - hemiballism in these patients. Histological findings in patients with C - H - BG syndrome have reported selective neuronal loss, gliosis, and reactive astrocytes, without evidence of hemorrhage or infarction at the striatal areas

Case 1: A 49-year male with a one - week history of uncontrolled choreiform movements affecting his left upper and lower extremity. The patient had a previous episode six months ago, lasting two days, which resolved spontaneously. With a medical history of uncontrolled type - 2 diabetes mellitus, systemic hypertension, and chronic kidney disease - diabetic nephropathy, the patient's initial glucose level was 577 mg/dl, and HbA1C was 18.0%. Neuroimaging revealed hypo - intensity on T2/FLAIR and hyperintensity on T1 in the right basal ganglia, consistent with choreoathetosis - hemiballismus (CHBG). The patient was non - compliant with diabetes medication. Treatment included insulin therapy and Tab TETRABENEZINE 25mg BD. Symptom resolution correlated with blood sugar control over 48 hours.

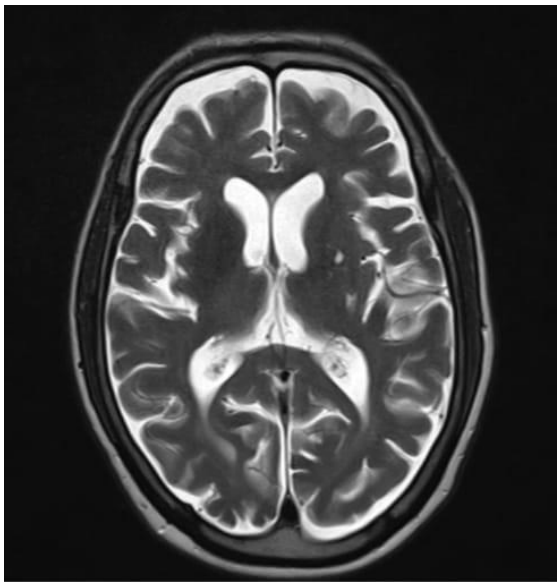

Figure 1: T2 Hypointensities seen in right basal ganglia

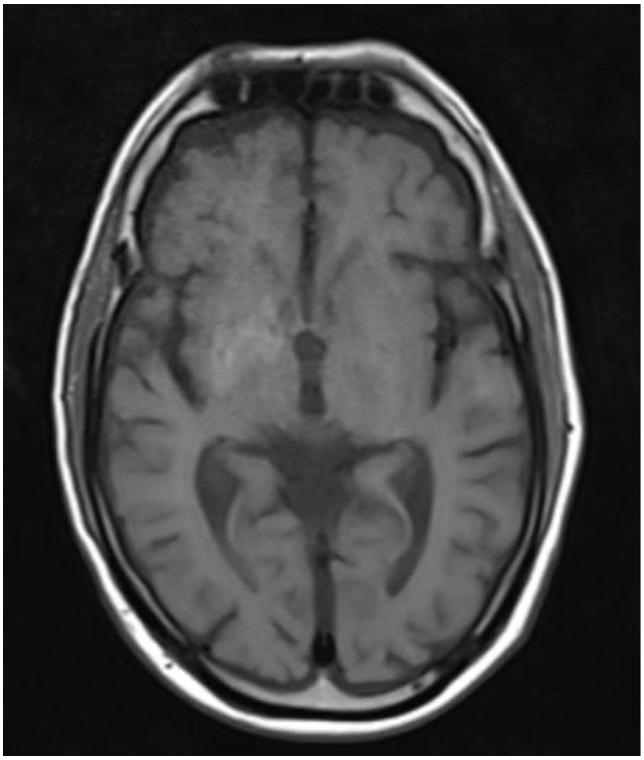

Figure 2: T1 Hyperintensities seen in right basal ganglia

Case 2: A 63-year male presented with a 3 days history of uncontrolled choreiform movements of his left upper extremity. His past medical history consisted of uncontrolled type - 2 diabetes mellitus, systemic hypertension, chronic kidney disease and old CVA with right hemiparesis secondary to left thalamus hemorrhage. He was on short acting insulin. The patient reported that he was not compliant with his diabetic medication. His initial glucose level was 740 mg/dl. HbA1C was 14.0% on the day of his admission. Vital signs were normal upon presentation, and physical exam was benign aside from the hemichorea movements of the left upper extremity. MRI BRAIN showing gliotic changes in the left thalamus secondary to old hemorrhage and micro hemorrhages in pons and left temperoparietal lobes. Inpatient treatment consisted of BASALOG 10 units every night and 30 units of HUMAN ACTRAPID 3 times a day. The patient's symptoms eventually resolved with control of his blood sugar levels. Control was achieved over roughly 7 days during which our patient's blood glucose levels dropped from an average of 740 to the 170–200 mg/dl range. The choreoathetosis progressed from continuous movements to intermittent and then finally to resolution

