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A case of hemichorea-hemiballismus secondary to a subacute hemorrhage in a patient with essential thrombocythemia: a case report

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Abstract

Background Hemiballismus is a rare movement disorder characterized by violent, involuntary movements, usually resulting from a stroke affecting the subthalamic nucleus. Risperidone, an atypical antipsychotic, has shown effectiveness in managing these symptoms.

Case presentation We present the case of a 75-year-old Asian female patient of Cambodian origin with hemichorea-hemiballismus secondary to a subacute hemorrhage in the left basal ganglia, in the context of essential thrombocythemia. The patient responded remarkably well to risperidone. The patient's story, along with video documentation of her condition and response to treatment, is discussed.

Conclusion This case highlights the potential of risperidone in treating hemichorea-hemiballismus, providing a non-surgical option for managing this debilitating condition.

Keywords Hemichorea, Hemiballismus, Essential thrombocythemia, Acquired von Willebrand syndrome, Subacute intracranial hemorrhage, Dyskinesia

Background

Hemichorea-hemiballismus is a hyperkinetic movement disorder characterized by irregular, flinging movements of one side of the body, often due to lesions in the basal ganglia [1, 2]. These involuntary movements can significantly impair quality of life and require effective management strategies [3, 4]. Although surgical interventions such as deep brain stimulation are sometimes considered, pharmacological treatments, particularly with atypical antipsychotics such as risperidone, have shown promise.

Essential thrombocythemia (ET) is a hematological disorder characterized by isolated thrombocytosis, often associated with a JAK2 mutation. It is a diagnosis of exclusion, with a prevalence of 38–50 cases per 100,000 people. In rare instances, ET can result in neurological complications such as chorea [5]. Chorea is a movement disorder described as irregular and involuntary movements, while ballismus involves large, violent, involuntary movements.

Patients with myeloproliferative neoplasms (MPNs), such as ET and polycythemia vera, are at increased risk of both thromboembolic and hemorrhagic complications [6]. Among the risk factors for bleeding is the development of an acquired qualitative von Willebrand factor (VWF) defect with a loss of larger VWF plasma

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multimers, resulting in an acquired von Willebrand syndrome (aVWS). The diagnosis of aVWS is challenging because no single automated test is sufficient to prove or exclude aVWS.

Case presentation

A 75-year-old Asian female patient of Cambodian origin, with a past medical history of essential thrombocythemia (24% positivity for the JAK2V617F mutation), hypertension, and hyperlipidemia, presented with a 5-day history of involuntary movements involving the right side of her body, including the arm, leg, and to a lesser extent, the torso. These involuntary movements were described as "severe twitching," aggravated by stress and minimized with sleep and relaxation techniques. She denied headache, did not report symptoms bulbar palsy, there was no ataxia or perturbation of special senses, no impairment of bowel and bladder function, no evidence of orthostasis, and no recent infections or vaccination. She had no family or personal history of similar dyskinesias, strokes, cardiac disease, arrhythmias, diabetes, lupus, luetic or lytic processes, rheumatic fever, or infectious etiology. She confirmed prior episodes of epistaxis. She denied trauma to her head and had not been on any anticoagulants or antiplatelet medications. Her blood pressure had consistently been well controlled on lisinopril 10 mg a day for several years.

Physical examination revealed an alert and oriented woman expressing mild distress and uncontrollable movements. Her blood pressure was 120/70 mmHg. No deficits of cranial nerve function were identified. Motor tone, configuration, and strength were normal in all major muscle groups of arms and legs. Despite expressing the symptoms of a "fat tongue," her speech was clear without dysarthria. Sensory exams to touch, pinprick, vibration, and proprioception were normal and without hemineglect. Romberg was negative and her gait was unremarkable. Ear, nose, and throat (ENT) exams were normal as well.

Right-sided hemichorea with hemiballismus was most evident in the right arm, and to a lesser extent, the right leg and torso. These movements were continuous and suppressible for 1–2 s with concerted effort. There were no abnormal movements noted in her eyes, pharynx, tongue, palate, or on the left side of her body. Her deep tendon reflexes were hypoactive in the right biceps, patella, and ankle. A slight asymmetry of plantar stimulation response was noticed on the right with subtle toe extension, though not easily reproducible. Coordination testing was difficult on the right due to the superimposition of the abnormal movements, but it was normal on the left.

