NONKETOTIC HYPERGLYCEMIC HEMICHOREA

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DISCUSSION
• Typical patient with non-ketotic hemichorea shows triad:

  • *Nonketotic hyperglycemia,*

  • *Hemichorea and*

  • *High signal in basal ganglia in MRI T1 sequence or high density in CT scan.*
• Most frequently reported in elderly patients, typically Asian who have type II diabetes.

• The majority of cases published involved female patients.
Clinical presentation

• Chorea develops rapidly and can be either unilateral or bilateral and is seen during episodes of non-ketotic hyperglycaemia.

• Symptoms usually resolve upon normalisation of glucose levels.
• The exact underlying pathophysiology of changes seen on imaging of patients with non-ketotic hyperglycaemic hemichorea is not fully understood.
Some hypothesised mechanisms include:

- Hyperviscosity secondary to hyperglycaemia, leading to regional blood-brain barrier disruption and metabolic damage.
- The augmented sensitivity of dopaminergic receptors in a postmenopausal period (possibly explaining the female predominance).
- Decreased gamma-aminobutyric acid (GABA) availability in the striatum secondary to the non-ketotic state.
CT

- CT of the brain initially is normal, but later it can demonstrate subtle hyperdensity in the striatal region (caudate nuclei and putamen). Findings tend to be contralateral to the body side affected by hemiballistic, hemichoreic movements.
MRI

• MRI of the brain is the modality of choice for assessing possible non-ketotic hyperglycaemic hemichorea and typically demonstrates signal changes particularly in the putamen and/or caudate. If the changes are unilateral, then they are contralateral to the symptomatic side.

• **T1**: hyperintense

• **T2/FLAIR**: variable but generally hypointense
Overall, the hyperintensity on T1 sequence is the most consistent finding of the disease. Other associated findings do not present the same frequency and tend to vary.
• Imaging findings gradually resolve after hyperglycaemia correction. However, they tend to return to baseline more slowly than the clinical findings.
Differential diagnosis

• The main MRI differential is that of other causes of increased T1 signal in the basal ganglia:

• Idiopathic calcification

• Hepatic failure
  • Acquired non-Wilsonian hepatocerebral degeneration
    • Wilson’s disease (copper)
• Toxins / Ischaemia
  • Carbon monoxide
  • hyperalimentation or long term parenteral nutrition (manganese)
  • Global hypoxia
• Haemorrhagic stroke
• Japanese encephalitis
• Hamartoma in NF1
• Some causes of striatal hyperdensity on CT include:
  
  • Hypertensive hemorrhages
  
  • Basal ganglia calcifications
  
  • Tuberous sclerosis
Treatment and prognosis

• Symptoms and imaging findings usually resolve upon normalisation of glucose levels.

• Central Monoamine-Depleting agent - Tetrabenazine
THANK YOU