



Figure 1: Round face

A rare case of Mauriac syndrome

Sir,

Mauriac syndrome is an uncommon syndrome described in Type 1 diabetic children treated with short-acting insulin. The clinical features consist of growth retardation, hepatomegaly, and cushingoid features. The incidence of this syndrome has decreased significantly with introduction of long-acting insulin and better control of sugars.^[1] Recently, there have been cases of this syndrome reported in improperly treated T1DM children, especially with premix insulin. We report one such case.

A 12-year-old boy was referred to us for evaluation of short stature. He was diagnosed to have Type 1 DM 7 years back, following an event of diabetic ketoacidosis, and was put on premix insulin (30/70). He had history of weight gain after starting insulin and had no further episodes of ketoacidosis. He was irregular in taking insulin and monitored sugars once in 1–2 months only.

Examination showed that he was significantly short for his age (height 120 cm, less than 3rd percentile), weight was 20 kg (less than 3rd percentile), and body mass index (BMI) was 13.88 (5th–10th percentile). Also, he had cushingoid features with round face [Figure 1] and abdominal fat deposition [Figure 2]. Liver was palpable clinically 7 cm below costal margin, with no splenomegaly or free fluid. He was prepubertal with secondary sexual characteristic A1, P1, and testicular volume of less than 4 ml bilaterally.

Investigations showed normal hemogram, normal liver and renal function tests, and urine routine examination was



Figure 2: Round face, prominent abdomen

normal. Ultrasound abdomen showed liver enlargement 13.5 cm, increased echotexture, with normal spleen and portal vein and no free fluid. On evaluation for diabetic status, he had an HbA1c of 8.3%, urine microalbumin 50 µg/g of creatinine, and fundus showing background diabetic retinopathy. He had a bone age of 8.5 years (Tanner Whitehouse 2), thyroid stimulating hormone (TSH) of 3.2 mIU/ml, and T4 of 9.8 µg/dl. Growth hormone (GH) following clonidine stimulation was 11.2 µg/l and serum cortisol following overnight Dexamethasone suppression was 0.98 µg/dl.

Based on the clinical history and investigation findings, the final diagnosis of Mauriac syndrome was made and the patient was switched over to basal bolus regime. He was followed up for 1 year, and has significant reduction of hepatomegaly and has gained 11 cm of height. He is P2 now and has a testicular volume of 5–6 ml.

Mauriac, in 1930, described growth failure and maturational delay with hepatomegaly and abdominal distension in children with Type 1 diabetes, who were treated with short-acting insulin.^[2] Equal incidence is reported in males and females, with most of the cases occurring during adolescence.^[3] With better control of sugars, the incidence of this syndrome has reduced rapidly and in the current era. Two different forms of Mauriac syndrome have been described based on the presence or absence of obesity.^[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 (IGF-1) and GH levels, hypercortisolism, and resistant or defective hormone receptor action contribute to stunted growth and delay in puberty.^[3] The period of supraphysiological levels of insulin is associated with hepatomegaly. The hepatomegaly is thought to be due to the deposition of glycogen in the liver,^[4] and similar subcutaneous deposition gives rise to the round, moon-like facies.^[5] Growth failure, delayed puberty, and hepatomegaly in Mauriac syndrome improve with glycemic control.^[1]

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