Short report

Biballism due to non-ketotic hyperglycaemia

A. Boughammoura-Bouatay , S. Chebel , S. Younes-Mhenni , M. Frih-Ayed *

Department of Neurology, University Hospital of Fattouma Bourguiba, avenue 1er-juin, 5000 Monastir, Tunisia

Received 19 March 2008; received in revised form 2 April 2008; accepted 6 April 2008
Available online 21 November 2008

Abstract

We describe the case of a 70-year-old woman, with type 1 diabetes mellitus, who suddenly developed a movement disorder on the left side of her body that rapidly extended to the right side, evoking biballism. There was no facial involvement and no vascular lesions on cerebral MRI but non-ketotic hyperglycaemia was present. A combination of a reduction in glucose levels and the use of neuroleptic drugs resulted in the disappearance of the abnormal movements. In this report, we discuss the association between non-ketotic hyperglycaemia and ballism along with a review of the literature.

© 2008 Published by Elsevier Masson SAS.

Résumé

Biballisme lors d’une hyperglycémie sévère sans cétose.

Nous rapportons l’observation d’une femme de 70 ans, diabétique de type 1, qui a présenté brutalement des mouvements anormaux de l’hémicorps droit et qui se sont étendus rapidement à l’hémicorps gauche sans participation faciale, évoquant un biballisme. Il n’y avait pas de lésions vasculaires à l’IRM cérébrale. Une hyperglycémie sévère sans cétose est probablement à l’origine de cette symptomatologie. La normalisation glycémique et l’utilisation des neuroleptiques ont permis une évolution favorable avec la disparition de ces mouvements. Dans cet article, est discutée l’association entre l’hyperglycémie sévère sans cétose et le biballisme avec revue de la littérature.

© 2008 Published by Elsevier Masson SAS.

Keywords: Non-ketotic hyperglycaemia; Hemiballism; Biballism; Diabetes mellitus

Mots clés : Hyperglycémie sans cétose ; Diabète sucré ; Hémiballisme ; Biballisme

1. Introduction

Hemiballism is a rare hyperkinetic movement disorder that is characterized by irregular, wide and vigorous involuntary movements of the limbs on one side of the body. The movements are usually continuous but may be intermittent and can be voluntarily restrained by the patient, albeit for only a few minutes at a time. These movements are more prominent during periods of rest but are absent during sleep. It may be seen with other types of involuntary movements such as chorea, dystonia or myoclonus. Hemiballism is reported to be most commonly due to a lesion in the subthalamic nucleus (STN) on the contralateral side of the involved limbs. There have been few reports of hemiballism in patients with non-ketotic hyperglycaemia, making biballism – and in particular, biballism secondary to non-ketotic hyperglycaemia – a rare condition [1–5]. Nevertheless, we describe here just such an unusual case of biballism, which completely disappeared after correction of severe hyperglycaemia.

2. Observation

A 70-year-old woman was brought to our hospital’s emergency department because of left-sided involuntary movements. Her medical history included type 1 diabetes diagnosed 2 months previously but incorrectly treated and hypertension treated with medications for 15 years. The uncontrollable, intermittent flinging movements of her left arm and leg started 2 weeks before coming to hospital. The movements were especially prominent during periods of rest but were absent during sleep. The left arm was more severely involved than the left leg. On admission, the patient had a blood pressure of 120/80 mmHg and a pulse rate
of 64 beats per minute. She was alert and aware of her surroundings. Apart from the involuntary movements, she presented with no other neurological abnormalities. Two days later, however, there was an extension of the involuntary movements to the right arm and leg.

Laboratory tests showed non-ketotic hyperglycaemia (35 mmol/L). Cerebral computed tomography (CT) showed a hyperdense lesion over both putamina (Fig. 1), whereas diffusion magnetic resonance imaging (MRI) revealed hyperintensities in both the caudate nuclei (Fig. 2A) and right globus pallidus (Fig. 2B). The movements were decreased with the administration of oral phenothiazine. She was discharged on the sixth day when her serum glucose level reached normal. The involuntary movements completely disappeared after 4 months.

3. Discussion

Biballism is rarely reported in the literature. It may be secondary to a double subthalamic vascular lesion, which is unusual, or to estroprogestative drugs, encephalitis of the basal ganglia or, less frequently to non-ketotic hyperglycaemia, as in the case of our patient. Bedwell in 1960 [1] reported a case of hemiballism in a patient with severe hyperglycaemia who was treated with blood glucose correcting drugs. Initially, hyperglycaemia, as a cause of hemiballism, was reported sporadically but now, it is considered the second most commonly reported cause after several recent reports from East Asia have provided a much more complete description of this disorder in more than 60 reported cases [3–5]. It tends to present in elderly people. The typical presentation begins with the development of a severe non-ketotic hyperglycaemia secondary to type 2 diabetes. As the hyperglycaemia develops, hemiballistic movements emerge but as soon as the blood glucose abnormality is corrected, the movements generally disappear within hours. However, 20% of patients present with persistent hemiballism for up to 3 months or longer.

In all of the reported cases, there was a high signal intensity on T1-weighted images in the putamen contralateral (and in some cases ipsilateral as well) to the involved side, with similar changes variably found in the globus pallidus and caudate nucleus [1,3,5–9]. Two-thirds of patients also have high signal abnormalities on T2-weighted sequences, and some have corresponding abnormalities on diffusion-weighted imaging (DWI), as seen in our patient [3,5,8]. MR spectroscopy has revealed a lower-than-normal ratio of N-acetylaspartate to creatine but an increased ratio of choline to creatine, suggestive of neuronal loss with hypercellularity [10].

On the basis of these findings, many different causes have been suggested, including petechial haemorrhages [2], decreased synthesis of GABA and acetylcholine secondary to metabolic changes [3,6,7] and temporary ischemia due to microinfarction (with or without hyperviscosity) [3,5,10]. Nev-
ertheless, the pathogenesis of this striking disorder remains uncertain.

Lesions outside of the STN, for example, at points along the afferent or efferent pathways connecting the STN to its projection areas – which include the globus pallidus, putamen, thalamus and brainstem, as found in our patient, are also reported to lead to hemiballism [11–13]. Surprisingly, a review of the more recent case-series has reported that lesions outside of the STN accounted for more than half of the reported hemiballism cases [12]. The striatum (caudate nucleus and putamen) is reported to be the most common non-STN site [14] and is usually associated with hyperglycaemia. Vigilance for this cause of biballism is important, as the movement disorder may be the first presentation of a potentially dangerous underlying hyperglycaemia.

References