

Hemichorea in the Setting of Diabetic Striatopathy Uniquely Associated with Concurrent Myelofibrosis: A Case Report



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ABSTRACT

Hemichorea, a hyperkinetic disorder characterized by involuntary, rapid, irregular movements on one side of the body, typically originates from cortical basal ganglia involvement, particularly the striatum. We present a 63-year-old Filipino female with poor glycemic control and known idiopathic myelofibrosis exhibiting chorea-ballism movements in the right distal and proximal extremities. Significant improvement in involuntary movements was observed upon optimal glycemic control and benzodiazepine therapy. This report underscores the noteworthy presentation of uncontrolled hyperglycemia in type 2 diabetes, while highlighting the potential contribution of myelofibrosis.

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INTRODUCTION

Diabetic striatopathy (DS), also known as “hyperglycemic non-ketotic hemichorea,” presents as an acute hyperkinetic movement disorder arising from nonketotic hyperglycemia, with an estimated prevalence of 1 in 100,000. It tends to affect females more frequently with advancing age as the primary risk factor.[1]

Myelofibrosis entails persistent inflammation, triggering insulin resistance and hyperinsulinemia through the elevation of inflammatory cytokines like tumor necrosis factor- α (TNF- α) and interleukins.[2-4]

Although movement disorders are rare in myeloproliferative neoplasms (MPNs), they have been reported. Reported cases include a JAK2 V617F-positive MPN that evolved to primary myelofibrosis (PMF) with generalized chorea,[5] hemichorea as a presenting feature of polycythemia vera[6] and hemichorea-hemiballismus following basal ganglia hemorrhage in essential thrombocythemia.[7] Coexistence of PMF with type 2 diabetes has also been reported,[8] but we found no prior reports specifically linking PMF with DS. Available literature indicates that hemichorea in myelofibrosis is rare, with only a handful of comparable cases reported worldwide.[5-7] Among all reported myeloproliferative cases, hemichorea accounts for a minority of presentations. More recent reports describe hemichorea in myelofibrosis

Table 1: Capillary blood glucose trends

Date	6 AM (mg/dL)	12 NN (mg/dL)	6 PM (mg/dL)	9 PM (mg/dL)
February 7, 2024	-	297	173	176
February 8, 2024	183	195	187	177
February 9, 2024	161	217	184	-
February 10, 2024	197	271	198	-
February 11, 2024	109	197	148	-
February 12, 2024	182	283	104	-

only with secondary triggers, such as opportunistic central nervous system infection after stem-cell transplantation, suggesting that myelofibrosis alone is rarely the direct cause.[5]

In contrast, DS is a recognized but rare complication of uncontrolled hyperglycemia. However, available reports reveal no clearly documented cases combining myelofibrosis and DS presenting as hemichorea, making this coexistence particularly unusual. We therefore report a case of DS in a patient with PMF presenting with hemichorea-hemiballismus, highlighting a plausible intersection between MPN-related inflammatory and metabolic vulnerability and severe hyperglycemia.

CASE REPORT

A 63-year-old Filipino with diabetes and known idiopathic myelofibrosis presented with abnormal, continuous, abrupt, rapid, brief and irregular movements affecting her distal right upper and lower extremities. These movements progressed to flailing motions of her right upper and lower extremities, causing difficulty in performing daily activities. She has been on Ruxolitinib 5 mg per tablet, taking four tablets daily for the past eight years. On admission, she was started on thalidomide 50 mg tablet once daily and prednisone 10 mg tablet once daily.

Upon neurologic examination, there was intact cognitive function with no cranial nerve deficits. Motor and sensory examinations were unremarkable. Hyporeflexia was noted on all extremities. Following initial evaluation, the patient underwent diagnostic testing, which showed elevated fasting blood sugar levels of 562 mg/dL. Treatment was initiated; however, HbA1c levels showed 7.81%. Capillary blood glucose levels were monitored (Table 1). Magnetic resonance imaging of the brain with contrast revealed striatal T1 hyperintense signals involving the body of the left caudate nucleus and ipsilateral putamen (Figure 1).

