A Case Report of Mauriac Syndrome: Diabetic Dwarfs

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ABSTRACT

Introduction: Mauriac syndrome is a severe form of growth retardation seen in patients with poorly controlled type 1 diabetes mellitus characterized by growth failure, delayed puberty, hepatomegaly and Cushingoid features. It is often referred to as diabetic dwarfism. The actual cause is unknown but is probably a combination of factors including inadequate glucose in the tissues, decreased IGF-1 and growth hormone levels, impaired bioactivity of the hormones, a circulating hormone inhibitor, resistant or defective hormone receptors, especially with the use of premix insulin. Case Report: We report a case of 15 year old female, known case of type 1 diabetes mellitus since the age of 5 years, presented to us with diabetic ketoacidosis. She had multiple similar episodes in the past. She was on premix insulin (30/70) and tablet metformin but had poor compliance to the treatment. On examination she was found to be significantly short for her age (height 129 cm, less than 3rd percentile) and weight was 27 kg (less than 3rd percentile) with cushingoid features (round face and protuberant abdomen). She was at Tanner stage P1. On detailed laboratory evaluation, IGF-1 levels were decreased (64), LH levels were below normal limits (0.2) and FSH levels were normal, while transaminases and HbA1C (8.3) were elevated. TSH was elevated (22.6), anti-TPO was elevated (252.6) with normal FT3, FT4. Hemogram, renal function test, GH, cortisol, estradiol, prolactin, S. iron, S. ferritin were normal. Ultrasound abdomen showed fatty liver with atrophic pancreas and USG neck showed bilateral bulky thyroid lobes. Fundus examination was normal. Now for the past 1 year she is on regular insulin 8/15/8 units (BBF/BL/BD) & Insulin Glargine 12 units. This led to a decrease in hepatomegaly and increase in height of 2 cms. Conclusion: Mauriac syndrome is a rare complication of poorly controlled diabetes mellitus in adolescence, but the treating physician should keep a high index of suspicion for this so that proper growth can be accomplished with timely intervention.

Key words: Mauriac syndrome

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