Brief Communication

Re-emergence of a rare syndrome: A case of mauriac syndrome

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ABSTRACT

Mauriac syndrome is a rare syndrome associated with type 1 diabetes (T1DM) in children presenting with growth retardation, hepatomegaly, and cushingoid features. Recently, there has been re-emergence of this syndrome, especially with the use of premix insulin. A 15-year old type 1 diabetic boy, who was on premix insulin with erratic blood glucose, was referred to us for evaluation of short stature. He had significant short stature, hepatomegaly, and cushingoid features. His growth hormone (GH) stimulation was normal, and so was the overnight dexamethasone suppression test, based on which the diagnosis of Mauriac syndrome was reported. He was made to switch over to basal bolus regime, and was advised to follow-up for 6 months. He had reduction in hepatomegaly and a height gain of 3 cms.

Key words: Growth retardation, hepatomegaly, mauriac syndrome, type 1 diabetes

Introduction

Mauriac syndrome is a rare syndrome associated with type 1 diabetes (T1DM) in children. The clinical features consists of growth retardation, hepatomegaly, and cushingoid features. The incidence of this syndrome had decreased significantly with introduction of long-acting insulin and better control of blood sugar.[1] Recently, there has been re-emergence of this syndrome, especially with the use of premix insulin.[2] Herein, we report such a case.

Case Report

A 15-year-old Type 1 diabetic boy was referred to us for evaluation of short stature. He was diagnosed to have T1DM, following an episode of diabetic ketoacidosis (DKA) 8 years back, and was started on premix (30/70) insulin. He was irregular with the treatment, with 4 hospitalisations for DKA till date.

Examination showed that he was significantly short for his age, height 131 cms (less than 3rd percentile), weight 28 kg (less than 3rd percentile) and body mass index of 16.31. Secondary sexual characteristic A1, P1, testicular volume less than 4 ml bilaterally. He had abdominal fat deposition [Figures 1 and 2] and liver was palpable clinically 5 cms below costal margin [Figure 3] with no splenomegaly or free-fluid. There were no signs of chronic liver failure.

Investigations showed haemoglobin of 13.5 gm/dl. Liver function tests, renal function tests, and urine routine examination were normal. Ultrasound abdomen showed liver enlargement 15.5 cms with increased echotexture, with normal spleen and portal vein and no free-fluid. On evaluation for diabetic status, he had an HbA1c of 10.3%, urine microalbumin 70 μg/gm of creatinine, and fundus showing background diabetic retinopathy. He had a bone age of 10.2 years, (Tanner Whitehouse 2) thyroid-stimulating hormone (TSH) of 2.4 mIU/ml and T4 of 8.8 microgm/dl. His growth hormone (GH) stimulation test and overnight dexa methasone suppression test were normal (after blood glucose control and testosterone priming).

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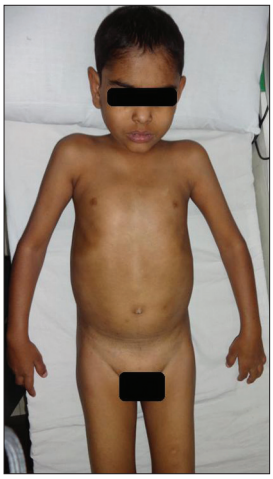

Figure 1: Short stature, abdominal obesity

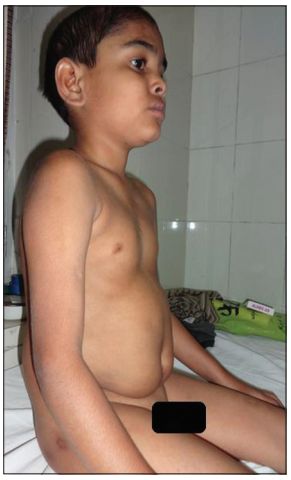

Figure 2: Short stature, abdominal obesity

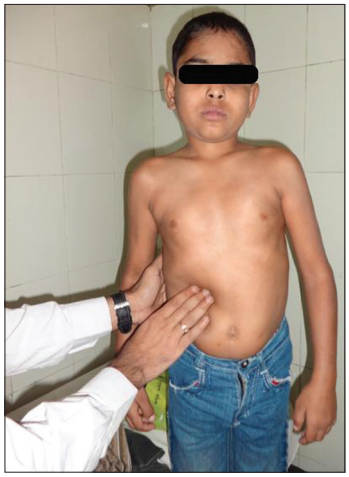

Figure 3: Hepatomegaly

Based on the clinical history and investigations, the final diagnosis of Mauriac syndrome was made and the patient was advised tight control of sugars. He was switched over to basal bolus regime, with Glargine in the night and three doses of short-acting insulin before meals. He was follow-up for 6 months. He had shown reduction in hepatomegaly and a height gain of 3 cms. After that he was lost to follow-up.

Discussion

Mauriac in 1930, described growth failure and maturational delay with hepatomegaly and abdominal distension in children with T1DM, who were treated with short-acting insulin.[3] Hepatomegaly was commonly observed in children in the earlier periods of diabetic treatment, when only short-acting insulin was available and aglycosuria was the objective of treatment. It was noticed that the hepatomegaly regressed when the children were given the newly introduced protamine zinc insulin, providing better sugar control. In the late 1930s Joslin clinic reported a case series of 60 youngsters with hepatomegaly, growth failure, delayed sexual maturation, and severe uncontrollable diabetes.[4] Equal incidence is reported in males and females, with most of the cases occurring during adolescence.[5] With better control of sugar, the incidence of this syndrome has reduced rapidly and in the current era this is a very rare syndrome.

Two different forms of Mauriac syndrome have been described, based on the presence or absence of obesity. In first form, as classically described, treatment with regular insulin alone there is associated Cushingoid obesity and documented-wide fluctuation between hyperglycemia and hypoglycemia, suggestive of a pattern of over-and under-insulinization, with secondary hyperadrenalism. Periods of over-insulinization appear to be essential for the development of obesity, and for the induction of hyperadrenalism. Recently, Mauriac syndrome has been reported in patients who are not obese and are without a history of alternating hypoglycemia and ketoacidosis. This occurs in patients who have been given regular, under the dose insulin.[1]

The pathogenesis of growth retardation is not clear but is thought to be multifactorial. Inadequate glucose to the tissues, decreased Insulin-like growth factor-1 and GH levels, hypercortisolism, resistant or defective hormone receptor action contribute to stunted growth and delay in puberty.[5] The periods of supraphysiological levels of insulin is associated with hepatomegaly. The cause of hepatomegaly is thought to be due to the deposition of glycogen in the liver,[6] and similar subcutaneous deposition gives rise to the round moon like facies.[7]

Growth failure, delayed puberty and hepatomegaly in Mauriac's syndrome improves with glycemic control.[1] Aggressive glycemic control has been associated with worsening of retinopathy, which should be monitored.[8]

Conclusion

Mauriac syndrome is a rare manifestation of poorly treated T1DM. With aggressive glycemic control, the manifestations of this syndrome can be reverted.

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