Long-term honeymoon period in Type 1 diabetes: True diagnosis MODY5; New mutation of HNF1B

Zekiye Kupcu, Aysun Bideci, Esra Doger, Orhun Camurdan, Peyami Cinaz
Gazi University Faculty of Medicine, Department of Pediatric Endocrinology

Introduction

MODY is an autosomal dominant inherited type of diabetes that has been diagnosed before the age of 25 and caused by pancreatic β-cell dysfunction. HNF1B-MODY is more rare than other MODY causes and its frequency is between 1-5%. Mutations in HNF1B (MODY 5) are associated with pancreatic agenesis, kidney anomalies, genital system malformations and liver dysfunction.

Case

The patient is 8 years 2 months old girl whom fasting blood glucose level was 370 mg / dl, HbA1c: 13.9%, c-peptide: 0.29. Anti-GAD (+), anti-insulin(-), islet cell antibody (+). In the follow-up period 1 month after the diagnosis of honeymoon period, the patient entered the honeymoon period, which is claimed to be good for the use of a plant mixture of diabetes was learned. During the follow-up, insulin requirement was 0.16 / kg / day at 3 months and insulin was completely discontinued at follow-up. One year after the diagnosis, the baseline rate was found to be 78 mg / dl, c peptide was 0.83. Genetic analysis with suspicion of MODY was found to be heterozygous mutation c.C146G (rs770078634), a new mutation in the HNF 1B gene. In the abdominal ultrasonography of the patient, the pancreas was normal and the kidney had a double collector channel. Liver enzymes were normal.

Result

MODY is mostly seen in adolescence and young adulthood. However, it can be seen at earlier ages. Antibody positivity may initially lead to patients diagnosed with type 1 DM in rare cases. Although our patient was initially diagnosed with Type 1 DM, the need for insulin for a long time in the follow-up period brought MODY to mind. The patient family thought that the patient's diabetes had improved due to herbal remedy until the result of genetics. In patients with type 1 DM, the honeymoon period should be stimulating for MODY.