Electromyography and nerve conduction studies were consistent with a length-dependent sensorimotor polyneuropathy, predominantly axonal.

DISCUSSION

Diabetic striatopathy is a term that describes a state of hyperglycemia linked to either or both of the following conditions: (1) acute onset chorea-ballism and (2) striatal hyperintensity or hyperdensity on T1-weighted magnetic resonance imaging (MRI) or computed tomography (CT). It manifests as involuntary, rhythmic, aimless, jerky motions involving the distal limbs. Additionally, ballism or chorea results from dysfunction in the basal ganglia and subthalamus, often presenting as larger-amplitude movements involving the proximal muscles.

Although the exact pathophysiology is still unclear, most studies suggest that DS develops in the setting of non-ketotic hyperglycemia. In non-ketotic hyperglycemia, the brain shifts to anaerobic metabolism, which inactivates the Krebs cycle and reduces levels of the inhibitory neurotransmitter GABA. This metabolic switch, together with energy depletion, can disrupt basal ganglia function and potentially lead to chorea.[1,9]

PMF, on the other hand, also known as chronic idiopathic myelofibrosis, is a condition in which normal bone marrow tissue gradually becomes replaced by fibrous scar-like material, eventually leading to progressive bone marrow failure.[2,3]

According to previous studies, PMF is associated with frequent JAK2 V617F mutation and a chronic pro-inflammatory state.[2,3] Similarly, type 2 diabetes mellitus has been linked to inflammatory signaling and insulin resistance.[4,8] Chronic inflammation is characterized by mild elevation of pro-inflammatory markers, such as TNF- and interleukin-6, which are associated with insulin resistance, diabetes, hypertension and metabolic

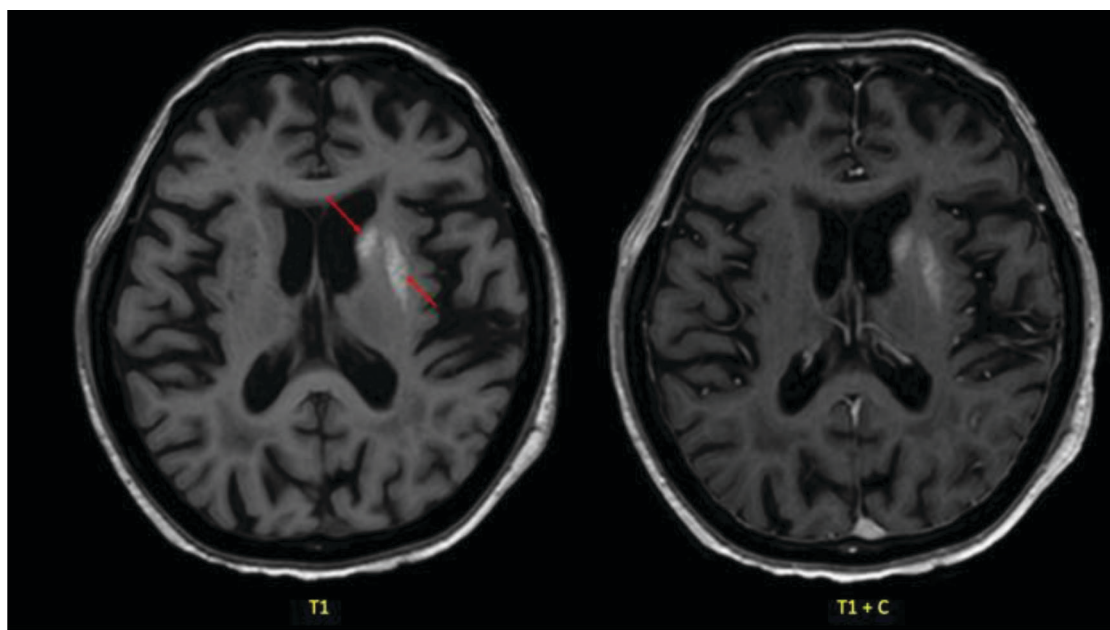


Figure 1: Contrast enhanced cranial MRI (T1 and T1 + C sequences on axial and coronal view), area of T1 hyperintense signal is seen at the body of the left caudate nucleus and left putamen.

