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Diabetes & its Complications

Nonketotic Hyperglycemic Hemichorea Hemiballism as the Debut of Diabetes in An 80-Year-Old Woman: Case Report

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ABSTRACT

Diabetes Mellitus is a chronic metabolic disease that constitutes one of the most serious public health problems worldwide. With the advent of new drugs and advances in the understanding of its etiopathogenesis, the quality of life of patients suffering diabetes mellitus has been improved. However, the large number of complications to which patients are exposed remains a challenge for health professionals, being the most frequent from cardiovascular nature, renal, retinal, peripheral nervous system and even increasing the risk of mortality.

Not much has been documented about the consequent relationship of movement disorders as a result of metabolic alterations, although cases of glucotoxicity and hyperkinetic disorders such as chorea and ballism have been described.

In the present article we describe the case of an 80-year-old woman with diabetes mellitus in poor metabolic control, associated with hemichorea and ballism.

Keywords

Hemichorea, Hemiballism, Diabetes mellitus, Hyperglycemia.

Introduction

Hemichorea / hemiballism (HH) is one of the hyperkinetic movement disorders characterized by acute, subacute or even chronic beginning of large amplitude, unpredictable, meandering and ataxic involuntary movements that affect body fluidity which can be hemicorporal or generalized [1,2]. The word chorea means dance, and emphasizes irregular, jerky, quick, brief and non-sustained involuntary movement. Choreic movements may appear at rest, are partially suppressible at willingness, and increase with stress and distracting maneuvers. They exhibit dramatic improvement during sleep [3]. On the other hand, ballism consists of proximal and broad movements, with more aggressive character. It frequently occurs unilaterally (hemiballismus), and although it may appear at rest, it tends to be exacerbated by movement [4].

The pathophysiology of this syndrome remains controversial. It is likely that a combination of hyperglycemia-induced metabolic alterations in the basal ganglia and failure of cerebral blood flow autoregulation contribute to the syndrome. A small number of histopathologic studies have found gliosis, swollen reactive astrocytes, and selective loss of neurons, without evidence of hemorrhage or infarction [5]. It is usually find and demonstrate lesions at the basal ganglia level that may be the result of different types of injury such as infection, genetic mutation, neoplasms, neurodegenerative disorders, stroke, metabolic disease, drug exposure and autoimmune disease. Hemichorea hemiballism describes such movements at the unilateral or hemibody level. In the case of association with metabolic disorders such as nonketotic hyperglycemia it is a rare complication of diabetes mellitus, wich is observed especially in elderly women with poor blood glucose control. It was described in 1960 and was characterized by non-ketotic hyperglycemia, unilateral involuntary choreiform movements and is currently known from hyperintensity of contralateral basal ganglia on T1-weighted MR images or high density on CT scans [6].

Several pathophysiological mechanisms have been proposed to explain hyperintensity on T1-weighted MRI sequences, including acute ischemia, petechial microhemorrhage, injury secondary to hyperviscosity, osmotic injury, and vasogenic edema [12,13] Hyperviscosity is currently suggested to be the most plausible mechanism due to the following findings: elevated serum osmolarity at the time of HH, variable T2-weighted MR signal changes reflecting the difference in patterns and severity of hyperviscosity, and elevated myoinositol and choline levels [7].

The diagnosis is made with findings of typical clinical and radiological features, including the presence of ballistic or choreiform movements in the context of marked hyperglycemia and the absence of ketoacidosis [8].

Determining the etiology of the condition is a matter of utmost importance and correcting non-ketotic hyperglycemia along with long-term diabetic control is the mainstay of treatment [9]. In cases of prolonged diabetic HH, dopamine receptor blocking agents (chlorpromazine or haloperidol), neuroleptics (olanzapine, clozapine) and dopamine-lowering agents (tetrabenazine) have been documented to be beneficial [10,11].