2. Discussion

Hyperglycemia - induced chorea is a rare neurological manifestation associated with poorly controlled diabetes mellitus (DM). It is characterized by involuntary hyperkinetic movements such as hemichorea or hemiballismus [1]. Diabetic striatopathy (DS) is defined as a hyperglycemic condition associated with either chorea/ballism or striatal abnormalities on neuroimaging [2]. This phenomenon has been reported predominantly in elderly individuals with uncontrolled DM, particularly type 2 DM [3]. Neuroimaging studies have shown characteristic findings such as unilateral or bilateral putamen involvement on CT and T1 - weighted MRI hyperintensity in the basal ganglia [4] [5]. However, it's important to note that normal brain imaging does not exclude the diagnosis of DS, as nearly half of cases may not exhibit characteristic neuroradiological stigmata [2]. The pathophysiological mechanisms underlying DS remain unclear. Chronic ischemia due to cerebral atherosclerosis combined with hyperglycemia has been proposed as an association with HC - NH (hemichorea associated with nonketotic hyperglycemia) [6]. Additionally, abnormal neuronal firing patterns in the internal segment of the globus pallidus have been suggested to be related to the pathogenesis of hemiballismus [7]. Treatment primarily involves addressing the underlying hyperglycemia. Insulin therapy alone or in combination with D2 - blockers has been effective in achieving dramatic improvement or complete resolution of chorea symptoms within days to weeks for most patients [8] [4] [9]. Symptomatic treatment using drugs like haloperidol can also be initiated if necessary [10] [11]. The prognosis for DS is generally good when prompt diagnosis and management are implemented. Patients typically experience complete clinical recovery after proper glycemic control [12]. Despite significant advancements in understanding DS over recent years, several aspects remain ambiguous and controversial. Clinical - radiological discordance is not uncommon among patients diagnosed with DS [5], emphasizing the need for further exploration through well - designed studies to better understand this condition. In summary, while DS remains a rare complication associated with uncontrolled DM primarily affecting elderly individuals, its recognition is crucial given its potential reversibility following appropriate management strategies focused on glycemic control. The coexistence of CKD and diabetes poses unique challenges in managing choreiform movements. The intricate interplay of metabolic and renal factors may contribute to the pathophysiology of chorea in this population. There is need for a collaborative, interdisciplinary approach involving nephrologists, endocrinologists, and neurologists for comprehensive patient care.

3. Conclusions

This case highlights the complexity of chorea in a patient with chronic kidney disease and diabetes mellitus. Managing choreiform movements in the presence of these comorbidities requires a holistic and collaborative approach, addressing both metabolic and renal aspects. Further research is warranted to better understand the underlying mechanisms and optimal management strategies for chorea in individuals with CKD and diabetes.

Acknowledgements
All participating authors in this study declare no financial, professional or personal conflicts. No grant support was received for this case report. All authors were involved in manuscript preparation and literature review.

References

[1] Spiegel JR I Med J. (R. I.) 105 (7): 46 - 48

[2] Wang W Medicine (Baltimore).2020 May29; 99 (22): e19801
[3] Gómez - Ochoa SA Funct Neurol Oct/Dec33; (4): 175 - 187
[4] Younes S Iran J Neurol.2014 Jul4; 13 (3): 172 - 6
[5] Hwang KJ, et. al. J Neurol, 260 Dec; (12): 2986 - 92
[6] Dwarak S Maedica (Bucur).2019 Mar; 14 (1): 59 - 62
[7] Collado - Saenz J et al., N Engl J Med.2022 Jul; 387 (3): e5
[8] Chua CB Sci Rep.2020 Jan31; 10 (1): 1594
[9] Arecco A Curr Med Res Opin Mar38; (3): 365 - 369
[10] Valle Alonso J et al., Med Clin (Barc).2023 Oct; 161 (7): 316 - 317
[11] Chen X et al. J Clin Endocrinol Metab.2023 Jul; 108 (8): e550 - e556
[12] Tsalta - Mladenov ME Curr Med Res Opin. Mar38; (3) 365 - 369