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Underdiagnosed Complications of Type 1 Diabetes Mellitus: Mauriac Syndrome

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ABSTRACT

Mauriac syndrome is a rare complication of poorly managed insulin dependent diabetes mellitus.
It is characterized by dwarfism, obesity, delayed puberty, cushingoid features, hepatomegaly and elevated transaminases. Although rare in developed countries, cases are seen in developing states where patients are not well aware about self management of type I diabetes mellitus. We present a 19 year old female of type 1 diabetes mellitus with chronic poor glycemic control with features of Mauriac syndrome from northern Himalayan region. Strict management of glycemic control can reverse the condition and may lead to better outcome.

Keywords: Mauriac syndrome, type I diabetes mellitus, insulin dependent diabetes mellitus

INTRODUCTION

Mauriac syndrome has been reported as rare complications of type I diabetes mellitus. It is characterized by hepatic glycogenosis, caused by chronic underinsulinzation leading to growth failure, delayed puberty, moon facies, hepatomegaly, protuberant abdomen, proximal muscle weakness, dyscholestro-lemia.(1,2) It occurs among patients of type 1 diabetes mellitus with poor glycemic control. It is more common in adolescent population but young children are also affected. Exact cause is not elucidated but multiple factors including decreased IGF-1 and growth hormone resistance play role. We present a case of type 1 diabetes mellitus with chronic poor glycemic control with features in spectrum of Mauriac syndrome.

CASE DESCRIPTION

We describe a 19-year-old female with Insulin dependent diabetes mellitus diagnosed 10 years ago, on biphasic insulin 50/50 (50% soluble Insulin and 50% Isophane Insulin). She presented with history of fever and vomiting with high blood sugar. She had pallor, growth retardation (height for age <3rd centile, weight for age 50th centile) and delayed puberty. Respiratory, CVS and CNS examination were normal. At admission, RBS was 465mg/dl and urine positive for ketone. The patient had anemia (Hb-6.3) with the microcytic hypochromic picture. HbA1c was high (12.6%), Triglyceride 265, Cholesterol 200. HBA1C was 12.6%, aspartate aminotransferase-115, alanine aminotransferase-96, with normal serum bilirubin. Serum urea and creatinine were 38 and 1.8 respectively. Thyroid functions, celiac screen and cortisol were normal. Ultrasound confirmed massive liver with normal spleen and unilateral hydronephrosis. Urine examination revealed acute Urinary tract infection and culture grew E. coli (sensitive to Piperacillin). Past history of long standing insulin dependent diabetes mellitus with history of multiple times hospital admissions with diabetic ketoacidosis in past 2 years. Father is primary care taker and illiterate. The patient herself is able to inject insulin, but there is

lack of compliance to insulin regimen advised. Patient managed with IV fluids, insulin infusion and IV antibiotics. As the patient had acute kidney injury, hence she was shifted to the higher centre for further management. As patient presented with clinical spectrum of Mauriac syndrome which is less commonly seen in current scenario as better insulin regimens are available but still it is prevalent in rural region where better health facilities and awareness are lacking.

DISCUSSION

Mauriac syndrome is initially described in early 20th century as clinical manifestation of poorly controlled type I diabetes mellitus including hepatomegaly, protuberant abdomen, dwarfism, moon-shaped face, and fat deposition in body.(3) We described a patient with the similar clinical picture from the rural region of northern Himalayas. Although this condition is common but underdiagnosed pertaining to lack of awareness among people. Index case had growth deceleration, cushingoid features, a protuberant abdomen, and hepatomegaly associated with long-term poor glycemic control. Exact causation for Mauriac syndrome is not well understood, but insufficient uptake of glucose due to insulin deficiency and decreased circulating IGF-I play some role. Glycogen deposition in hepatocytes cause hepatomegaly.(4) In poorly controlled diabetes mellitus type I, glucose can freely enter hepatocytes through facilitated diffusion and can be stored as glycogen.(2,4)

Few studies suggested that overinsulization may lead to deranged liver enzymes and liver enlargement and irregular dosing of insulin causing vicious cycle of hyperglycemia and hypoglycemia leading to glycogen deposition and hepatomegaly is associated with the syndrome.(1,2,4–7) Decrease in liver span and normalization of deranged live enzymes is reported with achieving good glycemic control. It may take few months or even years to achieve normalization of liver.

However, index case was followed for two months, but there was persistence of hepatomegaly and other features, although the liver span was decreased. Failure of decrease in liver size was attributed to failure to achieve tight glycemic control as parents and patient were not well understood the self-management and there was difficulty in procuring insulin secondary to financial issues.

CONCLUSION

Mauriac syndrome is rare complication of insulin dependent diabetes mellitus. It is commonly present in adolescence but few reported cases showed its presence in younger children too. Strict management of blood sugar leads to regression of clinical signs and hepatomegaly and patient growth can be resumed in normal manner. Long term poor management of blood sugar is prime cause.

Conflict of interest: None

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