Case Report

A Case of Hemichorea Caused by Cerebral Cavernous Angioma

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Chorea is a type of hyperkinetic movement, referring to involuntary, irregular, aimless, nonrhythmic, abrupt, rapid, unsustained movements. The term hemichorea refers to chorea of one side of the body. A 39-year-old woman presented with a three-week history of abrupt, involuntary movements of her right hand and right foot, which was compatible with hemichorea of the right side. MRI brain showed a 1.7x1.5x1.3 cm lesion in the posterior limb of the left internal capsule and the lateral part of the left thalamoganglionic region, which was seen as mixed iso-and hyperintense in T1W images, and heterogeneous and hyperintense in T2W/ FLAIR images. T2 gradient sequences showed a peripheral rim of decreased signal intensity, which is the hemosiderin ring, and no significant brain edema. Partial contrast enhancement of the lesion after contrast media injection suggested cavernous angioma. After medical treatment with haloperidol 2.5 milligrams per day, her symptom gradually improved within 2 months.

***Keywords**: Chorea, Choreiform movement, Hemichorea, Cavernoma, Cavernous angioma*

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Chorea is a type of hyperkinetic movement. It refers to involuntary, irregular, aimless, nonrhythmic, abrupt, rapid, unsustained movements that start in one part of the body and often continues to another part. Patients can partially and temporarily suppress the chorea, which may subside during sleep[1,2]. Chorea may be characterized as primary or genetic in origin, such as Huntington's disease, or may be secondary to infectious, post-infectious, or immunological process, tumors, vascular lesions, and pregnancy.

Other causes include certain drugs, toxins, hyperthyroidism, hypoparathyroidism with hypocalcemia, hyperglycemia, hypoglycemia, hypernatremia, hyponatremia, hypomagnesemia, hypocalcemia, and hepatic or renal failure[3,4]. In this report we discuss the clinical correlation of a case which presented with unilateral chorea, and the association of neuroanatomy, pathophysiology of the movement circuit, and brain imaging findings.

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Case Report

A 39-year-old married woman presented with a three-week history of sudden onset of involuntary movements in her right hand and foot which improved during sleep, consistent with the hemichorea. She had no significant past medical history and was a nonsmoker without any history of drug or alcohol abuse. She denied previous stroke or head trauma. Her blood pressure was 126/88 mmHg, and her pulse rate was 84/minute. Heart sounds were normal. Both eyes were negative for Kayser-Fleischer rings (KF rings). Neurological examination revealed good consciousness, right-sided periodic choreiform movement, normal memory and behavioral functions. Cranial nerves, muscle tone and power, deep tendon reflexes, and sensation were normal. Investigations showed Hb 13 gm%, WBC 6,520 cell/mm^3, platelet 400,000/mm^3, blood glucose 90 mg/dl. Serum electrolytes, liver function test, creatinine, calcium, magnesium and thyroid function test were normal. HIV antibody and VDRL titer were nonreactive.

The contrasted magnetic resonance imaging (MRI) of the brain (Fig. 1) showed a 1.7x1.5x1.3 cm lesion in the posterior limb of the left internal capsule and the lateral part of the left thalamoganglionic region. This was visualized as mixed iso- and hyperintense in

