

A case of a diabetic woman with an uncontrollable arm

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Abstract

We present the case of an 83-year-old female patient with left-side hemichorea and hemiballism associated with poorly controlled type 2 diabetes mellitus. Hyperintensity in the right basal ganglia was evident in T1-weighted brain magnetic resonance. Clinical and radiological findings were consistent with diabetic striatopathy.

Keywords

Diabetic striatopathy, non-ketotic hyperglycemic hemichorea

Key Points

1. Diabetic striatopathy often manifests as hemichorea and mainly affects type 2 diabetes mellitus patients with poor metabolic control.
2. A high signal intensity basal ganglia lesion on the T1-weighted brain magnetic resonance imaging is the most consistent imaging finding.
3. In the acute phase, controlling hyperglycaemia is paramount; and anti-choreic drugs like haloperidol can be used as needed.

Case description

An 83-year-old female patient presented to the Emergency Department complaining of bizarre and uncontrollable movements of the left side arm. The patient had a poorly controlled type 2 diabetes mellitus (DM) due to poor insulin therapy compliance. Last glycated haemoglobin fraction value reached 13.7%. One month prior, she was admitted with hyperglycaemia. Her vital signs were within normal range. Left-side hemichorea with hemiballism and signs of distal symmetrical sensitive neuropathy were found on neurological examination. The blood analysis revealed: glucose of 39.6mmol/L and creatinine of 162µmol/L. Arterial blood gases and ketone bodies were within normal range. Her brain magnetic resonance exam is shown in Figure 1.

Quiz

What is the most likely diagnosis?

1. Thalamic haemorrhage
2. Basal ganglia calcification
3. Diabetic striatopathy
4. Tay-Sachs disease
5. Brain neoplasm

Answer: 3

Concerning the diagnosis, the following statements are true except one. Identify that exception:

1. Underlying pathophysiology is not clearly understood.
2. Surgical options are reserved for a few refractory cases.
3. Glycaemic control and thorough target-organ lesion screening, like proliferative retinopathy, are paramount.
4. Elder type 2 diabetes mellitus constitute the majority of the affected patients.
5. A normal brain computerized tomography is sufficient to exclude the diagnosis.

Answer: 5

Diabetic striatopathy (DS) is also known as 'non-ketotic hyperglycemic hemichorea' or 'chorea, hyperglycemia, basal ganglia syndrome'. As reported in literature, chorea and/or ballismus associated with DS onset rapidly and affect the arm, leg or face alone or in combinations. The ratio of left-sided and right-sided involvement is 1.2:1. Bilateral involvement was noted in 9.7% of patients. The mean age of the affected patients was 67.6 years with a male-to-female ratio of 1:1.7. Although, generally described in type 2 DM patients, a small proportion (3.4%) of type

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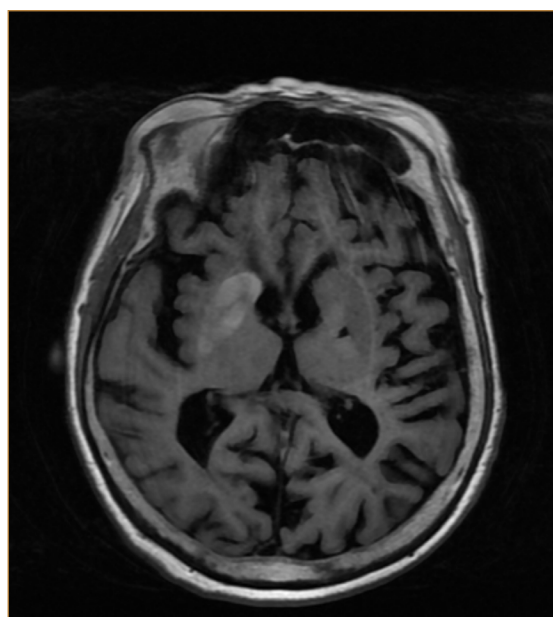


Figure 1. Axial fluid-attenuated inversion recovery T1-weighted brain magnetic resonance imaging showing a hyperintensity basal ganglia lesion.

1 DM is acknowledged. Most patients have poorly controlled DM. The average level of blood glucose among DS patients was estimated at 414 mg/dL and glycated haemoglobin at 13.1%.¹

Computerized tomography has poor sensitivity for brain changes due to DS. The most consistent imaging finding is a high signal intensity basal ganglia lesion on the T1-weighted brain magnetic resonance imaging. Signal intensity changes in T2-weighted differ among reports. Brain single photon emission computed tomography showed hypoperfusion at the basal ganglia or transitory hyperperfusion followed by hypoperfusion.²

DS primary pathologic mechanism is not fully clarified as few patients underwent biopsy. The pathologic changes found in DS patients

include: reactive astrocytosis in all studied patients; multiple infarcts in one patient; mild ischemia with gemistocytic accumulation in another; a third patient had patchy ischemic necrosis with oedema, foamy macrophage invasion into the parenchyma, red cell extravasation, and vascular proliferation, together with prominent arteriolar changes in which all layers of the wall were strikingly thickened. Some hyaline change of the vascular intima was noted in association with the narrowing of the lumen and obliteration. These changes are analogous to diabetic retinopathy.³

Acute treatment includes glycaemia level control and, as needed, anti-choreal treatment which includes haloperidol or other agents such as tiapride, chlorpromazine and benzodiazepines. Most patients show symptom remission over a period of days to several weeks upon glycaemic normalization. The overall recurrence rate is 18.2%. Intractable cases may be treated with pallidotomy, ventrolateral thalamotomy, transcranial magnetic stimulation, or *globus pallidus internus* deep brain stimulation.^{1,2}

Protection of the patient

The procedures were followed according to the regulations established by the institutional Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association.

Patient consent

Obtained.

Declaration of competing interests

Nothing to declare.

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