However, elderly patients are especially susceptible to developing neuroleptic-induced extrapyramidal syndrome. Anticonvulsants may also be useful for the treatment of hemiballismhemichorea syndrome. Topiramate is a broad-spectrum anticonvulsant widely used for adult and pediatric epilepsy, and has been reported to benefit hyperkinetic movement disorders, particularly essential tremor because of its GABAergic properties. Its clear advantage over neuroleptics is the absence of extrapyramidal syndrome with long-term use. Therefore, it may be reasonable to try topiramate even before neuroleptics are instituted [14]. The prognosis of hemichorea induced by non-ketotic hyperglycemia such as diabetic ketoacidosis is favorable [15] when hyperglycemia is detected and corrected, the movement disorder in most cases improves in a few days or weeks. The course of hemichorea hemiballism caused by hyperglycemia, whether ketosis or not, can be reversible and have a good prognosis [16,17].

Case Description

An 80-year-old female patient with a history of coronary artery disease with myocardial revascularization surgery, arterial

hypertension, dyslipidemia and smoking, who was hospitalized for clinical symptoms of polyuria, polydipsia and weight loss with glycemia found at 500 mg/dl at admission, finally generating a diagnosis of de novo Diabetes Mellitus at discharge.

Two months later, she presented clinical symptoms of sudden onset consisting of involuntary movements in the right hemibody, which disappear during sleep and interfered with the patient's quality of life. On physical examination she was found alert, conscious, oriented in time, place and person, fluent, coherent, without alteration in mental functions, without neurological focality in cranial pairs, however, irregular, uncontrollable, non-stereotyped, not very predictable, serpentiginous movements, moderate amplitude, with distal predominance, at the right brachicrural level were documented. On the other hand, the left hemibody showed no abnormal movements, preserved muscle tone and strength, in addition to presenting normal myotendinous reflexes.

Considering the mentioned medical history and the clinical presentation, complementary studies were requested. Cobalamin deficiency was found and a Nuclear Magnetic Resonance Imaging (NMR) in which a hyperintense lesion was observed in T1-weighted sequence located in basal ganglia, at the level of the head of the caudate nucleus extending to the left lenticular nucleus, describing other sequences (Figure 1 and 2). Being diagnosed by semiology and complementary studies as a left hemichorea and hemiballismus in the context of non-ketotic hyperglycemia. Ruling out, in turn, infectious or pharmacological etiologies causing the syndrome.

Treatment with Haloperidol and Topiramate is started, as well as metabolic control of her recently diagnosed pathology (DM2), adjusting the basal-bolus scheme to the patient's needs. 13 days after the start of the pharmacological treatment, the patient was found with adequate evolution and resolution of the clinical symptoms in 50 percent with respect to the initial presentation, so she was discharged and it was decided to continue in outpatient management with clinical neurology and internal medicine. In outpatient imaging control, substantial improvement of hyperintensity in the T1 sequence is evidenced with respect to the