A magnetic resonance imaging (MRI) of her brain revealed a subacute hemorrhage involving the left basal ganglia (Fig. 1). A complete blood count at the time of presentation was remarkable, with a platelet count of 359,000/μL. As previously mentioned, the patient had a 24% positivity for the JAK2V617F mutation, and other causes of thrombocythemia were ruled out by her hematologist 3 years prior, confirming her diagnosis of ET.

After being diagnosed with ET, the patient was initially prescribed 500 mg of hydroxyurea daily. Her platelet count was then under control with hydroxyurea daily, with a peak of 806,000/μL 3 years prior. Pertinently, her thyroid panel, renal function, and rapid plasma reagin were negative. Hemoglobin A1c was 6.0%, and low-density lipoprotein (LDL) was 70 mg/dL. Workup for ischemic etiologies was deferred since her risk factors were under control and her medical history was negative. She did not undergo testing for qualitative platelet defects toward aVWS due to the complex nature of investigative and interpretation algorithms.

Treatment and outcome

To alleviate her significant dyskinesia, which interfered with her daily life, risperidone 0.25 mg twice daily was initiated. Although the patient tolerated it well, there was only a slight improvement. The dose was increased to 0.50 mg twice daily for several months, which was well tolerated, with marked attenuation of the dyskinesias. The patient's platelet count doubled, leading to an increase in her hydroxyurea dosage. Interestingly,

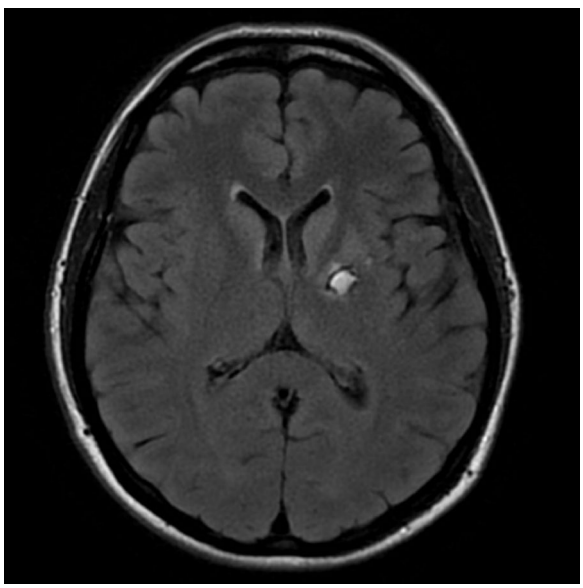

Fig. 1 Brain magnetic resonance imaging revealing subacute hemorrhage involving the left basal ganglia

5 weeks after this rise in platelet count, she developed dyskinetic movements on the right side of her body.

Over the next 4 weeks, with the escalation in the dose of risperidone to 0.50 mg twice daily, our patient's symptoms and dyskinesia improved significantly. The amplitude of her movements had diminished remarkably and was barely noticeable. Her movements now occurred intermittently every 20–60 seconds (as demonstrated in the additional file: Video). Quality of life improved significantly, and the patient was able to resume her hobbies, such as gardening, without any weakness or sensory changes. A repeat MRI of her brain 3 months later revealed a normal evolution of the intracerebral hemorrhage in the left basal ganglia region, and 3 months after the abatement of the movement disorder, risperidone was tapered but the dyskinesias returned, recurring in the right arm more than the leg. Risperidone was reintroduced at 0.25 mg twice a day and movements have since abated.

Discussion and conclusion

Hematological disorders such as essential thrombocythemia occasionally can result in hemichorea-hemiballismus. Our search was positive for only three cases of chorea, including generalized chorea and hemichorea associated with ET [6, 7]. One case report described a 55-year-old female patient with acute generalized chorea, who was also diagnosed with ET (platelet count of 1,092,000/μL) after the onset of chorea and was symptomatically treated with haloperidol [5]. In this case, the brain MRI was normal. A second case reported a 79-year-old female patient with progressive worsening of chorea affecting the face and jaw [8]. However, this patient was already diagnosed with ET (JAK2V617F mutation-positive) and on hydroxyurea treatment, with a platelet count of 341,000/μL. The third case report reported a 68-year-old female patient that developed acute right-sided hemichorea in the setting of essential thrombocythemia (platelet count of 1,883,000/μL), and a MRI showed a lesion affecting the left basal ganglia that were attributed to either microcirculatory failure or small cerebral hemorrhages [9]. The severity of thrombocytosis varied greatly between the cases, ranging from 341,000/μL to 1,883,000/μL.

