Estriatopatía Diabética: Dos casos clínicos

Diabetic Striatopathy: Two clinical cases

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ABSTRACT

Movement disorders associated with diabetes mellitus (DM) are rare. The diagnosis of diabetic striatopathy (DS) is based on the presence of a triad characterized by hyperglycemia, hemiballismus/chorea and hypersignal of the basal ganglia in T1 on the MRI. The treatment is, in most cases, glycemic control.
The DS should be considered as differential diagnosis in extrapyramidal movements episodes, especially when associated with hyperglycemia in the elderly.
We present two cases of diabetic striatopathy.

Keyword: Striatopathy, diabetes mellitus, chorea, ballism, hyperglycemia.

RESUMEN

Los trastornos del movimiento asociados a la diabetes mellitus (DM) son raros. El diagnóstico de estriatopatía diabética (DS) se sustenta en la presencia de una tríada caracterizada por hiperglucemia, hemibalismo/corea e hiperseñal en T1 en los ganglios basales en la resonancia magnética (RM). El tratamiento es, en la mayoría de los casos, control glucémico.
La DS debe ser considerada dentro de los diagnósticos diferenciales de los cuadros de movimientos extrapiramidales, especialmente cuando estos se presentan con hiperglucemia y en pacientes ancianos.
Presentamos dos casos de estriatopatía diabética.

Palabras clave: estriatopatía, diabetes mellitus, corea, balismo, hiperglucemia.

INTRODUCTION

Diabetic striatopathy (DS) is a rare diabetes mellitus (DM) complication that affects 1 in 100,000 people[1]. Bedwell in 1960 described 53 cases, mainly elderly women.[2]

The term DS was introduced in the last decade to describe a hyperglycemic state associated with involuntary limb movements, usually chorea or ballism, and unique reversible abnormality of basal ganglia on computed tomography (CT) or magnetic resonance imaging (MRI). In the last years the use of term DS has been expanded to cases with hyperglycemia and either involuntary movements or an abnormal MRI.[3]

DS is usually described in type 2 diabetes mellitus (T2DM) patients with poor metabolic control, especially among older Asian women[3]. Main treatment is glycemic control and, in some cases, neuroleptic drugs.[4]

Both anatomic location and pathophysiologic mechanism have not been fully clarified[5]. It has been suggested that the DS has a microvascular etiology.

We present two cases of DS: one in a man without diagnosed diabetes mellitus and other one in a woman with long-standing history of diabetes.

CLINICAL CASE 1

A 79-year-old male with arterial hypertension, dyslipidemia, and benign prostatic hyperplasia was admitted to the emergency room with five days of polydipsia, polyuria, speech disturbance and left limbs involuntary movements. Movements were dystonic and choreoathetoid, the most severe in the upper limb. In the blood test he had hyperglycemia (890 mg/dl), negative ketonemia and HbA1c of 14.3%. An CT angiography ruled out ischemic or hemorrhagic stroke. An MRI showed a bright hyperintense signal at both lenticular nuclei without putaminal or caudal nuclei involvement. He started insulin with complete remission of the neurological findings once glycemic control was achieved.

CLINICAL CASE 2

An 84-year-old woman with long-standing history of T2DM, with suboptimal metabolic control, coronaropathy and cerebrovascular disease, was admitted to the emergency room due to one week of hyperglycemia, urinary incontinence, psychomotor agitation, and unconsciousness. On the physical examination at admission, she had conjugated eye deviation to the left side and choreatic movements of the left limbs.

The blood test showed severe hyperglycemia (799 mg/dL), elevated inflammatory markers, and leukocyturia. An MRI revealed hyposignal, bilaterally involving the *corpus striatum* and extended to the medial portion of the cerebral crus in T2/T2-flair, and a light hypersignal in T1 in the basal nuclei.

At first, the patient was treated with diazepam and insulin with partial clinical response, mantained during admission despite the metabolic improvement. Consequently, treatment with tetrabenazine was initiated, leading to full resolution of the extrapyramidal symptoms.

A year later, the patient was admitted once again for an episode of diabetic hiperosmolar syndrome. At that time, she also presented choreoathetotic movements of right and left upper limbs. Tetrabenazine treatment was restarted, however, the patient continued to have some myoclonus in the left limbs after discharge.

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Image 1. The first case axial section on T1 of the MRI.

