LETTER TO THE EDITOR

A case report on pituitary macroadenoma presented as hemichorea hemiballism syndrome

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Sir,

A 36 year old woman was referred to the emergency room with a preliminary diagnosis of pituitary macroadenoma presenting as hemichorea-hemiballismus (HC-HB). She reported an acute onset of abnormal movement in the left extremities. An initial physical examination showed an acromegalic appearance. Neurological examination revealed severe HC-HB, especially prominent in the left upper extremity.

Laboratory and endocrine evaluation showed: elevated blood glucose, 471 mg/dL (normal range, 70–110); glycosuria; and elevated GH, 40.00 ng/mL (normal range, 0.06–5.00). Ketones and serum were not detected, and no electrolyte imbalance was observed. MRI revealed a 4x4x5 cm homogeneous intrasellar-suprasellar mass extending into the sphenoid and cavernous sinuses bilaterally. T1-weighted hyper intensity of the bilateral caudate

and right lentiform nuclei was observed (Fig. 1a, b). The patient was diagnosed non-ketotic hyperglycaemia with HC-HB. While the patient received insulin therapy treatment, the abnormal movements continued for three weeks. Such patients typically have elevated serum glucose levels, but are unaware of existing diabetes mellitus. Initially, our patient underwent conventional MRI with gadolinium enhancement. T1- and T2-weighted MR images showed high signal intensities in the right and left heads of the caudate nucleus and in the putamen. At the beginning of therapy, symptoms were treated with, but not controlled by, insulin alone. The addition of a dopamine receptor blocking agent quetiapine (25 mg twice a day) was effective over time. Diffusion-weighted (DW)-MRIs were obtained during the persistence of symptoms. DW-MRIs did not show high signal intensities reflecting ischaemia in the corresponding lesions. The movement disorders improved gradually followed by satisfactory regulation of blood glucose levels within two months. Transphenoidal surgery was planned to remove the

Sudden onset HC-HB syndrome caused by hypergly-caemia often presents in elderly patients who have a history of diabetes mellitus. MR images from these patients often reveal contralateral putaminal hyperintensity [1]. The main reason of non-ketotic hyperglycaemia induced HC-HB remains unknown. It is thought that reduction of regional blood flow in basal ganglia caused by hyperglycaemia leads the metabolism into an anaerobic pathway which damages GABA-inhibitory neurons in the striatum that disinhibits the external segment of the globus pallidus (GPe) and subthalamic nucleus resulting in hyperkinetic abnormal movements expressed as HC-HB. It has also been reported that metabolic acidosis causes reversible ischemia in basal ganglia without developing any infarction [2]. All these circumstances give rise to a hyperin-

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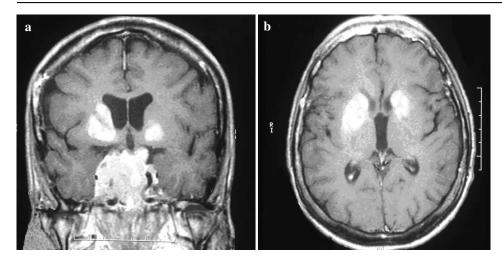


Fig. 1 Coronal T1-weighted magnetic resonance imaging (a) reveals a 4x4x5 cm enhancing homogeneous intrasellar-suprasellar mass infiltrating into the sphenoid and cavernous sinuses bilaterally. The axial image (b) displays increased intensity involving the bilateral caudate and right lentiform nuclei, partially sparing the left lentiform nucleus

tense image of cranial MR T1 sequence as it also reported in this presented case.

Considering all these former reports along with clinical and radiological findings we speculate that the diagnosis of our case is hyperglycaemia-induced HC-HB.

There are many previous reports of uncontrolled diabetes mellitus first presenting as HC-HB [1, 3–6]. This is the first report of a GH secreting, invasive pituitary macroadenoma causing HC-HB through long lasting hyperglycaemia

Extrinsic or intrinsic tumours may involve structures of basal ganglia, or their connections, by interruption of neuronal circuits as the possible mechanism for movement disorders. Our observations, however, support a mechanism that contrasts with the one postulated by Bhatoe [7]. We concluded that HB-HC may be an initial presentation of long lasting GH secreting pituitary adenoma by hormonal hyperfunction, but not by direct compression.

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