dysregulation. The connection between DS and myelofibrosis can be potentially traced through their shared link with chronic inflammation and its impact on glucose metabolism.[3,4,8]

Firstly, DS is often associated with prolonged, poorly controlled hyperglycemia. Chronic inflammation has been implicated in the development of insulin resistance and diabetes. Inflammatory cytokines like $\text{TNF-}\alpha$ and IL-6 can interfere with insulin signaling pathways, leading to reduced glucose uptake by cells and subsequent hyperglycemia.[3,4]

Secondly, myelofibrosis-associated inflammation may exacerbate existing insulin resistance in diabetic patients, leading to difficulty in glycemic control. This perpetuates the cycle of hyperglycemia and further exacerbates DS.[4,8]

Additionally, chronic inflammation in myelofibrosis may contribute to endothelial dysfunction and vascular complications, which can exacerbate cerebral microvascular disease, potentially worsening DS.[2,3]

Overall, the chronic inflammatory milieu associated with myelofibrosis may exacerbate hyperglycemia and insulin resistance, contributing to the development and progression of DS.

CONCLUSION

This case presents a patient with chorea ballismus that significantly improved with optimized glycemic control. It also identifies the unique comorbidity of

myelofibrosis on how chronic inflammation can amplify insulin resistance, leading to poorly managed hyperglycemia and neurological complications, ultimately connecting neuroinflammation to DS.

ETHICAL CONSIDERATIONS

This study complies with the ethical principles set out in relevant guidelines as specified in the certificate of agreement and compliance in this research; as well as the National Ethical Guidelines for Research Involving Human Participants (NEGRIHP) 2022 Edition. Paper documents (ie, consent form, printouts, case tracking sheets containing identifying information) is in a locked file cabinet when not in use. These documents are handled only by the primary investigator and corresponding co-authors. Electronic confidential data stored on transportable media such as a USB, CD and portable external drive is stored securely in the said locked file cabinet as well. Documents are also password-protected and handled only by the authorized investigators and co-authors. Data will be stored for five years, after which the hard copies will be shredded and thrown away, and the soft copy files will be deleted.

INFORMED CONSENT PROCESS

A clearly written informed consent form was obtained, understood and signed by the patient and sister, a legally acceptable representative, securing

their consent for presentation, publication of the case, including other diagnostic results (cranial magnetic resonance imaging of the brain, sugar levels, hematologic blood levels). The primary investigator, not the primary physician of the patient, obtained consent. A written informed consent form in Tagalog was used to obtain consent. The authors declare that there are no conflicts of interest associated with this research.

VULNERABILITY

The subject in this research was a patient of the primary investigator. Since the patient is considered to be part of the vulnerable population, informed consent was obtained from the patient and sister by the principal investigator.

PRIVACY AND CONFIDENTIALITY

Patients' confidentiality is protected by removing patient identifiers in the case report, with full compliance to the Data Privacy Act of 2012 and its implementing rules and regulations in 2016.

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None

Authors' Contributions

Conceptualization - ICT and RLR; Data curation - ICT and RLR; Formal analysis - Not applicable; Funding acquisition - Not applicable; Investigation - ICT

and RLR, Methodology - Not applicable; Project administration - Not applicable; Resources - ICT and RLR; Software - Not applicable; Supervision - ICT and RLR; Validation - ICT and RLR; Visualization - ICT and RLR; Roles/Writing - original draft - ICT and RLR; Writing ICT and RLR. All the authors have read and approved the manuscript.

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Data Availability

All data generated or analyzed during this study are included in this published article and its supplementary information files.

Declaration of Generative AI And AI-Assisted Technologies

None.

Declaration of Interests

None of the authors has any conflict of interest to disclose.

Consent For Publication

The consent for publication of collected data is secured as part of informed consent of the participant. It was reassured to the participant that all data will be anonymized and that privacy will be upheld.

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