Severe reversible chorea complicating non-ketotic hyperglycaemia

V Chikthimmah*, C Hopkins, P Wai, C Soh, M Silverdale

Case presentation
An 87-year-old female was started on prednisolone treatment for giant cell arteritis. Four weeks later she developed increasing confusion and slurring of speech. Her conscious level rapidly deteriorated, and she was admitted to hospital in a moribund state.

Investigations revealed steroid-induced hyperglycaemia with a random glucose of 39.7mmol/L. Serum osmolarity was 320mOsm/kg and urinary ketones were negative. U&Es on admission were Na 132mmol/L, K 4.7mmol/L, creatinine 156µmol/L, bicarbonate 23.6mmol/L. Arterial blood gas analysis revealed a pH of 7.43, pO2 11.1kPa, pCO2 4.4kPa, base excess of -1.7mmol/L. Other investigations including full blood count, liver function tests, calcium, magnesium, antinuclear antibodies and anticardiolipin antibodies were all normal or negative. Erythrocyte sedimentation rate done a week prior to admission was 2mm/hr when seen in a rheumatology clinic for follow up of her giant cell arteritis. The patient was managed with a sliding scale insulin regimen and IV normal saline in line with the protocol for the management of hyperosmolar non-ketotic state.

Her condition gradually improved over the next 48 hours, with normalisation of the blood glucose and electrolytes. However, it was noted that she had severe generalised chorea. Apart from the involuntary movements, clinical examination was unremarkable.

ABSTRACT
Movement disorders can manifest as initial presentations in a variety of metabolic and endocrine disorders. Chorea, though rare, has been documented to occur in patients with non-ketotic hyperglycaemia and usually has a good prognosis once the causal association is recognised and the hyperglycaemia treated. We report the case of an 87-year-old lady who presented with chorea complicating non-ketotic hyperglycaemia, and this was confirmed with MRI brain imaging demonstrating the classical feature of this condition. A high T1-weighted signal in the putamen is quite characteristic of this condition. Copyright © 2009 John Wiley & Sons.

KEY WORDS
reversible chorea; non-ketotic hyperglycaemia; T1-weighted MRI; putaminal petechial haemorrhage

Due to the clinical context of the movement disorder, it was felt that she had non-ketotic hyperglycaemic chorea, which was confirmed with MRI brain imaging demonstrating the classical features of this condition (Figure 1), consisting of high T1-weighted signal in the putamen.

She was treated with low dose tetrabenazine and her chorea gradually settled. When reviewed five months later, her chorea had settled almost completely and it was possible to slowly wean her off the tetrabenazine. Genetic testing for Huntington’s disease and other potential genetic causes of chorea was not performed in view of the acute presentation after non-ketotic hyperglycaemia, the relatively rapid recovery and classical MRI appearances.

Discussion
Chorea is a rare but important complication of non-ketotic hyperglycaemia.1,2 The condition usually recovers spontaneously over several weeks although, when severe, anti-choreic drugs such as tetrabenazine may be required to control the symptoms during the acute phase. Occasionally the chorea may be persistent, and it is sometimes associated with other neurological problems including cognitive impairment and personality change.1,4

A wide range of conditions have been known to be associated with chorea which need to be ruled out in the initial list of differential diagnosis. The following have all been associated with chorea: exposure to toxins and pharmacologic agents affecting dopaminergic neuronal pathways; encephalitis; neoplastic conditions; hereditary neurodegenerative disease such as late-onset Huntington’s disease; hereditary conditions, e.g. Wilson’s disease; endocrine conditions such as hyperthyroidism; Sydenham’s chorea following rheumatic fever (typically occurring in childhood); and autoimmune conditions such as systemic lupus erythematosus.
CASE REPORT

Severe reversible chorea complicating non-ketotic hyperglycaemia

The pathogenesis of chorea associated with non-ketotic hyperglycaemia is not fully understood. There is evidence that hyperglycaemia causes blood brain barrier dysfunction allowing erythrocytes to migrate across into the putamen which (in a poorly understood fashion) impairs the function of the putamen, leading to the development of chorea. Putaminal petechial haemorrhage has been documented pathologically in a single patient who died after chorea secondary to non-ketotic hyperglycaemia. Putaminal petechial haemorrhage would explain the MRI findings as red cells cause a high signal on T1-weighted MRI scans due to the iron molecule in haemoglobin. In a SPECT (single photon emission computerised tomographic) study done on non-ketotic hyperglycaemic chorea subjects, the researchers have detected reduced blood flow of the striatum on the contralateral site of the chorea, suggesting hypometabolism of the striatum as a possible mechanism in the development of chorea associated with non-ketotic hyperglycaemia.

Severe chorea is a rare complication of non-ketotic hyperglycaemia. It is important that general physicians involved in managing acute diabetic presentations are aware of this complication as well as the associated MRI appearances, so that appropriate treatment can be started when required and unnecessary investigations can be avoided.

Conflict of interest statement
There are no conflicts of interest.

References
References are available at www.practicaldiabetesinternational.com.