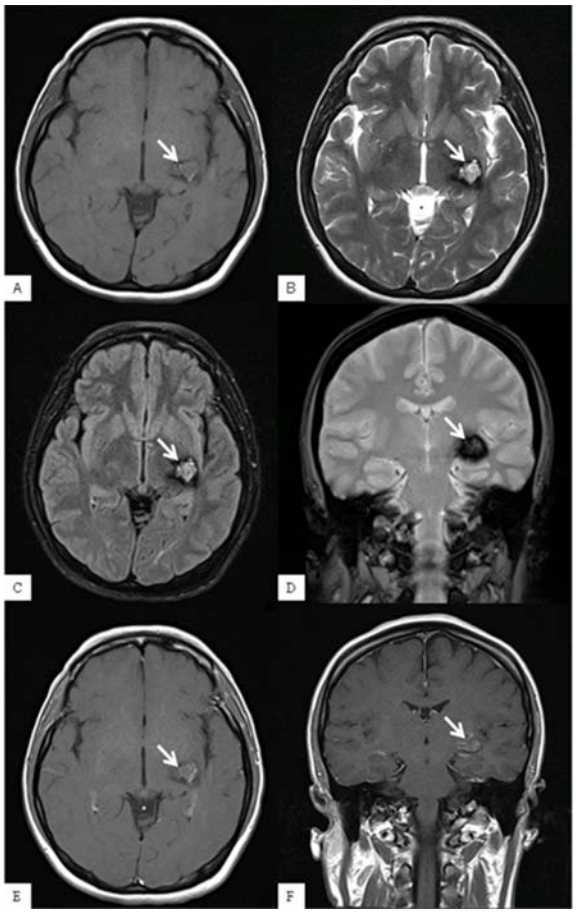

Fig. 1 Contrasted MRI brain showed a 1.7x1.5x1.3 cm of "popcorn-like" or "raspberry-like" appearance, suggesting a cavernous angioma (arrow) in the posterior limb of the left internal capsule and the lateral part of the left thalamoganglionic region. (A) axial T1W, (B) axial T2W, and (C) FLAIR imaging. Coronal T2GRE (D) showed a peripheral hemosiderin ring and partial enhancement after gadolinium injection (E and F).

T1-weighted (T1W), while being heterogeneous and hyperintense in T2-weighted (T2W) and fluid-attenuated inversion recovery (FLAIR) sequences. T2 gradient sequence (T2GRE) showed a peripheral rim of decreased signal intensity, which was the blooming hemosiderin ring, and no significant brain edema. Partial enhancement of the lesion after contrast media injection did not identify any nidus or feeding vessels. This MRI finding revealed a "popcorn-like" or "raspberry-like" appearance that suggested a cavernous angioma or cavernoma. The patient refused surgical management because the vascular lesion was located in the deep area of the thalamic region. After treatment with haloperidol 2.5 milligrams per day, her symptom gradually improved within 2 months.

Discussion

Diagnosis of the chorea is based on history and clinical findings, including age of onset, time course (acute or insidious), distribution of chorea (focal/hemichorea or generalized), past medical history, history of recent infection such as Group A beta hemolytic streptococcus infection, HIV infection, history of meningitis, encephalitis, drugs/toxins exposure, and family history of abnormal movement or neurodegenerative diseases[1-4].

Laboratory studies and brain imaging are useful for distinguishing secondary causes of chorea from primary causes. Laboratory investigations which are useful for the differential diagnosis of chorea include complete blood count and red blood cell morphology, serum glucose, electrolyte, calcium, magnesium, phosphorus, liver, renal, and thyroid function tests, HIV antibody, and VDRL titer. Erythrocyte sedimentation rate (ESR) and antinuclear antibody titer would be useful in cases suspected of autoimmune diseases. Genetic studies might be indicated in cases with family history of Huntington's disease or neuropsychiatric disease, whereas serum ceruloplasmin and 24-hour urine copper are recommended as screening tests for Wilson's disease, especially in young-onset chorea or other abnormal movements and/or positive family history of neuropsychiatric diseases and history of liver disease. Brain imaging should be done to detect intracranial structural lesions, such as tumors, vascular lesions, including ischemic or hemorrhagic lesions, and vascular malformations[3,4].

MRI brain of this patient revealed a lesion with mixed iso- and hyperintensity in T1, and heterogeneous hyperintensity in T2/FLAIR imaging. T2GRE sequence showed a peripheral rim of hemosiderin. The finding of "popcorn-like" or "raspberry-like" appearance and partial enhancement after contrast injection strongly supports the diagnosis of cavernous angioma or cavernoma in the posterior limb of the left internal capsule and the lateral part of the left thalamoganglionic region[5,6]. This locality was thought to be consistent with hemichorea. Regarding the pathophysiology and pathogenesis of this structurally-related abnormal movement disorder, lesions may involve the caudate nucleus, putamen, external and internal segments of the globus pallidus

externa (GPe), globus pallidus interna (GPi), subthalamic nucleus, substantia nigra, or interconnecting pathways. The abnormal movement results from damage or dysfunction of these structures, which causes an imbalance between indirect and direct movement pathways in the basal ganglia circuit. Disruption of the indirect pathway with loss of inhibition to the globus pallidus allows hyperkinetic movements, including chorea, to occur[3,7].

The disorders of the basal ganglia circuit may be due to structural damage, neuro-degeneration[8], neurotransmitter receptor blockade, electrolyte or metabolic disturbance, and autoimmune conditions[9-12]. An important etiology of chorea is vascular lesions, the most common being ischemic or hemorrhagic cerebrovascular diseases and vascular malformations, such as venous angiomas, arteriovenous malformations and cavernous angiomas[13-17]. Cerebral cavernous angiomas may be asymptomatic and often found incidentally. Common symptoms of cerebral cavernous angiomas may result from acute hemorrhage, and include headache, focal seizures, complex-partial and generalized seizures followed by focal neurological deficits, and rarely, abnormal movements[3,5,6,9,18-23].