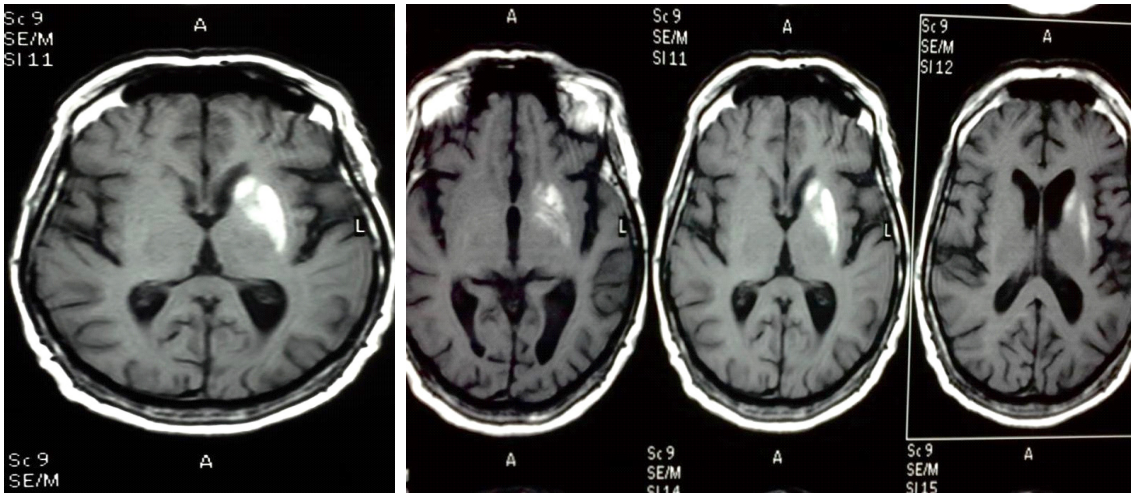

Figure 1 and 2: Axial T1-weighted nuclear magnetic resonance image in axial plane at the level of the basal nucleus where there is evidence of increased signal intensity in the head of the caudate nucleus and part of the posterior arm of the internal capsule.

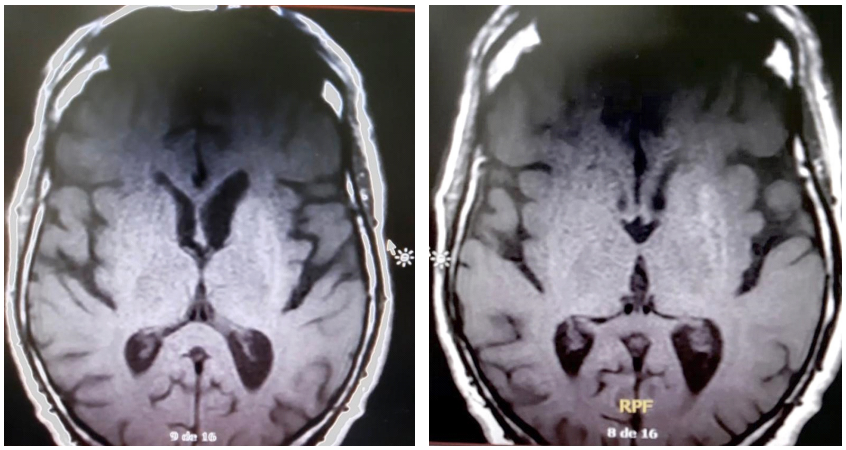

Figure 3: T1-weighted axial plane Nuclear Magnetic Resonance image showing a slight increase in signal intensity in the basal nucleus, especially in the head of the caudate nucleus and the lenticular nucleus, with less involvement in the internal capsule on the left side compared to the first images.

findings of the first study (Figure 3 and 4).

Discussion

Hemichorea hemiballism syndrome can result from multiple etiologies, the most frequent being ischemic stroke and hyperglycemia-type metabolic disorders. Hemichorea hemiballism due to non-ketotic hyperglycemia is a rare syndrome, more common in type 2 Diabetes Mellitus compared to type 1 Diabetes Mellitus, with predominance in the female sex and in advanced ages of life [18].

The presentation is generally the product of chronic accumulation and occurs without ketoacidosis, although it has been described in association with this metabolic emergency [19]. HH affects and interferes with the performance of daily activities in patients who suffer from it, generating deterioration in their quality of life, as well as in their physical and mental health.

Its diagnosis is a challenge for the clinician, because a variety of entities that may cause similar manifestations should be taken into account. A complete physical examination should be performed with emphasis on neurological evaluation, adequate description of the movement disorder, requesting complementary studies aimed at a timely diagnosis including confirmatory diagnostic images, individualizing the needs of each patient.

Diabetic striatopathy lesions have been attributed to gemistocyte accumulation and neuronal loss while biopsies have demonstrated hyaline degeneration, arteriolar narrowing and vascular proliferation. It has been hypothesized that GABA depletion in hemichorea-hemiballism syndrome is the cause of dyskinesias although this is less applicable to diabetic ketoacidosis because of GABA is produced from acetoacetate synthesised by the liver [20].

Once the diagnosis is made, the control of the underlying cause is essential and has shown good clinical outcomes, as in the case described above, being a patient with Diabetes Mellitus type 2, the metabolic control of glycemia was a main support, combined with dopamine receptor blocker and pharmacological treatment appropriate to the clinical scenario of the patient, giving satisfactory results in the short term.

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