



Hemichorea Revealing Type 2 Diabetes about a Presentation

Halladain Mpung Mansoj*, Anna Modgi BASSE, Adjaratou Dieynabou Sow and Moustapha Ndiaye

Department of Neurology, Fann Teaching hospital, Dakar, Senegal

*Corresponding author: Halladain Mpung Mansoj, Department of Neurology, Fann Teaching hospital, Dakar, Senegal

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Introduction

Neurological complications of diabetes mellitus, such as peripheral neuropathy and stroke, are common. However, hemichorea is a rare complication seen in less than 1 in 1,000,000 diabetics [1]. Additionally, chorea associated with nonketotic hyperglycemia usually occurs in older people, women, and people of Asian descent [2]. The first case of nonketotic hyperglycemia hemichorea was first described by Bedwell et al. in 1960 [3]. Hence the interest of our case.

Report Case

An 80-year-old woman presented to our neurology department with involuntary movements of her upper and lower right limbs. Movements increased with action, decreased with relaxation, and disappeared during sleep. The patient had no medical history of diabetes and hypertension. No history of abnormal movements

or other neurological disorders was reported. On admission, she was conscious and well oriented in time and space. A neurological physical examination revealed normal muscle tone and strength on both sides; . There were no signs of sensory impairment and the cranial nerves were normal. Deep tendon reflexes were Laboratory tests revealed poorly controlled diabetes mellitus, with fasting blood sugar of 4g/dl and hemoglobin A1C of 11.0%. Urine examination was negative for ketone bodies. Shortly after her consultation, the patient's blood sugar was checked with insulin; this led to a steady correction of blood sugar in the following days. On admission, the patient had a non-enhanced cerebral computed tomography (CT) scan, and the result was normal (Figure 1). The treatment made of haloperidol gout as well as metformin was instituted with good clinical improvement for weeks and then the patient returned to appointments for her treatment.



Figure 1

Discussions

Nonketotic hyperglycemia hemichorea is a rare syndrome characterized by the triad of chorea, hyperglycemia and basal ganglia damage [4]. Typically, patients with this symptom have long, poorly controlled diabetes [5]. Our case had no history of diabetes. Previous literatures [6] has shown that abnormal movements may be early manifestations of undiagnosed diabetes. The exact pathogenesis of this syndrome is unclear. Several hypotheses have been proposed to provide an explanation for this condition. Nonketotic hyperglycemia may be one of the possible mechanisms involved in the genesis of hemichorea. In hyperglycemia, cerebral metabolism switches to the anaerobic pathway with inactivation of the tricarboxylic acid (Krebs) cycle [7]. To provide an alternate energy source, the brain metabolizes GABA into succinic acid. However, such a GABA shunt provided 10-40% of the energy required by the basal ganglia, which leads to metabolic acidosis.

Unlike in ketotic hyperglycemia, where GABA can be resynthesized, GABA and acetate are depleted rapidly in nonketotic hyperglycemia [7]. As a result, acetylcholine synthesis is reduced by acetate depletion. A reduction in both GABA and acetylcholine in the basal ganglia, together with metabolic acidosis and lack of energy production, can then lead to basal ganglia dysfunction and subsequent chorea. However, with this hypothesis, it is difficult to explain the persistence of chorea after normalization of blood sugar. Additionally, chorea can also occur with hypoglycemia [8,9] and ketotic hyperglycemia [10]. Some patients develop chorea after rapid correction of hyperglycemia [11-13]. Most patients with Chorea, hyperglycemia, basal ganglia syndrome (CH-BG) in a diabetic patient develop hemichorea. It is unusual for dyskinesia to be caused by systemic metabolic disorders. Taken together, these results suggest that nonketotic hyperglycemia itself may not be the sole mechanism responsible for the development of chorea. This hypothesis seems to be for our patient who had only nonketotic hyperglycemia.

Ischemia as a possible mechanism Ischemia may be a possible mechanism of NKH chorea. Kim [14] reported 4 cases of hemichorea with acute cerebral infarction. Suzuki [15] reported a 66-year-old woman with diabetes mellitus who suddenly developed right hemichorea. However, this theory is difficult to explain the presence of chorea in some patients. Microhemorrhage as a possible mechanism High signal intensity of T1-MRI has been reported in many cases, [16] suggesting that it could be caused by bleeding. But pathological and autopsy studies of chorea [17] found s-selective neuronal loss, glial hyperplasia, and reactive astrocytosis, but no significant hematoma was reported. Therefore, the pathogenesis of this disease requires further study. However, not all of the theories above have been backed by solid evidence but would explain many obscure aspects of this symptom. More experiments and histopathology studies on large series of patients are needed. The neuroradiological findings of NCNH are unique. CT scans usually show hyper attenuation in the striatum contralateral

to the affected side, with no signs of mass effect, edema, or volume loss. The typical result of MRI is hyperintensity on weighted images, this assumption does not correlate with our patient who had normal brain imaging.

Clinical Manifestation

Oh [18] mentioned in his research that out of 53 patients with chorea, hyperglycemia, basal ganglia syndrome (CH-BG) in a diabetic patient, 6 were chorea B (11.4%) and the other 47 were hemichorea (88.6%). Fourteen (26.4%) patients had facial involvement. Most patients' symptoms improved during sleep. In addition to chorea, some patients had other neurological deficits, such as a pyramidal sign, transient dyskinesia, hemifacial muscle spasm and dystonia [19].

Treatment and Prognosis

The most common treatment is blood sugar control therapy. Based on blood sugar control, HPD is used as monotherapy or in combination with other medications to control symptoms. Most patients receiving sugar control and HPD treatment of dance symptoms will be significantly improved or even gone. A few serious patients, in addition to hypoglycemic therapy, can also use various drugs (tipride, chlorpromazine, diazepam). For patients with refractory chorea, [20] they can be treated by ventral lateral thalamotomy. Some patients relapsed after stopping HPD, but relapsed patients responded well to piperazine and valproic acid [21]. Our patient had benefited from haloperidol drops for the treatment of abnormal movements who completely recovered. And rapid glycemetic control with parenteral monotherapy made of metformin 500 mg morning and evening. The prognosis of NCNH has been reported to be excellent [22], and it was indeed excellent in our patient as well. Hyperglycemia needs to be quickly controlled.

Conclusion

Hemichorea is a rare complication of hyperglycaemia. Neuroradiological images show typical features. Physicians should be aware that hemichorea may be the first manifestation of undiagnosed diabetes so that proper diagnosis and treatment can be initiated.

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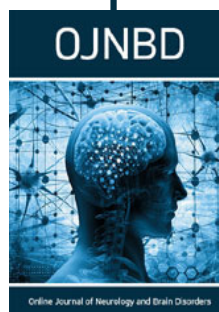
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