CASE REPORT

DOI: 10.5336/caserep.2019-72836

Non-ketotic Hyperglycaemic Hemichorea with Unilateral Magnetic Resonance Imaging Putaminal Hyperintensity

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ABSTRACT Non-ketotic hyperglycaemic hemichorea is a rare syndrome characterized by a triad of hemichorea, hyperglycemia, and peculiar imaging abnormalities. Regression of symptoms is typically associated with normalisation of serum glucose levels and plasma osmolality. An 85 year-old female was admitted to the neurology clinic with left-sided involuntary movements. Magnetic resonance imaging of brain demonstrated hyperintensity on T1 weighted images. She was treated with clonazepam (12. 5 mg/ day) for symptomatic therapy. Hyperglycaemia was controlled using a combined therapy with insulin aspart and insülin glargine. The patient showed prompt improvement in her involuntary movements in a day. Acute or subacute onset of unilateral involuntary movements should suggest non-ketotic hyperglycaemic hemichorea, the recognition of which may result in prompt control of the symptoms by normalization of blood glucose.

Keywords: Chorea; hyperglycaemia; magnetic resonance imaging; hemichorea

Hemichorea is a hyperkinetic movement disorder characterized by involuntary, rapid, random and irregular contractions of one side of the body.[1]

Non-ketotic hyperglycaemic hemichorea (NHH) is rare syndrome characterized by a triad of hemichorea, hyperglycaemia, and peculiar imaging abnormalities.[2] This syndrome has been reported more frequently in elderly patients, mostly Asian women, who have type 2 diabetes mellitus.[3] Regression of symptoms is typically associated with normalisation of serum glucose levels and plasma osmolality.

We herein report a case of non-ketotic hyperglycaemic hemichorea with unilateral putaminal hyperintensity.

CASE REPORT

An 85 year-old female was admitted to the neurology clinic with left-sided involuntary movements of upper and lower extremities progressing over the past two weeks. She had difficulty in her activities of daily living. She had type 2 diabetes mellitus in her medical history. There was no movement disorder in her family history.

Physical examination was normal. Neurological examination revealed chorea involving the left upper and lower extremities, was otherwise unremarkable. Random serum glucose level was 326 mg/dl with no ketones either in the blood or urine. High glycated haemoglobin (HbA1c) was 11.5%. The other laboratory tests were normal.

Magnetic resonance imaging (MRI) of brain demonstrated right sided putaminal hyperintensity on T1 weighted images (Figure 1). Diffusion-weighted images and the apparent diffusion coefficient map were normal.

18 F-Fluorodeoxyglucose positron-emission scan revealed right sided unilateral putaminal hypometabolism (Figure 2).

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Peer review under responsibility of Turkiye Klinikleri Journal of Case Reports.

Received: 16 Dec 2019 ***Received in revised form:*** 21 Mar 2020 ***Accepted:*** 23 Mar 2020 ***Available online:*** 15 Apr 2020

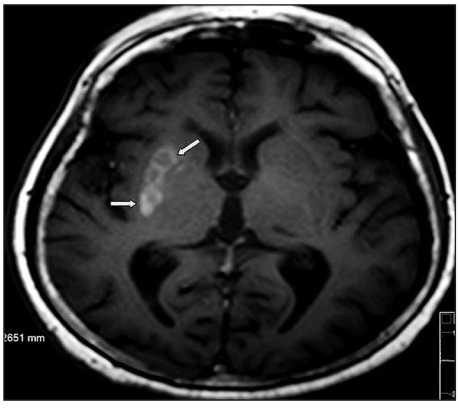

FIGURE 1: Right sided putaminal hyperintensity on T1 weighted images.

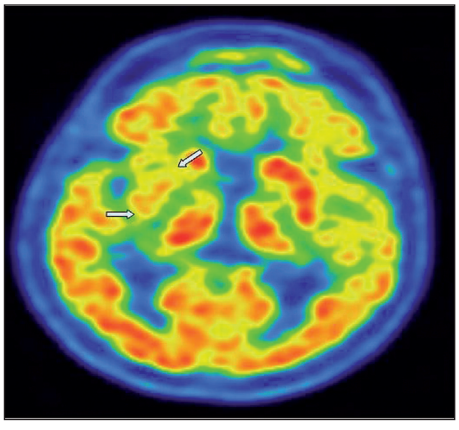

FIGURE 2: Right sided unilateral putaminal hypometabolism on 18 F-Fluorodeoxyglucose positron-emission scan.

Hyperglycaemia was controlled using a combined therapy with a fast-acting insulin analogue (insulin aspart) and long-acting insulin (glargine), subcutaneously. The patient showed prompt improvement in her involuntary movements in a day. Abnormal involuntary movements had totally disappeared 48 hours after the commencement of treatment. It was observed that the patient did not have any complaints at the 1-month follow-up.Control radiological imaging was not performed due to lack of complaints. Written consent was obtained from the patient for publishing case report.

DISCUSSION

In this case, the triad of NHH, acute hemichorea, and the contralateral bright putamen (hyperintense on T1 weight MRI) suggested a rare syndrome known as NHH.[4]

NHH occurs predominantly in elderly women mostly Asian.[2] The exact underlying psychophysiology is still unclear. Some hypothesized mechanisms include focal metabolism balance secondary to hyperglycaemia-induced hyperviscosity, increased sensitivity of dopaminergic receptors, decreased gamma-aminobutyricacid availability in the basal ganglia secondary to non-ketotic state, microvascular ischaemic injury and resulting hypoxia.[3]

Hyperintensity on T1 images in contralateral putamen is the typical brain MRI finding.[3,4]

NHH carries a good prognosis. Most of the literature report that rapid glucose-lowering therapy and adequate hydration resolve symptoms quickly in most patients.[5] In our case, the patient showed complete resolution after control of the hypergycaemia.

In conclusion, acute or subacute onset of unilateral involuntary movements should suggest NHH, the recognition of which may result in prompt control of the symptoms by normalization of blood glucose.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Cansu Eğilmez Sarıkaya, Pervin İşeri;* ***Design:*** *Cansu Eğilmez Sarıkaya, Pervin İşeri;* ***Control/Supervision:*** *Pervin İşeri;* ***Data Collection and/or Processing:*** *Cansu Eğilmez Sarıkaya, Pervin İşeri;* ***Analysis and/or Interpretation:*** *Cansu Eğilmez Sarıkaya, Pervin İşeri, Gür Akansel;* ***Literature Review:*** *Cansu Eğilmez Sarıkaya;* ***Writing the Article:*** *Cansu Eğilmez Sarıkaya;* ***Critical Review:*** *Pervin İşeri, Gür Akansel;* ***References and Fundings:*** *Cansu Eğilmez Sarıkaya;* ***Materials:*** *Cansu Eğilmez Sarıkaya, Pervin İşeri.*

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