

P86. An extremely rare complication of type 1 diabetes mellitus: Pierre Mauriac syndrome.

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Context:

Pierre Mauriac syndrome is characterized by delayed in development and hepatomegaly in patients with insulin-dependent diabetes mellitus. It is associated with poor control of type 1 diabetes mellitus (T1DM) in adolescents. It is associated with growth failure and delayed pubertal maturation.

Objective:

To present a case report of Mauriac syndrome as an extremely rare complication of type 1 diabetes.

Methods:

A direct questioning was carried out as well as a review of the patient's clinical record, which is in regular control in the endocrinological gynecology outpatient clinic of the National Medical Center "20 de Noviembre".

Patient:

A 16-year-old woman with a 15-year history of T1DM was admitted for evaluation of growth retardation, primary amenorrhea and uncontrolled diabetes. She debuted before reaching the first year of life with diabetic ketoacidosis.

She does not follow the diet or take his treatment adequately, bad adherence to insulin therapy.

The physical examination revealed hypertension (BP 140/90 mmHg in repeated recordings), and hepatomegaly. The patient was prepubertal with stage 1 Tanner and primary amenorrhea.

Anthropometric data revealed height 137 cm (<3 rd percentile), weight 33 kg (<3 rd percentile), body mass index 17.58 kg/m², height age 10.5 years, weight age 11 years.

Laboratory analysis revealed: hemoglobin 13.4 g/dl, total protein 8.1 g/dl, albumin 4 g/dl, triglycerides 354 mg/dl, total cholesterol 261 mg/dl, aspartate aminotransferase 45 U/L, alanine aminotransferase 38 U/L. Serum cortisol was 499 nmol/L and glycosylated hemoglobin 11.1%. Ultrasound revealed a grade II fatty change in the liver.

Interventions:

Adjusted therapy with long-acting insulin (12-0-12 IU) and rapid insulin according to daily requirements.

Main Outcome Measures:

The patient has improved her tolerance to the diet with carbohydrate counting, uses long-acting insulin that every 24 hours and according to her daily needs uses fast-acting insulin.

Results: According to clinical characteristics and compelling studies, Mauriac syndrome is diagnosed.

Conclusions:

Mauriac syndrome is a rare complication of poorly controlled diabetes mellitus in adolescence, nowadays, with better treatment modalities, this syndrome is rarely seen. Is necessary treating physician

should keep a high index of suspicion for this so that proper growth can be accomplished with timely intervention in order to improve the quality of life of these patients.

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