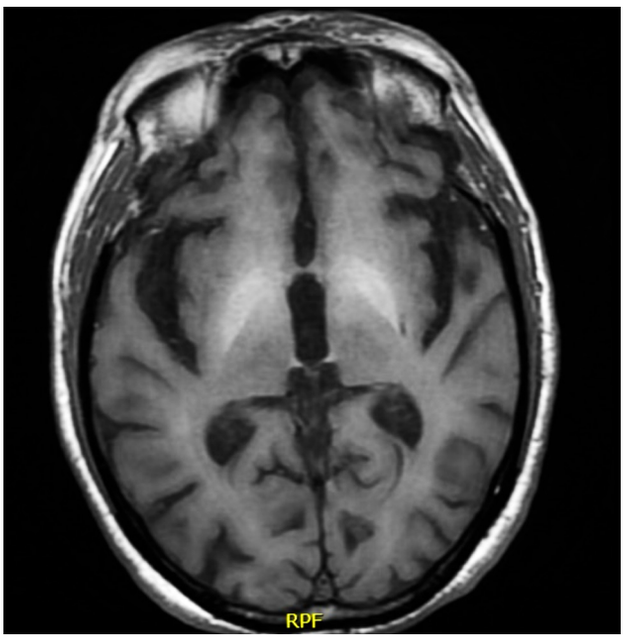
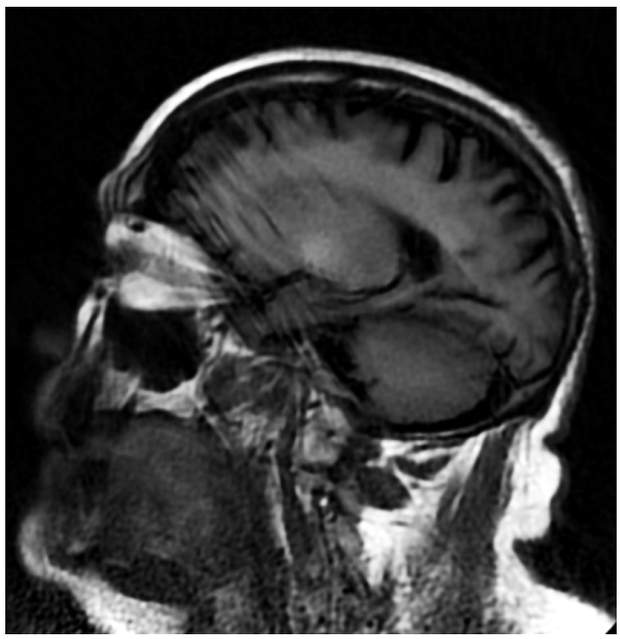


Image 2. The second case sagital section on T1 of the MRI.



DISCUSSION

The DS is more common in patients with long-standing diabetes, usually with poor metabolic control. The underlying vascular disease, and therefore, vascular insufficiency makes brain more susceptible, given basal ganglia irrigation and high metabolic demands[3]. Ischemia-induced dysfunction of GABA projection neurons is a proposed mechanism[6], however, there also seems to be a reduction in inhibition in the thalamus and the corpus striatum, resulting from decreased GABA levels due to elevated cerebral acetate and glucose concentrations. This is thought to occur due to disruption of the blood-brain barrier caused by plasma hyperviscosity[7], leading to disinhibition of the basal ganglia and the onset of involuntary movements[3]. This mechanism would explain the occurrence of DS in diabetic patients with mild diabetes and without any previously known vascular complications.

In both our cases, the MRI showed an hypersignal in T1 on the basal nuclei. In a study of 153 cases, this radiological finding was often found in putamen and/or caudate nucleus and in a lower proportion in globus pallidus[3]. To differentiate the hypersignal from hypertensive hemorrhage, there must be no mass effect and preservation of the internal capsule[8]. It has been reported one case of DS where a parenchymal transcranial sonography (PTcS) was performed, that revealed hyperechogenicity in the right lenticular nucleus with subsequent MRI showing hypersignal in the ipsilateral stratum in T1[9]. None of our cases underwent PTcS, but it could be a particularly useful diagnostic tool in environments where immediate MRI are not available.

CONCLUSION

DS is a rare complication of the DM; however, it should be considered as differential diagnosis in extrapyramidal movements episodes, especially when associated with hyperglycemia in the elderly.

CONFLICT OF INTEREST
The authors declare that they have no conflict of interests.

SOURCE OF FUNDING
This research had no funding sources.

ETHICAL ASPECTS
All participants submitted a consent form to be included in this study.

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