Haemichorea associated with transient hyperglycemia

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Introduction: *Chorea has a rare association with hyperglycemia . The triad of chorea, non-ketotic hyperglycemia and a high signal basal ganglia lesion on the T1 weighted brain MRI (C-H-BG), is a pattern described by Oh SH in a meta-analysis of 53 cases (2)*

Case presentation: *In ER a 75 years old woman, with glucose intolerance, hypertension, chronic renal failure, HCV-related cirrhosis, FAC in TAO, has shown for right-sided haemichorea involving upper and lower limbs. The chorea had been gradually progressive over 3 weeks. The movements have worsened with distraction and furthelrly slightly improved with intentional use of the affected limbs.*

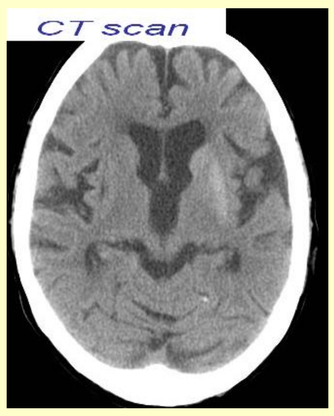
Parallel with the history of haemichorea she reported polydipsia, polyuria, bewilderment during the time that the chorea was developing.

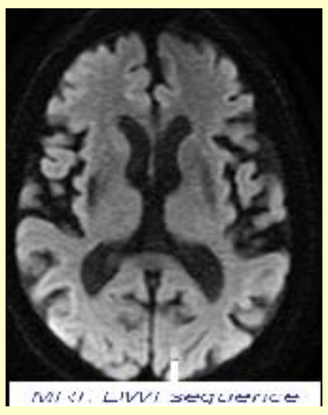
HbA1c was 11.9% and fasting glucose level 581 mg/dl, without evidence of ketoacidosis

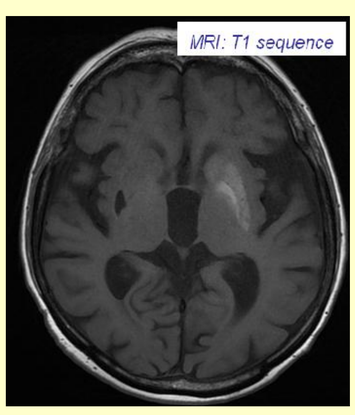
A CT scan of the brain revealed a hyperdense area in the left basal ganglia.

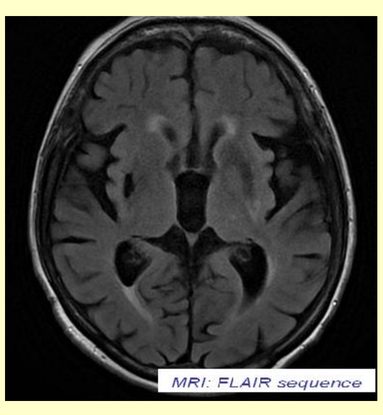
A MRI of brain showed T1 hyperintense signal in the left caudate and lenticular nuclei with sparing of the internal capsule. Vascular event were escluded, as the lenticular nucleus and internal capsule are in the same vascular distribution, yet the internal capsule was unaffected.

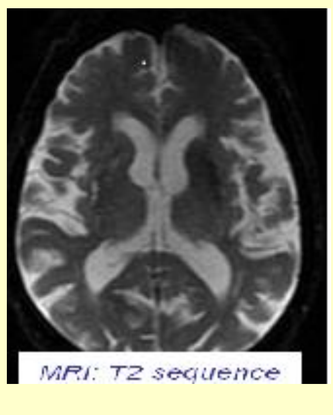
With the introduction of insulin, oral hypoglycemic medications, hydration, haloperidol (2 mg/ml, up to 8 + 8 + 5 drops daily) the patient improved gradually, by 1 month. Haloperidol was reduced to 5 drops twice daily. It was almost complete resolution of MRI and CT signs

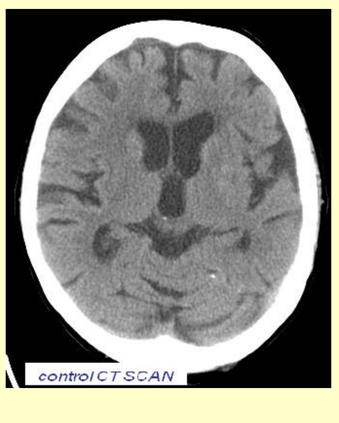












Discussion: ***The onset of chorea coincided with the exacerbation of hyperglycemia. There was a temporal relationship between restoration of euglycemia and improvement of the chorea. The imaging findings were typical for C-H-BG syndrome that characteristically demonstrates an unilateral increasing signal within the striatum on T1 weighted MRI. Theories include putaminal petechial haemorrhage, calcification and deficiency of basal ganglia γ-aminobutyric acid due to its use as an alternative energy source during hyperglycemia. A recent autopsy report of C-H-BG by Ohara showed that multiple infarcts were associated with a reactive astrocytic response (3). Thus it is proposed that hyperviscosity, induced by hyperglycemia, causes transient ischemia of vulnerable striatal neurons in predisposed individuals. This could cause astrocytic hypertrophy and oedema resulting in the characteristic MRI changes. This reversible mechanism would explain those cases, like ours, where transient MRI and clinical alterations occur***

Conclusion: ***C-H-BG appears to be a rare but potentially important cause of chorea although generally self-limiting, the chorea can persist. Further pathological studies of this syndrome would provide important insights into the pathogenesis of this disorder.***

References: *1. Permanent haemichorea associated with transient **hyperglycemia** Hannah Slabu, Sheila Savedia-Cayabyab, Peter Senior, Terra Arnason BMJ Case Rep. 2011*

2. Oh SH, Lee KY, Im JH, et al. Chorea associated with non-ketotic hyperglycemia and hyperintensity basal ganglia lesion on T1-weighted brain MRI study: a meta-analysis of 53 cases including four present cases. J Neurol Sci 2002;200:57–62

3. Ohara S, Nakagawa S, Tabata K, et al. Hemiballism with hyperglycemia and striatal T1-MRI hyperintensity: an autopsy report. Mov Disord 2001;16:521–5