Cavernous angiomas causing chorea have been rarely reported. Previously reported patients varied in age of onset and clinical course[5,13,18-23]. Two cases were of childhood-onset[18,19], while other patients were older compared with our case[20-23]. In all cases, the clinical presentation was described as hemichorea. There was only one patient, a 77-year-old man, who presented with choreiform movements and behavioral disturbance[22]. Brain imaging of these cases showed a vascular lesion localized in the caudate nucleus[13,18,19,22], lentiform nucleus and subcortical structures[20,21,23]. Barut et al reported a case of an elderly woman with hemichorea related to a cavernous angioma in the posterior crus of the left internal capsule, sublentiform and retrolentiform region, and lateral left thalamus[5], which is similar to the brain imaging of our case.

Treatment of chorea depends upon the etiology[1-4]. For example, in drug-induced chorea, cessation of that particular drug may help. Wilson's disease is treatable and potentially curable by liver transplantation and the use of copper chelating agents, of which penicillamine is the first line therapy. For other types of chorea, dopaminergic antagonists are effective. Typical neuroleptics include haloperidol and fluphenazine. Atypical neuroleptics, such as risperidone, olanzapine, clozapine and quetiapine may have less potential than typical neuroleptics to induce parkinsonism. GABAergic drugs, such as clonazepam, valproate, and gabapentin, can be used as adjunctive medications[2,3]. Drugs that reduce the amount of dopamine release, dopamine-depleting agents, such as reserpine and tetrabenazine may also help, but the improvement may be limited[3,8]. Regarding treatment of cavernous angioma-related hemichorea, most cases were controlled by pharmacological treatment, although some were intractable[5,20]. Complete surgical resection of the lesion also led to resolution of the abnormal movement[19,21]. In one case, the hemichorea improved spontaneously[13].

Conclusion

We present a rare case of chorea associated with cerebral cavernous angioma. Although our patient was treated conservatively, antipsychotic medication could control the abnormal movement. Neuroimaging is an important part of the work up of chorea because it may reveal potentially treatable or reversible etiologies. Treatment depends on the type of chorea and associated diseases.

What is already known on this topic ?

Cerebral cavernous angiomas are usually clinically silent. However, headache, seizures, focal neurological deficits or abnormal movements may manifest when the lesion is complicated by hemorrhage.

What this study adds ?

This study adds the information that the brain imaging is mandatory in the work up cause of any abnormal movements because it may demonstrate intracranial structural lesions.

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Potential conflicts of interest

None.

References

1. Kumar A, Calne DB. Approach to the patient with a movement disorder and overview of movement disorders. In: Watts RL, Koller WC, editors. Movement disorders: neurologic principles and practice. 2nd ed. New York: McGraw-Hill; 2004: 3-13.