Key points
- Chorea is a rare but known complication of non-ketotic hyperglycaemia
- It usually recovers spontaneously over a few weeks after achieving euglycaemia
- Hyperintense signal in the putamen on a T1-weighted MRI is usually characteristic

Figure 1. T1-weighted coronal MRI brain scan done shortly after chorea developed. There is generalised cerebral atrophy. High signal change is seen in the putamen (arrow)

The pathogenesis of chorea associated with non-ketotic hyperglycaemia is not fully understood. There is evidence that hyperglycaemia causes blood brain barrier dysfunction allowing erythrocytes to migrate across into the putamen which (in a poorly understood fashion) impairs the function of the putamen, leading to the development of chorea. Putaminal petechial haemorrhage has been documented pathologically in a single patient who died after chorea secondary to non-ketotic hyperglycaemia. Putaminal petechial haemorrhage would explain the MRI findings as red cells cause a high signal on T1-weighted MRI scans due to the iron molecule in haemoglobin. In a SPECT (single photon emission computerised tomographic) study done on non-ketotic hyperglycaemic chorea subjects, the researchers have detected reduced blood flow of the striatum on the contralateral site of the chorea, suggesting hypometabolism of the striatum as a possible mechanism in the development of chorea associated with non-ketotic hyperglycaemia.

Severe chorea is a rare complication of non-ketotic hyperglycaemia. It is important that general physicians involved in managing acute diabetic presentations are aware of this complication as well as the associated MRI appearances, so that appropriate treatment can be started when required and unnecessary investigations can be avoided.

Conflict of interest statement
There are no conflicts of interest.

References
References are available at www.practicaldiabetesinternational.com.

Key points
- Chorea is a rare but known complication of non-ketotic hyperglycaemia
- It usually recovers spontaneously over a few weeks after achieving euglycaemia
- Hyperintense signal in the putamen on a T1-weighted MRI is usually characteristic

Figure 1. T1-weighted coronal MRI brain scan done shortly after chorea developed. There is generalised cerebral atrophy. High signal change is seen in the putamen (arrow)

The pathogenesis of chorea associated with non-ketotic hyperglycaemia is not fully understood. There is evidence that hyperglycaemia causes blood brain barrier dysfunction allowing erythrocytes to migrate across into the putamen which (in a poorly understood fashion) impairs the function of the putamen, leading to the development of chorea. Putaminal petechial haemorrhage has been documented pathologically in a single patient who died after chorea secondary to non-ketotic hyperglycaemia. Putaminal petechial haemorrhage would explain the MRI findings as red cells cause a high signal on T1-weighted MRI scans due to the iron molecule in haemoglobin. In a SPECT (single photon emission computerised tomographic) study done on non-ketotic hyperglycaemic chorea subjects, the researchers have detected reduced blood flow of the striatum on the contralateral site of the chorea, suggesting hypometabolism of the striatum as a possible mechanism in the development of chorea associated with non-ketotic hyperglycaemia.

Severe chorea is a rare complication of non-ketotic hyperglycaemia. It is important that general physicians involved in managing acute diabetic presentations are aware of this complication as well as the associated MRI appearances, so that appropriate treatment can be started when required and unnecessary investigations can be avoided.

Conflict of interest statement
There are no conflicts of interest.

References
References are available at www.practicaldiabetesinternational.com.

Key points
- Chorea is a rare but known complication of non-ketotic hyperglycaemia
- It usually recovers spontaneously over a few weeks after achieving euglycaemia
- Hyperintense signal in the putamen on a T1-weighted MRI is usually characteristic

Figure 1. T1-weighted coronal MRI brain scan done shortly after chorea developed. There is generalised cerebral atrophy. High signal change is seen in the putamen (arrow)

The pathogenesis of chorea associated with non-ketotic hyperglycaemia is not fully understood. There is evidence that hyperglycaemia causes blood brain barrier dysfunction allowing erythrocytes to migrate across into the putamen which (in a poorly understood fashion) impairs the function of the putamen, leading to the development of chorea. Putaminal petechial haemorrhage has been documented pathologically in a single patient who died after chorea secondary to non-ketotic hyperglycaemia. Putaminal petechial haemorrhage would explain the MRI findings as red cells cause a high signal on T1-weighted MRI scans due to the iron molecule in haemoglobin. In a SPECT (single photon emission computerised tomographic) study done on non-ketotic hyperglycaemic chorea subjects, the researchers have detected reduced blood flow of the striatum on the contralateral site of the chorea, suggesting hypometabolism of the striatum as a possible mechanism in the development of chorea associated with non-ketotic hyperglycaemia.

Severe chorea is a rare complication of non-ketotic hyperglycaemia. It is important that general physicians involved in managing acute diabetic presentations are aware of this complication as well as the associated MRI appearances, so that appropriate treatment can be started when required and unnecessary investigations can be avoided.

Conflict of interest statement
There are no conflicts of interest.

References
References are available at www.practicaldiabetesinternational.com.
Severe reversible chorea complicating non-ketotic hyperglycaemia

References