

Mauriac Syndrome: A Rare Complication With an Easy Solution

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Abstract : Mauriac syndrome (MS) is a rare complication of type 1 diabetes mellitus (DM1). It is related to low insulin concentrations. Therefore is a complication mainly found in developing countries. The main clinical features are hepatomegaly, edema, growth and puberty delay, and the presence of elevated transaminases and serum lipids. The MS incidence is decreasing due to the new types of insulin and intensive glycemic control. Therefore is a rare diagnosis in Europe nowadays, being described mainly in developing countries or with socio-economic limitations to guarantee an adequate management of diabetes. Edema secondary to fluid retention is a rare complication of insulin treatment, especially in young patients. Its severity is variable and is mainly related to the start of a proper treatment and the improvement in glycemic control after diagnosis or after periods of poor metabolic control. Edema resolves spontaneously without requiring treatment in most cases. The Pediatric Endocrinology Unit of Hospital Motril could diagnose a 14-year-old girl who presented very poor metabolic control during the last 3 years as a consequence of the socioeconomic conditions of the country of origin during the last years. Presents up to 4 admissions for ketoacidosis during the last 12 months. After the family moved to Spain our patient began to be followed up in our Hospital. Initially presented glycosylated hemoglobin figures of 11%. One week after the start of treatment, the patient was admitted in the emergency room due to the appearance of generalized edema and pain in the limbs. The main laboratory abnormalities include: blood glucose 225mg/dl; HbA1C 10.8% triglycerides 543 mg/dl, total cholesterol 339 mg/dl (LDL 225) GOT 124 U/l, GPT 89U/l. Abdominal ultrasound shows mild hepatomegaly and no signs of ascites were shown. The patient presented a progressive improvement with resolution of the edema and analytical abnormalities during the next two weeks. During admission, the family received diabetes education, achieving adequate glycemic control at discharge. Nowadays the patient has a good glycemic control having glycosylated hemoglobin levels around 7%.

Keywords : Mauriac, diabetes, complication, developing countries

Conference Title : ICP 2024 : International Conference on Pediatrics

Conference Location : Singapore, Singapore

Conference Dates : May 02-03, 2024