2. Fahn S, Jankovic J, Hallett M. Chorea, ballism, and athetosis In: Fahn S, Jankovic J, Hallett M, editors. Principles and practice of movement disorders. 2nd ed. Philadelphia:Saunders Elsevier; 2011: 335-49.
3. Cardoso F, Seppi K, Mair KJ, Wenning GK, Poewe W. Seminar on choreas. Lancet Neurol 2006; 5: 589-602.
4. Bhidayasiri R, Truong DD. Chorea and related disorders. Postgrad Med J 2004; 80: 527-34.
5. Barut BO, Demiryurek BE, Turksoy O. Choreiform Movement Disorder Secondary to Intracerebral Cavernoma: Case Report. Turk J Neurol 2013; 19: 60-2.
6. Raychaudhuri R, Batjer HH, Awad IA. Intracranial cavernous angioma: a practical review of clinical and biological aspects. Surg Neurol 2005; 63: 319-28.
7. Mink JW. The Basal Ganglia and involuntary movements: impaired inhibition of competing motor patterns. Arch Neurol 2003; 60: 1365-8.
8. Grove VEJr, Quintanilla J, DeVaney GT. Improvement of Huntington's disease with olanzapine and valproate. N Engl J Med 2000; 343: 973-4.
9. Alp R, Alp SI, Ure H. Cavernous hemangioma: a rare cause for secondary parkinsonism: a case report. Int J Neurosci 2009; 119: 2112-7.
10. Piccolo I, Defanti CA, Soliveri P, Volonte MA, Cislaghi G, Girotti F. Cause and course in a series of patients with sporadic chorea. J Neurol 2003; 250: 429-35.
11. Wintermark M, Fischbein NJ, Mukherjee P, Yuh EL, Dillon WP. Unilateral putaminal CT, MR, and diffusion abnormalities secondary to nonketotic hyperglycemia in the setting of acute neurologic symptoms mimicking stroke. AJNR Am J Neuroradiol 2004; 25: 975-6.
12. Oh SH, Lee KY, Im JH, Lee MS. Chorea associated with non-ketotic hyperglycemia and hyperintensity basal ganglia lesion on T1-weighted brain MRI study: a meta-analysis of 53 cases including four present cases. J Neurol Sci 2002; 200: 57-62.
13. Kobayashi K, Aoyama N, Sasaki J, Oshima H, Fukaya C, Yamamoto T, et al. MRI appearance of a cerebral cavernous malformation in the caudate nucleus before and after chorea onset. J Clin Neurosci 2011; 18: 719-21.
14. Larrosa D, Ramon C, Santamarta E, Zeidan N, Pascual J. Hemichorea secondary to contralateral pontine haemorrhage. Parkinsonism Relat Disord 2013; 19: 271-2.
15. Chang MH, Chiang HT, Lai PH, Sy CG, Lee SS, Lo YY. Putaminal petechial haemorrhage as the cause of chorea: a neuroimaging study. J Neurol Neurosurg Psychiatry 1997; 63: 300-3.
16. Mestre TA, Ferreira JJ, Pimentel J. Putaminal petechial haemorrhage as the cause of non-ketotichyperglycaemic chorea: a neuropathological case correlated with MRI findings. J Neurol Neurosurg Psychiatry 2007; 78: 549-50.
17. Krauss JK, Kiriyanthan GD, Borremans JJ. Cerebral arteriovenous malformations and movement disorders. Clin Neurol Neurosurg 1999; 101: 92-9.
18. Yakinci C, Durmaz Y, Korkut M, Aladag A, Onal C, Aydinli M. Cavernous hemangioma in a child presenting with hemichorea: response to pimozide. J Child Neurol 2001; 16: 685-8.
19. Carpay HA, Arts WF, Kloet A, Hoogland PH, Van Duinen SG. Hemichorea reversible after operation in a boy with cavernous angioma in the head of the caudate nucleus. J Neurol Neurosurg Psychiatry 1994; 57: 1547-8.
20. Kuwahara H. Images in clinical medicine. Persistent hemichorea. N Engl J Med 2013; 368: e26.
21. Zabek M, Sobstyl M, Dzierzecki S, Gorecki W, Jakucinski M. Right hemichorea treated successfully by surgical removal of a left putaminal cavernous angioma. Clin Neurol Neurosurg 2013; 115: 844-6.
22. Carella F, Caraceni T, Girotti F. Hemichorea due to a cavernous angioma of the caudate. Case report of an aged patient. Ital J Neurol Sci 1992; 13: 783-5.
23. Donmez B, Cakmur R, Uysal U, Men S. Putaminal cavernous angioma presenting with hemichorea. Mov Disord 2004; 19: 1379-80.

รายงานผู้ป่วยที่มีการเคลื่อนไหวแบบ chorea ที่บริเวณร่างกายครึ่งซีกที่มีสาเหตุจากเส้นเลือดดำที่ผิดปกติในเนื้อสมอง (Cavernous angioma)

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Chorea เป็นลักษณะการเคลื่อนไหวที่ผิดปกติชนิดการเคลื่อนไหวแบบเร็วประเภทหนึ่ง ซึ่งมีการเคลื่อนไหวที่เป็นลักษณะที่เกิดไม่สม่ำเสมอ ไม่เป็นจังหวะ เกิดขึ้นทันทีอย่างรวดเร็วและไม่ต่อเนื่อง โดยที่ผู้ป่วยไม่สามารถควบคุมอาการที่เกิดขึ้นได้ อาการเกิดได้ที่บริเวณร่างกายครึ่งซีกที่เรียกว่า hemichorea รายงานนี้บรรยายถึงผู้ป่วยหญิงอายุ 39 ปีที่มีการเคลื่อนไหวที่เกิดขึ้นอย่างรวดเร็วทันทีและไม่สามารถควบคุมบังคับได้ที่มือและเท้าข้างขวานาน 3 สัปดาห์ก่อนมาพบแพทย์ อาการเข้าได้กับการเคลื่อนไหวผิดปกติแบบ chorea ครึ่งซีกของลำตัวข้างขวา ผลการตรวจสมองด้วยคลื่นแม่เหล็กไฟฟ้าพบรอยโรคขนาด 1.7x1.5x1.3 เซนติเมตร ที่สมองด้านซ้ายบริเวณ internal capsule และสมองส่วน thalamus โดยมีลักษณะที่เข้าได้กับเส้นเลือดดำที่ผิดปกติในเนื้อสมองที่เรียกว่า cavernous angioma ผู้ป่วยได้รับการรักษาโดยรับประทานยา haloperidol ขนาด 2.5 มิลลิกรัมต่อวัน อาการค่อยๆ ดีขึ้นจนหายเป็นปกติภายใน 2 เดือนหลังได้รับการรักษา