The pathophysiological association between ET and hemichorea-hemiballismus remains unclear, though several mechanisms have been proposed. ET is characterized by an increased platelet count, which can lead to a hypercoagulable state. This predisposes patients to microthrombus formation, potentially causing ischemic damage by occluding small cerebral vessels, particularly those supplying the basal ganglia. Even in the absence of microthrombi, chronic hypoperfusion in the basal ganglia, resulting from increased blood viscosity and reduced cerebral blood flow, can lead to metabolic dysfunction and the development of movement disorders [10]. Additionally, ET is associated with a systemic inflammatory response, marked by elevated levels of proinflammatory cytokines such as interleukin (IL)-4, IL-8, IL-10, platelet-derived growth factor, and vascular endothelial growth factor [11]. These cytokines can contribute to neuroinflammation, further implicating basal ganglia dysfunction in the pathogenesis of hemichorea-hemiballismus. Moreover, excessive platelets in ET may locally adhere, aggregate, and release neurotoxic substances, potentially altering neuronal signaling within the basal ganglia [12].

ET is also associated with a dual propensity for thrombosis and bleeding. Hemorrhage involving the basal ganglia, particularly near the subthalamic nucleus, can result in movement disorders such as hemichorea-hemiballismus [6, 13]. In the case described, subacute hemorrhage involving structures adjacent to the subthalamic nucleus likely contributed to the symptoms, occurring in the context of a reasonably well-controlled platelet count, hypertension, and hyperlipidemia.

The association of involuntary motor movements and stroke are relatively common, including the manifestation of movement disorders, and extreme thrombocytosis can increase the risk of bleeding. One study of 74 young women with ET showed that 3 patients (4%) had a major hemorrhagic event [14]. In our case, our patient's blood pressure (120–130 s systolic) and lipids (LDL 70 mg/dL) were under control with medications (lisinopril-hydrochlorothiazide and atorvastatin), and importantly, she was not on any anticoagulants or antiplatelet agents. Because our patient's hypertension, hyperlipidemia, and essential thrombocythemia were well controlled, it is possible her hemorrhagic insult could be due to an acquired platelet defect. Studies have shown that patients with MPNs, such as ET, may develop a loss of large VWF multimers, resulting in aVWS and a paradoxical increased risk of bleeding [6, 15–17]. Interestingly, our patient also had confirmed prior episodes of epistaxis with normal blood pressure and ENT exams. We did not conduct aVWS tests, since a diagnosis of VWS is challenging, with no single automated test being sufficient to prove or exclude aVWS.

The treatment of hemichorea-hemiballismus typically includes dopamine receptor blockers such as haloperidol and perphenazine, antipsychotics from both first and second generations (for example, risperidone and clozapine), and catecholamine-depleting agents such as reserpine and tetrabenazine [18]. In this case, risperidone was selected not only for its demonstrated efficacy in managing severe hemichorea-hemiballismus, but also

for its unique advantages over alternative treatments. First, while sharing a similar mechanism of action with other dopamine receptor blockers, risperidone selectively targets D2 receptors with fewer extrapyramidal side effects, especially at lower doses, compared with first-generation antipsychotics. Second, risperidone has a more favorable side effect profile than older medications such as haloperidol, with reduced risks of sedation and cognitive impairment, making it more suitable for long-term use [19]. Additionally, risperidone's high affinity for 5-HT2A receptors provides added efficacy in managing psychiatric comorbidities such as anxiety and depression, which are often associated with movement disorders or post-stroke conditions, as seen in our patient [20]. Furthermore, the adverse effects commonly associated with alternative treatments for hemichorea-hemiballismus, such as depression and sedation with tetrabenazine or agranulocytosis with clozapine, underscore the advantages of risperidone as an optimal choice in this context [21].

Our case shows that patients with reasonable platelet control and excellent control of hypertension and hyperlipidemia may still develop neurological insult, with limited reported cases in literature [5, 8–10]. In our case, intracerebral hemorrhage likely contributed to the dyskinesias, with risperidone significantly alleviating symptoms and enhancing the patient's quality of life.

Abbreviations

aVWS	Acquired von Willebrand syndrome
ENT	Ear, nose, and throat
ET	Essential thrombocythemia
LDL	Low-density lipoprotein
MPN	Myeloproliferative neoplasms
MRI	Magnetic resonance imaging
VWF	Von Willebrand factor
IL	Interleukin

Supplementary Information

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Additional file 1.

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Availability of data and materials
All data generated during this study can be accessed through direct communication with the corresponding author and the agreement of all research team members.

Declarations

Ethics approval and consent to participate
Ethics approval is not required for this type of study. Written consent to participate in this study was obtained from the patient.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that there is no competing interest to be reported.

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