A rare case of DKA in a patient with maturity diabetes of young.

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INTRODUCTION:

Maturity Onset Diabetes of the Young (MODY) accounts for upto 2% of all patients with diabetes. Hepatocyte Nuclear Factor 1 alpha (HNF1-A) MODY is the most common subtype accounting for 30-70% of all MODY cases. Typically, it presents in young adults below the age of 45, frequently < 25 with autosomal dominant family history of diabetes, absence of autoimmune markers and insulin resistance and c-peptide positivity.

DKA is a rare complication of MODY particularly in situations of non-compliance. We describe a case of DKA in a genetically confirmed HNF1-A MODY patient presented to our hospital.

CASE REPORT:

A 26-year-old female was admitted with severe vomiting. She had a background history of HNF1- alpha MODY diagnosed at the age of 15 when she was found to have hyperglycaemia during pregnancy. She was on Gliclazide 40mg daily but stopped taking it about a year ago. On admission her pH was 6.96, blood glucose of 31.4 mmol/L and blood Ketones of 5.8 mmol/L. This was consistent with DKA which was successfully treated. There was no evidence of sepsis. Her HbA1c was high at 101mmol/mol suggesting poor glycaemic control. She had uneventful recovery and was discharged home on Gliclazide with appropriate follow up arranged.

DISCUSSION:

The presence of DKA was previously considered an exclusion criterion for MODY according to the International Society for Paediatrics and Adolescent Diabetes (IPSAD) 2009 guidelines. It is presumed that patients with MODY do not develop DKA due to the presence of residual insulin production that prevents ketogenesis. However, this was withdrawn in the 2014 update. The majority of patients with genetically proven MODY are initially incorrectly diagnosed as Type 1 or Type 2 diabetes. Exclusion of DKA from the diagnostic criteria will lead to further misdiagnosis which will have implications for the patient and family members. MODY should be included in the differential diagnosis of patients presenting with DKA particularly if there are other features to suspect. This should of course be balanced with the limitation of resources for carrying out genetic testing.

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